CHAPTER 42

Skin and subcutaneous tissue

LEARNING OBJECTIVES

To understand:
- the structure and functional properties of skin
- the classification of vascular skin lesions

To be aware of:
- the cutaneous manifestations of generalised disease as related to surgery

To know:
- the classification of benign skin tumours
- the management of malignant skin tumours

FUNCTIONAL ANATOMY AND PHYSIOLOGY OF SKIN

Skin can be divided into an outer layer: the epidermis and an inner layer: the dermis. Deep to the dermis is the hypodermis which is composed of subcutaneous fat and remnants of the panniculus carnosus.

Epidermis

The epidermis is composed of keratinised stratified squamous epithelium and can be further subdivided into five layers: the stratum basale (deepest), the stratum spinosum, the stratum granulosum, the stratum lucidum and the stratum corneum (superificial) (Figure 42.1). It accounts for 5 per cent of the total skin (Summary box 42.1).

Summary box 42.1

Epidermis
- Stratum basale
- Stratum spinosum
- Stratum granulosum
- Stratum lucidum
- Stratum corneum

Dermis
- Papillary layer
- Reticular layer

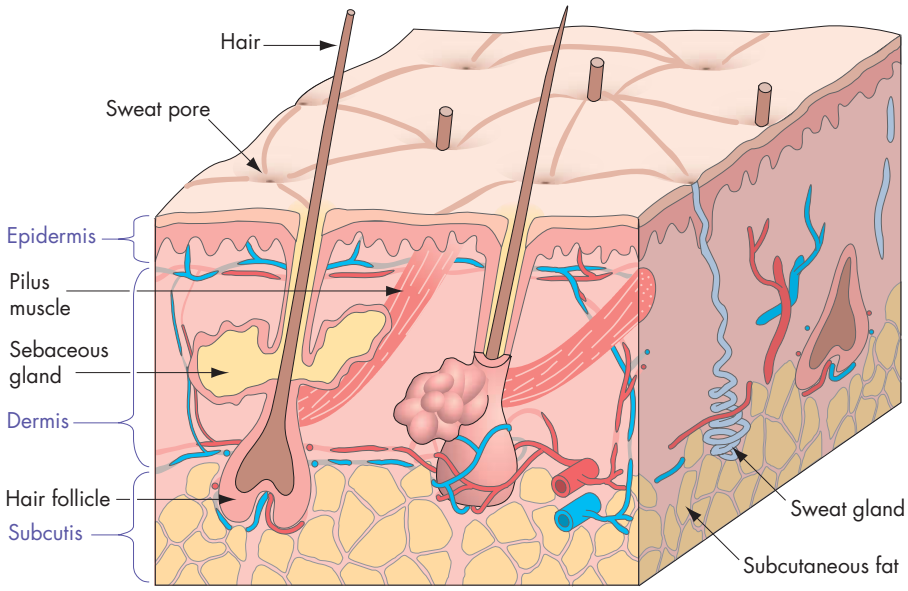

Figure 42.1 Three-dimensional diagram of the structural layers of the skin and its adnexal structures. (Reproduced from Simonsen T, Aarbakke J, Kay I *et al. Illustrated pharmacology for nurses*. London: Hodder Arnold, 2006 with kind permission of the illustrator Roy Lysaa.)

The majority of epidermal cells are keratinocytes arranged in layers. The basal epidermis (stratum basale) also contains melanocytes. Keratinocytes are classified according to their depth in the epidermis and their degree of differentiation. Keratinocytes grow and are replaced via mitoses in the cells of the stratum granulosum as they progress from deep to superficial, losing their nuclei and organelles as they progress upwards before forming the stratum corneum. The other keratinocyte layers in the skin (the strata lucidum; granulosum and spinosum) are variably present according to body site – for instance all three are present in the glaberous skin of the palms and soles of the feet.

Melanocytes are dendritic cells of neural crest origin usually located in the basal epidermis. Each melanocyte synthesises the brown-black pigment melanin, which is transferred via membrane processes to the keratinocytes in the strata granulosum and spinosum. Melanin provides protection against ultraviolet radiation. Ethnic differences in skin colour are determined by variations in the amount, combination and distribution of melanin within the keratinocytes, rather than differences in the number of melanocytes.

Dermis

The dermis comprises 95 per cent of the skin and is structurally divided into two layers. The superficial papillary layer is composed of delicate collagen and elastin fibres in ground substance, into which a capillary and lymphatic network ramifies. The deeper reticular layer is composed of course branching collagen, layered parallel to the skin surface.

The epidermis and dermis meet at the dermoepidermal junction in a three-dimensional wave-like arrangement in which epidermal rete pegs project down and interdigitate with upward-pointing, dermal papillary ridges containing vascular and lymphatic plexi.

The skin also contains specialised cells such as Langerhan's cells, whose role is to engulf antigens and present them to T cells. Merkel cells, Meissner's and Pacinian corpuscles have a role in mechanosensation.

Skin adnexia

Adnexial structures such as hair follicles, sebaceous and sweat glands span both the epidermal and dermal layers and contain some keratinocytes in their ducts. In injuries where epidermis is lost, re-epithelialisation occurs from these structures as well as from the wound margins.

Hair follicles

The human body is covered by fine downy hair (vellus) for three months *in utero*. This is eventually shed before birth, apart from the eyebrows and lashes. Hair which grows out from a hair bulb at the base of a follicle (tubular invaginations of the epidermis) is a shaft of dead keratinised tissue. Strips of smooth muscle (erector pili) are inserted into the wall of the hair follicle and lead to hair elevation in times of stress and cold.

Sebaceous glands

Most are hair follicle appendages situated between each hair follicle and its erector pili muscle. When the erector pili muscle contracts to elevate the hair, it compresses the gland and sebum is released (holocrine secretion). The function of the sebum is to act as a skin lubricant and physical protection barrier.

Sweat glands

Eccrine and apocrine are simple sweat glands that open into pores in hair follicles. Eccrine glands are distributed throughout the entire body surface except on the lips. These glands secrete sweat in response to emotion or as part of thermoregulation. Apocrine glands are found in the axillary and groin areas and become active at puberty. Their secretion, which is characteristically malodourous, varies in response to emotion, hormone secretion and bacterial degradation.

Skin thickness

Skin thickness varies with age and body area. It is thinner in children than in adults in any given region. The dermis is between 15 and 40 times thicker than the epidermis, but starts to thin during the fourth decade as part of the ageing process. The epidermis is thickest on the palms, soles, back and buttocks and thinnest on eyelids (0.5–1 mm on sole of the foot, 0.05–0.09 mm on the eyelid).

Blood supply of the skin

In the last 25 years, the 'angiosome model' has furthered our understanding of the anatomical blood supply of skin and therefore the ability to reconstruct soft tissue defects using vascularised flaps of various tissue compositions. With respect to its blood supply, the body can be envisaged as three-dimensional segments of tissue called angiosomes, each with an arterial supply and a venous drainage. Blood equilibrates and flows between neighbouring angiosomes via 'choke' vessels, which tend to be situated within muscles. Cutaneous arteries, direct branches of segmental arteries (concentrated at the dorsoventral axes and intermuscular septae), perforate the underlying muscles or run directly within fascial layers to the skin from the deep tissues (Figure 42.2).

The blood supply to the skin anastomoses in subfascial, fascial, subdermal, dermal and subepidermal plexi. The epidermis contains no blood vessels so cells there derive nourishment by diffusion.

The venous drainage of the skin is via both valved and un-valved veins. The unvalved veins allow an oscillating flow between cutaneous territories within the subdermal plexus – equilibrating flow and pressure. The valved cutaneous veins drain via plexi to the deep veins.

Anomalies of skin metabolism

Blood flow to the skin can vary between 5 and 100 mL 100 g/min in the temperature range 20–40°C. The skin thus has a potential blood supply that is 20–100 times greater than its metabolic and thermoregulatory requirements. Despite this, the blood supply is inadequate to support wound healing alone – primary closure or granulation tissue is therefore required for healing to occur.

A teleological explanation for this apparent excess blood supply is in the restitution of mechanical integrity after the myriad injuries such as scratching, stretching, compressing, heating and cooling to which our skin is constantly subjected.

Skin functions optimally at temperatures below body core temperature and can tolerate long periods of ischaemia (allowing it both to be grafted and to be expanded and used in reconstructive surgery, see Chapter 31).

George Meissner, 1829–1905, successively Professor of Anatomy and Physiology, Basle, Switzerland, Professor of Zoology and Physiology, Freiberg, Germany and Professor of Physiology, Göttingen, Germany.

Paul Langerhans, 1847–1888, Professor of Pathological Anatomy, Freiberg, Germany.
Friedrich Sigmud Merkel, 1845–1919, Professor of Anatomy successively at Rostock, Koningsberg (now Kaliningrad in Russia) and Göttingen, Germany.
Filippo Pacini, 1812–1883, Professor of Anatomy and Physiology, Florence, Italy.

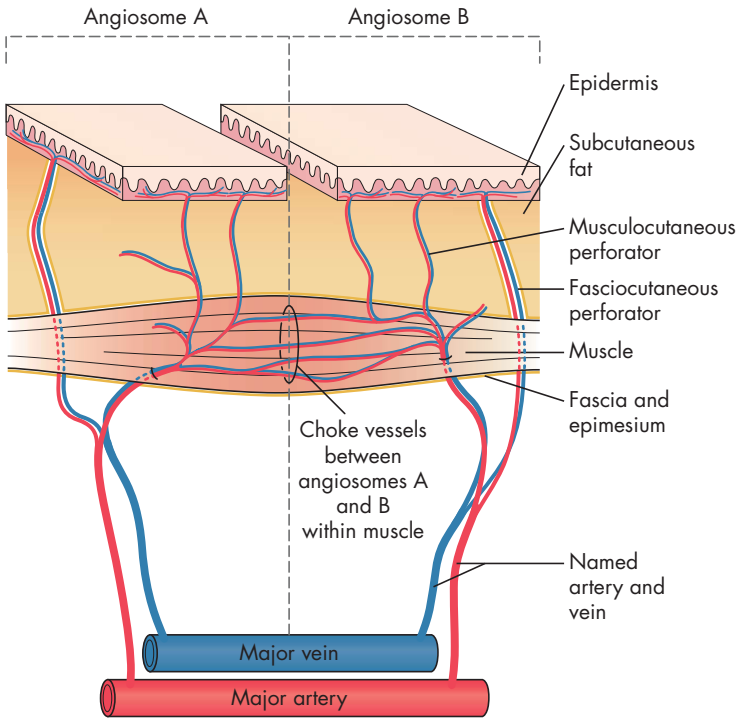

Figure 42.2 Schematic showing two neighbouring angiosomes. Note the choke vessels within the muscle spanning the two cutaneous territories of angiosome A and B – two common examples of myocutaneous flaps which utilise this physiology include the rectus abdominus and the latisimus dorsi flaps.

FUNCTION OF THE SKIN

Human skin and subcutaneous tissue have several important functions (Summary box 42.2).

Summary box 42.2

Function of the skin

- Barrier to the environment: trauma, radiation, pathogens
- Temperature and water homeostasis
- Excretion (e.g. urea, sodium chloride, potassium, water)
- Endocrine and metabolic functions
- Sensory organ for pain, pressure, movement

Skin grafts

Grafts of the skin can be used to reconstruct wounds having been harvested as split (leaving some epidermal components) or full thickness. The process by which a skin graft adheres to and heals a wound is a unique and unnatural process, in which normal wound healing at the recipient site is altered by the presence of the graft. The survival of a skin graft is largely dependent on how fast the graft derives new blood supply from the wound on which it is placed. Until the graft establishes a new blood supply, nutrition is derived by diffusion through the fibrin layer formed between it and the wound bed. After 48–72 hours, a fine capillary network grows into the graft and anastomoses with the native vasculature of the graft. Factors that inhibit this process (haematoma, seroma or bacterial exudates) will decrease the likelihood that the graft will successfully 'take' (see Chapter 31).

Ulcers

An ulcer is a discontinuity of an epithelial surface. It is characterised by progressive destruction of the surface epithelium and a granulating base. Ulcers can be classified as non-specific, specific and malignant (Table 42.1 and Figure 42.3).

Sinus

A sinus is a blind-ending tract that connects a cavity lined with granulation tissue (often an abscess cavity) with an epithelial surface. Sinuses may be congenital or acquired (Table 42.2). Congenital sinuses arise from the remnants of embryonic ducts that persist instead of being obliterated and involuted during embryonic development. Acquired sinuses occur as a result of the presence of a retained foreign body (for example suture material), specific chronic infection (for example tuberculosis (TB) or actinomycosis), malignancy or inadequate drainage of the cavity (Figure 42.4).

Treatment of the sinus is directed at removing the underlying cause. Biopsies should always be taken from the wall of the sinus to exclude malignancy or specific infection.

For specific management of the disease conditions please refer to the appropriate chapter.

Fistula

A fistula is an abnormal communication between two epithelium lined surfaces. This communication or tract may be lined by granulation tissue but may become epithelialised in chronic cases. Fistulas may be congenital or acquired.

Examples of congenital fistula include tracheo-oesophageal and branchial fistulas. Acquired fistulas include fistula in ano, enterocutaneous fistula following Crohn's disease or postoperative anastomotic complications, arteriovenous fistula which may

Table 42.1 Classification of common types of ulcer.

Ulcer	Type
Peptic	Non-specific
Pressure sores (decubitus ulcers) and ischaemic ulcers	Non-specific
Gravitational ulcers – venous insufficiency	Non-specific
Secondary infective – wound infection and abscess drainage	Non-specific
Traumatic ulcers	Non-specific
Neuropathic ulcers – diabetes, tabes dorsalis, leprosy	Non-specific
Iatrogenic – intravenous fluid extravasation	Non-specific
Dermatitis artefacta – self-mutilation	Non-specific
Aphthous	Non-specific
Primary infective – herpes simplex, tuberculosis, fungal, syphilis	Specific
Gastrointestinal tract and skin	Malignant

Table 42.2 Sinuses.

Congenital	Acquired
Preauricular	Post surgical (abdominal or perineal)
Umbilical	Pilonidal
Urachal	Suture
Coccygeal	TB
Sacral	Actinomycosis
	Osteomyelitis
	Crohn's disease

be traumatic or iatrogenic (for haemodialysis).

Again, management of the fistula is directed to treating the underlying aetiology (see appropriate chapter).

PATHOPHYSIOLOGY OF THE SKIN AND SUBCUTANEOUS TISSUES

Radiation damage

Ultraviolet radiation (UVR) and ionising radiation (IR) damage cellular DNA via the tumour suppressor gene *p53*, inhibiting cellular repair and apoptotic mechanisms. There is also evidence that efferent immune responses are impaired after skin exposure to ultraviolet radiation facilitating neoplasia.

Ultraviolet radiation

UVR is divisible into A, B and C according to wavelength. UVR is the principal cause of skin cancer in all skin types. Its effects are attenuated by melanin and there is an inverse relationship between melanin content and skin susceptibility to UV-induced neoplasia. Some protection is afforded by the stratum corneum, which reflects and refracts UVR and by clothing, protective creams, cloud cover and buildings.

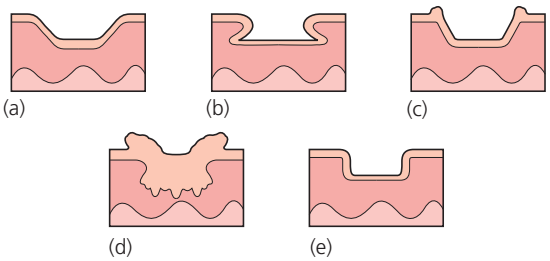

Figure 42.3 Some characteristic shapes of the edges of ulcers. (a) Non-specific ulcer: note the shelving edge. (b) Tuberculous ulcer: note the undermined edge. (c) Basal cell carcinoma (rodent ulcer): note the rolled edge, which may exhibit small blood vessels. (d) Epithelioma: note the heaped-up, everted edge and irregular thickened base. (e) Syphilis: note the punched-out edge and thin base, which may be covered with a 'wash-leather' slough.

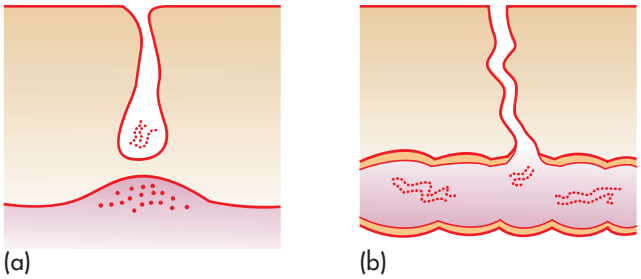

Figure 42.4 A sinus (a) and a fistula (b); both usually arise from a preceding abscess. (a) This is a blind track, in this case a pilondial abscess. (b) This is a track connecting two epithelium-lined surfaces, in this case a colocutaneous fistula from colon to skin.

Ionising radiation

The effects of IR are dose and time dependent. The skin with its rapid cellular turnover exhibits signs soon after exposure. High frequency rays cause electron coupling at the molecular level damaging proteins, polysaccharides and lipids.

Infrared radiation

Infrared radiation generates heat with cumulative exposure posing the risk of thermal burns (see Chapter 30).

Congenital/genetic disorders

Neurofibromatosis

There are two distinct neurofibromatosis syndromes where Schwann cells form tumours (Figure 42.5). Each is caused by different genes on different chromosomes. Seventy per cent are

Friedrich Theodor Schwann, 1810–1882, Professor of Anatomy successively at Louvain (1839–1848) and Liege, Belgium (1849–1880). Original researches before the age of 27 laid the foundation of physiology of nerve and muscle. The first to deal with problems related to living matter on a purely physical and chemical basis, and to recognise the cell as the unit of living matter. Discoverer of pepsin, and role of living organisms in fermentation.

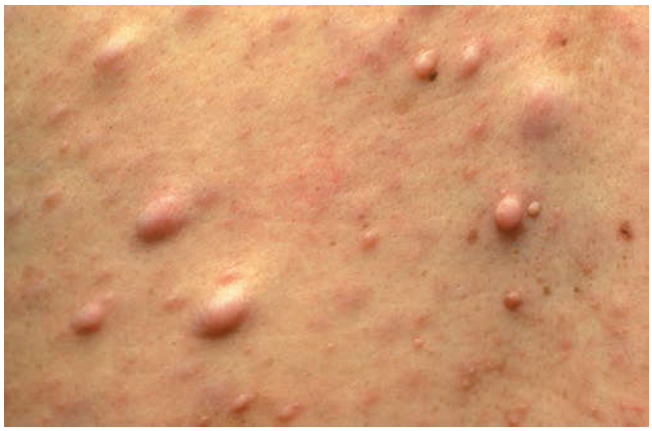

Figure 42.5 Neurofibromatosis (courtesty of St John's Institute for Dermatology, London, UK).

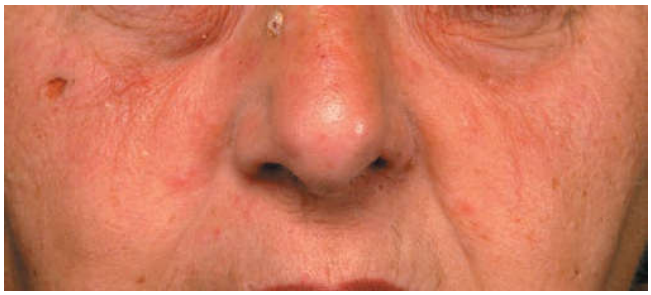

Figure 42.6 Mid-face of a patient with Ferguson–Smith syndrome. Note the two tumours at different stages of development: on the nose and the right cheek (courtesy of St John's Institute for Dermatology, London, UK).

autosomal dominant and 30 per cent arise from sporadic mutations. Neurofibromatosis (NF) 1 or von Recklinghausen's disease is the more common variant, affecting approximately 1:4000 births. It arises from a gene mutation on chromosome 17. Skin manifestations can appear in early life, with the development of more than five smooth-surfaced café-au-lait spots, subcutaneous neurofibromata, armpit or groin freckling and Lisch nodules.

Naevoid basal cell carcinoma (Gorlin's) syndrome

This is an autosomal dominant inherited condition caused by an abnormality in the tumour suppressor gene on chromosome 9q22-31 that codes for the 'patched' protein. Ninety per cent of patients develop multiple basal cell carcinomas. Patients may also exhibit specific phenotypical characteristics including overdeveloped supraorbital ridges, broad nasal roots, hyperteliorism, bifid ribs, scoliosis, brachymetacarpalism, palmar pits and molar odontogenic cysts.

Xeroderma pigmentosum

Described by Kaposi in 1874, this syndrome is caused by an abnormality on the 'patched' gene of chromosome 9q resulting in aberrant nucleotide repair during cellular DNA maintenance. It has an autosomal recessive inheritance confering >2000-fold increase in skin cancer risk. Sufferers have an intolerance to UVR manifested as erythema, pigmentation and photophobia. This leads to premature skin ageing and the development of multiple neoplasms, with most affected individuals dying in early adulthood from metastatic disease (60 per cent mortality by 20 years of age).

Gardner's syndrome

An autosomal dominant disease variant of familial adenomatous polyposis (FAP) is caused by an abnormal gene on chromosome 5. Gardner's syndrome can cause the development of cutaneous pathology such as multiple epidermoid cysts and lipomata.

Ferguson-Smith syndrome

This is a rare, autosomal-dominantly inherited abnormality on chromosome 9q (Figure 42.6). This results in a syndrome that can be traced to a single familial line from western Scotland with affected individuals developing multiple self-healing squamous cell carcinomas.

Cutaneous manifestations of generalised disease

Many diseases have cutaneous manifestations that may present in surgical practice. These include necrobiosis lipoidica, granuloma annulare in diabetes mellitus and pyoderma gangrenosum in inflammatory bowel disease.

Hyperhydrosis

This condition involves excessive eccrine sweating of the palms, soles of the feet, axillae and groins. This can cause functional and social problems, but can be controlled depending on severity, with anti-perspirants or periodic local injections with botulinum toxin A. More resistant cases are treated by laparoscopic cervical sympathectomy.

Lipodystrophy

Lipodystrophy (lipoatrophy) is a localised or generalised loss of fatty tissue which can have primary or secondary causes. It is most commonly seen as a complication of long-term administration of insulin, following treatment of HIV with protease inhibitors or in transplant recipients.

It can be treated in selected cases by autologous fat grafting, injections of poly-L-lactic acid and free tissue transfer.

Inflammatory conditions

Hidradenitis suppurativa

This is a chronic inflammatory disease culminating in suppurative skin abscesses, sinus tracts and scarring (Figure 42.7). It most commonly occurs in the apocrine gland containing skin, namely in the axillary and groin areas. Less common sites include the scalp, breast, chest and perineum.

Hidradenitis suppurativa appears to have a genetic predisposition with variable penetrance, and is strongly associated with obesity and smoking. Women are four times more likely affected than men.

The pathophysiology involves follicular occlusion followed by folliculitis and secondary infection with skin flora (usually *Staphylococcus aureus* and *Propionibacterium acnes*). Clinically, patients develop tender, subcutaneous nodules which may not

Friedrich von Recklinghausen, 1833–1910, Professor of Pathology, Strasbourg, France, described generalised neurofibromatosis in 1882.
Karl Lisch, 1907–1999, ophthalmologist, Wörgl, Austria.
Moritz Kaposi, 1837–1902, Professor of Dermatology, Vienna, Austria.
John Francis Ferguson-Smith, 1888–1978, dermatologist, The Victoria Infirmary, Glasgow, UK.
Eldon J Gardner, 1909–1989, Professor of Zoology, Utah State University, Salt Lake City, UT, USA.

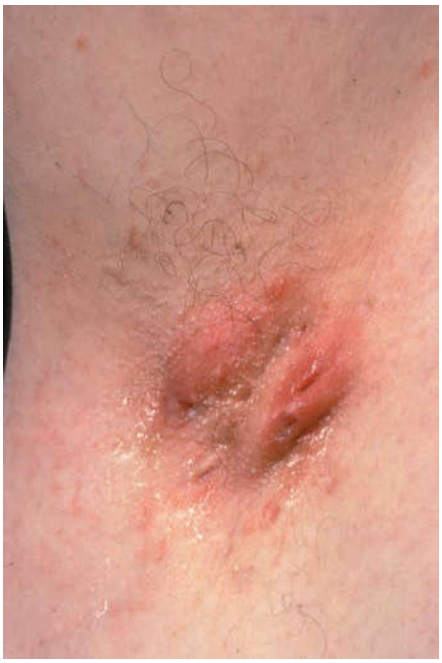

Figure 42.7 Hidradenitis suppurativa affecting the axilla (courtesy of St John's Institute for Dermatology, London, UK).

point and discharge, but which usually progress to cause chronic inflammation and scarring.

Management

Patients should be advised to stop smoking and lose weight where appropriate. Symptoms can be reduced by the use of antiseptic soaps, tea tree oil, non-compressive and aerated underwear.

Medical treatments include topical and oral antibiotics and anti-androgen drugs.

In selected cases, patients may require radical excision of the affected skin and subcutaneous tissue with reconstruction. Healing by secondary intention more frequently leads to contracture and functional impairment than when plastic surgical techniques, such as skin grafting or flap transposition are employed.

Pyoderma gangrenosum

Pyoderma gangrenosum is characterised by cutaneous ulceration with purple undermined edges (Figure 42.8). It is secondary to heightened immunological reactivity, usually from another disease process such as inflammatory bowel disease; rheumatoid arthritis, non-Hodgkin's lymphoma or Wegener's granulomatosis.

Cultures from ulcers often grow Gram-negative streptococci. These skin lesions generally respond to steroids. Surgery is rarely indicated and may exacerbate the condition.

Infections

Skin and soft tissue infections can be localised or spreading, necrotising or non-necrotising. Localised or spreading, non-necrotising infections usually respond to broad-spectrum antibiotics. Localised necrotising infections will need surgical debridement as well as antibiotic therapy. Spreading, necrotising soft tissue infection constitutes a life-threatening surgical

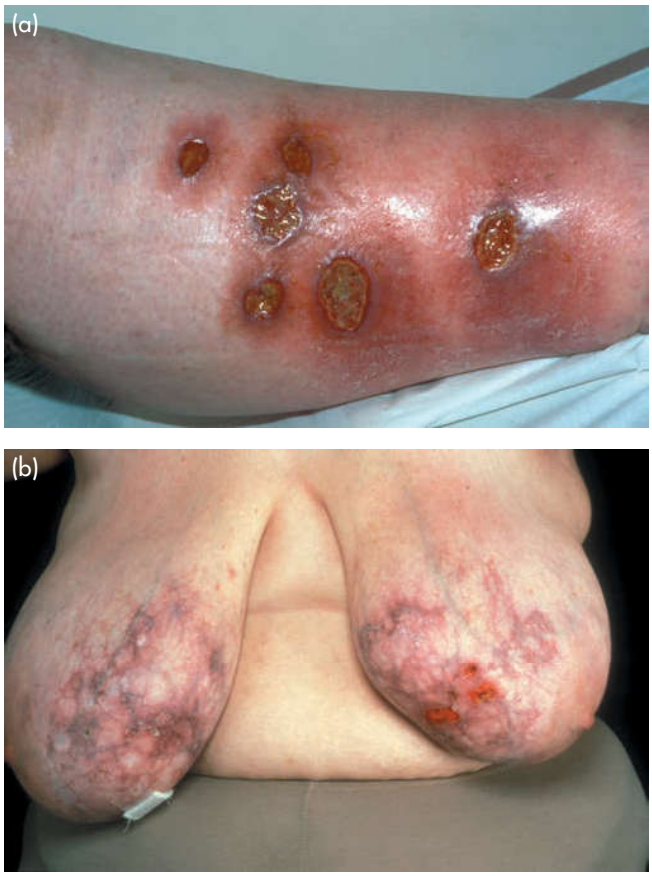

Figure 42.8 Pyoderma gangrenosum affecting the legs (a) and the breasts (b) (courtesy of St John's Institute for Dermatology, London, UK).

emergency, requiring immediate resuscitation, intravenous antibiotic therapy and urgent surgical intervention with radical debridement.

Impetigo

Impetigo is a superficial skin infection with staphylococci, streptococci or both (Figure 42.9). It is highly infectious and usually affects children. Impetigo is characterised by blisters that rupture and coalesce to become covered with a honey-coloured crust. Treatment is directed at washing the affected areas and applying topical anti-staphyloccocal treatments, and broad-spectrum oral antibiotics if streptococcal infection is implicated.

Erysipelas

Erysipelas is a sharply demarcated streptococcal infection of the superficial lymphatic vessels, usually associated with broken skin on the face (Figure 42.10). The area affected is erythematous and oedematous. The patient may be febrile and have a leukocytosis. Prompt administration of broad-spectrum antibiotics after swabbing the area for culture and sensitivity is usually all that is necessary.

Cellulitis/lymphangitis

This is a bacterial infection of the skin and subcutaneous tissue that is more generalised than erysipelas (Figure 42.11). It is usually associated with broken skin or pre-existing ulceration. Cellulitis is characterised by an expanding area of erythematous,

Thomas Hodgkin, 1798–1866, Curator of the Museum and Demonstrator of Morbid Anatomy, Guy's Hospital, London, UK.

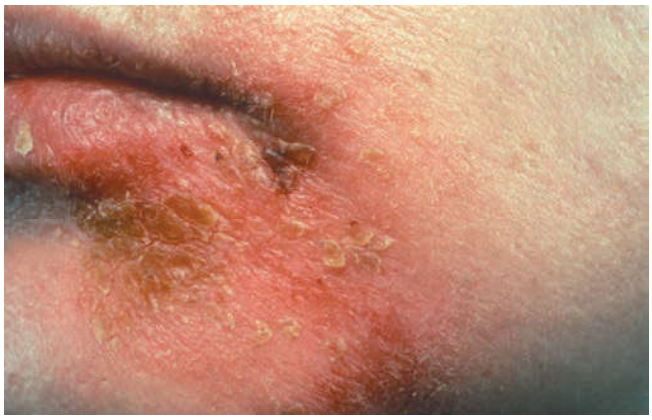

Figure 42.9 Impetigo. Note the honey-coloured crust (courtesy of St John's Institute for Dermatology, London, UK).

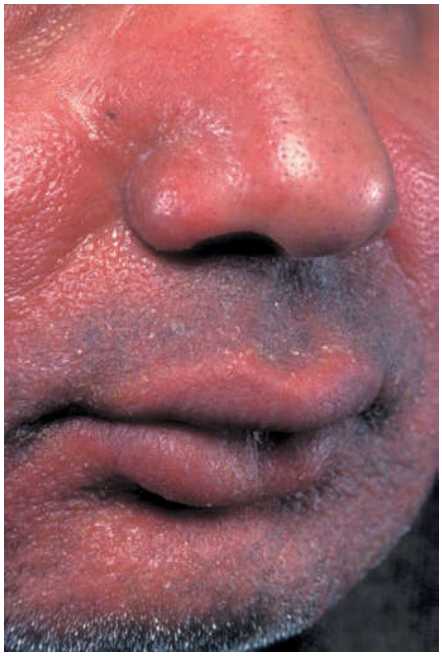

Figure 42.10 Erysipelas (courtesy of St John's Institute for Dermatology, London, UK).

oedematous tissue that is painful and associated with a fever, malaise and leukocytosis. Erythema tracking along lymphatics may be visible (lymphangitis). The most common causative organism is streptococcus. Blood and skin cultures for sensitivity should be taken before prompt administration of broad-spectrum intravenous antibiotics and elevation of the affected extremity.

Necrotising fasciitis

Necrotising fasciitis was first described by Paré in the sixteenth century. Meleney's synergistic gangrene and Fournier's gangrene are all variants of a similar disease process (Summary box 42.3).

Necrotising fasciitis results from a polymicrobial, synergistic infection, most commonly a streptococcal species (group A β haemolytic) in combination with *Staphylococcus*, *Escherichia coli*, *Pseudomonas*, *Proteus*, *Bacteroides* or *Clostridia*. Eighty per cent have a history of previous trauma/infection and over 60 per cent

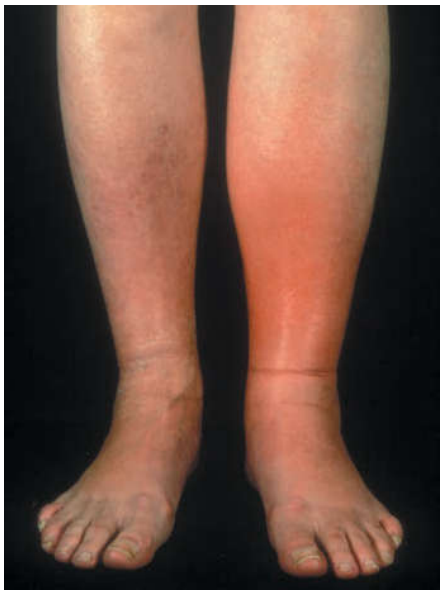

Figure 42.11 Cellulitis affecting the left leg (courtesy of St John's Institute for Dermatology, London, UK).

Summary box 42.3

Necrotising fasciitis

- Surgical emergency
- Polymicrobial synergistic infection
- 80 per cent history of previous trauma or infection
- Rapid progression to septic shock
- Urgent resuscitation, antibiotics and surgical debridement
- Mortality 30–50 per cent

commence in the lower extremities. Predisposing conditions include:

- diabetes;
- smoking;
- penetrating trauma;
- pressure sores;
- immunocompromised states;
- intravenous drug abuse;
- perineal infection (perianal abscess, Bartholin's cysts);
- skin damage/infection (abrasions, bites, boils).

Classical clinical signs include: oedema stretching beyond visible skin erythema; a woody hard texture to the subcutaneous tissues; an inability to distinguish fascial planes and muscle groups on palpation; disproportionate pain in relation to the affected area with associated skin vesicles and soft tissue crepitus (Figure 42.12). Lymphangitis tends to be absent. Early on, patients may be febrile and tachycardic, with a very rapid progression to septic shock. Radiographs should not delay treatment but if taken, they may demonstrate air in the tissues.

Ambrose Paré, 1510–1590, a French military surgeon who also worked at the Hôtel Dieu, Paris, France. He was regarded as the great official royal surgeon for the kings. He devised a dressing of egg white, oil of roses and turpentine which he applied to the wounds of soldiers successfully. He also developed the ligature as a means to stop bleeding.

Frank Lamont Meleney, 1889–1963, Professor of Clinical Surgery, Columbia University, New York, NY, USA.
Jean Alfred Fournier, 1832–1915, syphilologist, founder of the Venereal and Dermatological Clinic, Hôpital St Louis, Paris, France.

Management should commence with urgent fluid resuscitation, monitoring of haemodynamic status and administration of high-dose broad-spectrum intravenous antibiotics. This is a surgical emergency and the diseased area should be debrided as soon as possible until viable, healthy, bleeding tissue is reached. Early re-look and further debridement is advisable together with the use of vacuum-assisted dressings. Early skin grafting in selected cases may minimise protein and fluid losses. Mortality of between 30 and 50 per cent can be expected even with prompt operative intervention.

Purpura fulminans

This is a rare condition in which intravascular thrombosis produces a rapid skin necrosis with haemorrhagic skin infarction. This progresses rapidly to septic shock and disseminated intravascular coagulation. It is usually seen in children, but can occur in adults. It may be sub-divided into three types based on aetiological mechanism.

Acute infectious purpura fulminans

This is the most common form of purpura fulminans and is caused by either an acute bacterial or viral infection (Figure 42.13). *Neisseria meningitidis* and varicella are the most common causal organisms. Acute infectious purpura fulminans causes an acquired protein C deficiency as endotoxins produce an imbalance in the procoagulant and anticoagulant endothelial activity.

Acute infectious purpura fulminans is most common in children under seven years, following an upper respiratory tract infection or in asplenia. Clinically, an initial petechial rash is observed. This develops into confluent ecchymoses and haemorrhagic bullae, which in turn necrose to form well-demarcated lesions that form hard eschars. Extensive tissue loss is common which often culminates in limb amputation. Acute infectious purpura fulminans is associated with a mortality rate of 40–50 per cent, usually a result of multiorgan failure.

Neonatal purpura fulminans

This is an inherited deficiency of protein C and protein S and primarily affects children causing extensive venous thrombosis of the skin and viscera in the first days of life.

Idiopathic purpura fulminans

Usually follows a viral illness after a latent period before the development of the clinical picture of purpura fulminans.

Skin and soft tissue cysts

Milia

These are tiny hard keratin retention cysts seen both in babies and the elderly after chronic sun exposure damage (Figure 42.14).

Epidermal cysts

These are cysts lined with true stratified squamous epithelium derived from the infundibulum of the hair follicle or traumatic inclusion. They are commonly known as sebaceous cysts and are often found on hairy areas of the body, such as the scalp, trunk and face. They are fixed to the skin and usually have a central punctum that is often indentable (Figure 42.15).

Treatment depends on the clinical state of the cyst. When they are inflamed or infected they should be incised and drained initially, and subsequently removed approximately 6 weeks later once the inflammation and induration has subsided. It is important to excise the cyst in its entirety as failure to do so usually results in recurrence.

Meibomian cysts are epidermal cysts found on the free edge of the eyelid. A chronic Meibomian cyst is called a chalazion. Tricholemmal (pilar/pilosebaceous) cysts can be confused with epidermal cysts, except they are derived from the epidermis of the external root sheath of the hair follicle. Ninety per cent are found in the scalp and 70 per cent are multiple.

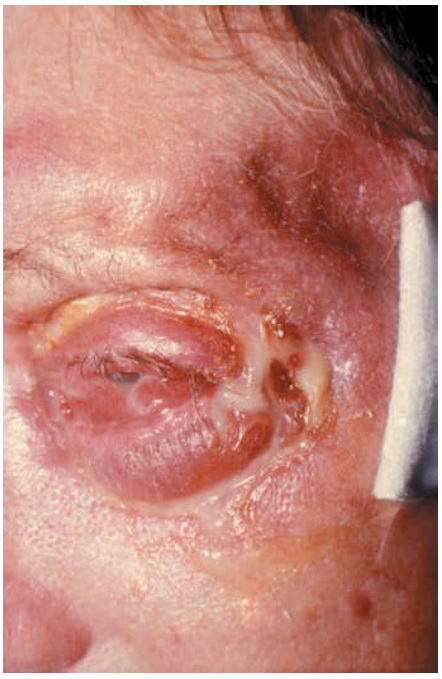

Figure 42.12 Necrotising fasciitis affecting the left orbit and facial skin (courtesy of St John's Institute for Dermatology, London, UK).

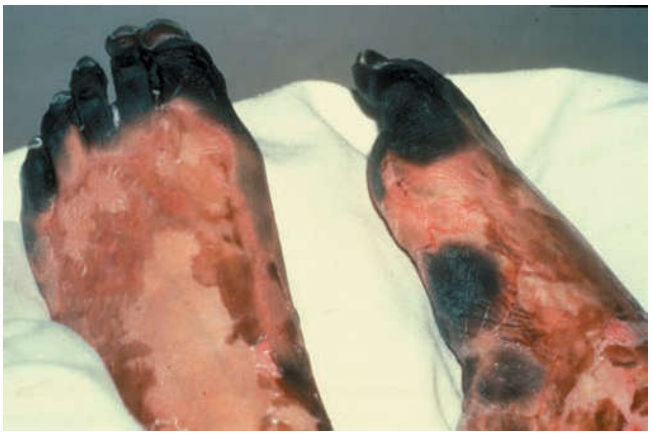

Figure 42.13 Acute infectious purpura fulminans caused by meningococcal septicaemia. Note the sharply demarcated necrotic areas distal to the affected end or perforating arteries with surrounding normal skin (courtesy of St John's Institute for Dermatology, London, UK).

Albert Ludwig Seigmund Neisser, 1855–1916, Director of the Dermatological Institute, Breslau, Germany (now Wroclaw, Poland).
Heinrich Meibom (Meibomius), 1638–1700, Professor of Medicine, History and Poetry, Helmstadt, Germany, described these glands in 1666.

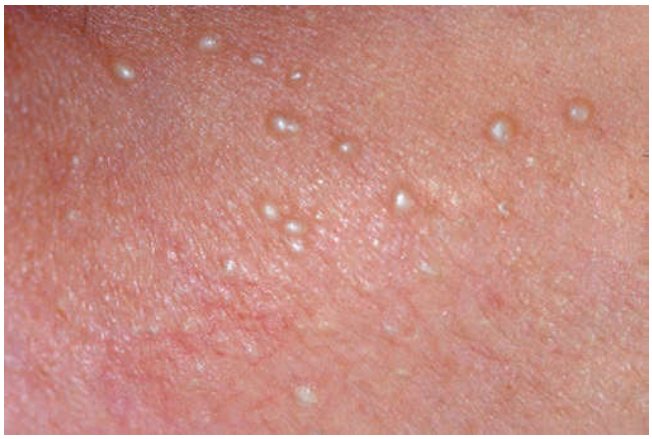

Figure 42.14 Milia (courtesy of St John's Institute for Dermatology, London, UK).

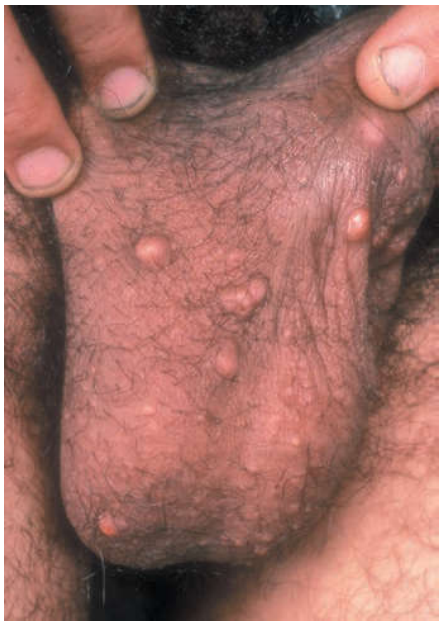

Figure 42.15 Multiple scrotal epidermal cysts (courtesy of St John's Institute for Dermatology, London, UK).

SKIN TUMOURS

Benign lesions

Basal cell papilloma (seborrhoeic keratosis, senile keratosis, verruca senilis)

These are soft warty lesions, which are often pigmented and hyperkeratotic. They are formed from the basal layer of epidermal cells and contain melanocytes. They are one of the most common benign skin tumours in the elderly (Figure 42.16).

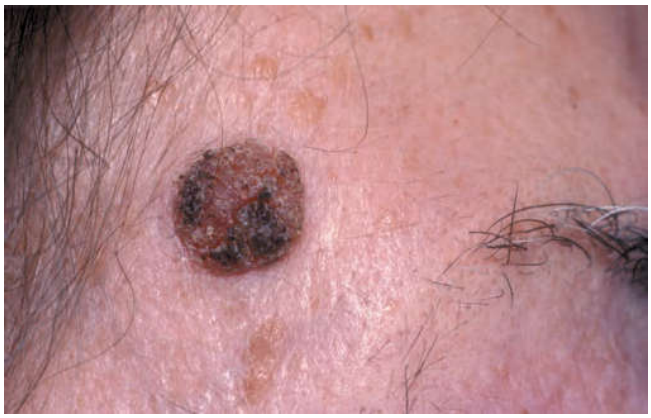

Figure 42.16 Basal cell papilloma (courtesy of St John's Institute for Dermatology, London, UK).

Papillary wart (verruca vulgaris)

This is a benign skin tumour arising from infection with the human papilloma virus, which is also responsible for plantar warts and condylomata acuminata.

Freckle (ephilis)

A freckle is an area of skin that contains a normal number of melanocytes, producing an abnormally large number of melanin granules.

Lentigo

Lentigens are small, sharply circumscribed pigmented macules which are a marker for sun damage and some systemic syndromes. Solar lentigenes are more common in fairer skins. An example of a systemic syndrome associated with lentigenes is Peutz–Jeghers syndrome.

Moles/naevi

Melanocytes migrate from the neural crest to the basal epidermis during embryogenesis. When these melanocytes layer in the epidermis they form a simple mole. Melanocytes that aggregate in the dermis or at the dermoepidermal junction are called naevus cells.

Junctional naevus

A junctional naevus is a deeply pigmented macule or papule that occurs commonly in childhood or adolescence (Figure 42.17). It represents a dermoepidermal proliferation of naevus cells, which

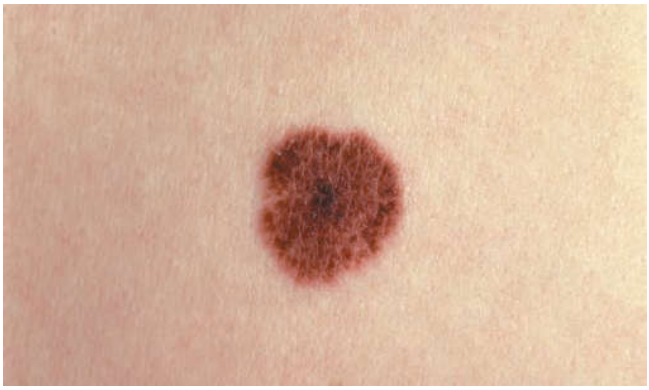

Figure 42.17 Junctional naevus (courtesy of St John's Institute for Dermatology, London, UK).

John Law Augustine Peutz, 1886–1968, Chief Specialist for Internal Medicine, St John's Hospital, The Hague, The Netherlands.
Harold Joseph Jeghers, 1904–1990, Professor of Internal Medicine, New Jersey College of Medicine and Dentistry, Jersey City, NJ, USA.

usually progresses to form a compound or intradermal naevus with advancing age. It may be found on any part of the body but has no malignant potential. Benign mucosal lesions tend to be junctional naevi.

Compound naevus

This is a maculopapular, pigmented lesion that becomes most prominent during adolescence (Figure 42.18). It represents a junctional proliferation of naevus cells with nests and columns in the dermis.

Intradermal naevus

Intradermal naevi are faintly pigmented papules in adults showing no junctional proliferation but a cluster of dermal melanocytes (Figure 42.19).

Spitz naevus

These are reddish brown (occasionally deeply pigmented) nodules previously termed 'juvenile melanoma' (Figure 42.20). They most commonly occur on the face and legs, growing rapidly initially then remaining static. The differential diagnosis is melanoma and excision biopsy is warranted if there is doubt as to the diagnosis.

Spindle cell naevus

Spindle cell naevi are dense black lesions which contain spindle cells and atypical melanocytes at the dermoepidermal junction. They are commonly seen on the thighs and affect women more frequently. They may have malignant potential.

Halo naevus

The halo of depigmentation around any benign naevus represents an antibody response to melanocytes (Figure 42.21). The importance of this depigmentation is that it may also be a feature of a malignant melanoma. A halo naevus is associated with vitiligo.

Café-au-lait spots

These are coffee-coloured macules of variable size (from a few millimetres to 10 cm) (Figure 42.22). Multiple lesions are associated with neurofibromatosis type 1 and McCune–Albright syndrome. They are more common in dark-skinned races.

Naevus spilus

This is also known as speckled lentiginous naevus (Figure 42.23). It is similar in appearance to a café-au-lait spot but with hyperpigmented speckles throughout. It is a benign lesion that is associated with various cutaneous diseases. The mainstay of management is observation and serial photography as malignant transformation is rare.

Mongolian spot

A Mongolian spot is a congenital blue-grey macule found over the sacral skin area (Figure 42.24). Pigmentation initially deepens and then regresses completely by age seven years.

Blue naevus

This is a benign skin lesion that is four times more common in children, typically affecting the extremities and face (Figure 42.25).

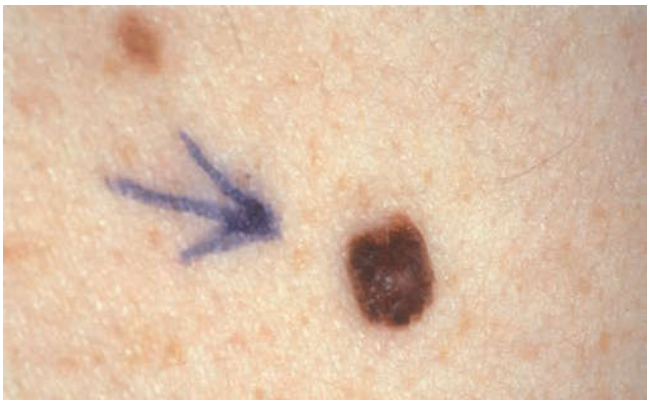

Figure 42.18 Compound naevus (courtesy of St John's Institute for Dermatology, London, UK).

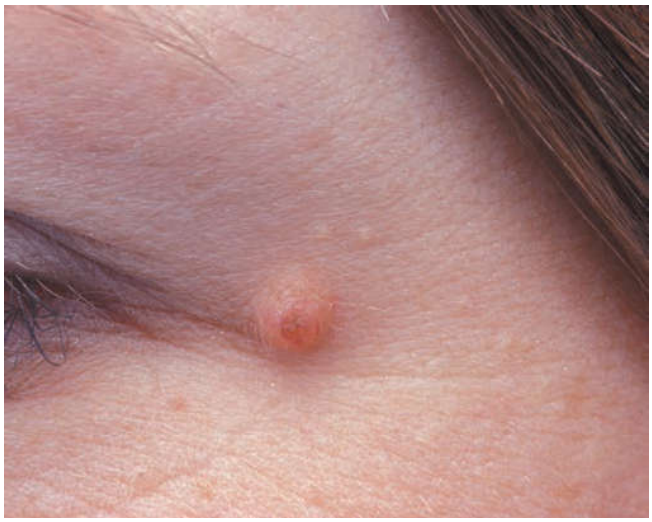

Figure 42.19 Intradermal naevus (courtesy of St John's Institute for Dermatology, London, UK).

Naevi of Ota and Ito

The naevus of Ota is a dermal melanocytic hamartoma with a characteristically blue or grey macule in the trigeminal V_1 and V_2 dermatomes (Figures 42.26 and 42.27). It is four times more common in women and most frequently seen in Oriental and African races.

The Naevus of Ito is characterised by dermal melanocytosis in the shoulder region and can occur simultaneously in a patient with naevus of Ota.

Hair follicles

Trichoepithelioma

These are small skin-coloured nodules most often found in the nasolabial fold. It is clinically and histologically similar to a basal cell carcinoma.

Sophie Spitz, 1910–1956, American pathologist. Dermatopathologist at Sloan-Kettering Cancer Center, published the first case series of 'juvenile melanoma' in 1948. Died at the age of 46 from carcinoma of the colon.

Donovan James McCune, 1902–1976, American paediatrician.
Fuller Albright, 1900–1969, physician, Massachusetts General Hospital, Boston, MA, USA.
Minoru Ito, 1892–1986, Professor of Dermatology, Tohoko University, Sendai, Honshu, Japan.

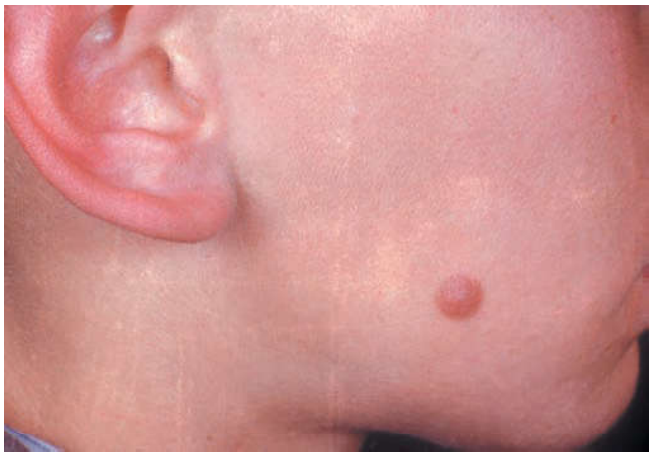

Figure 42.20 Spitz naevus (courtesy of St John's Institute for Dermatology, London, UK).

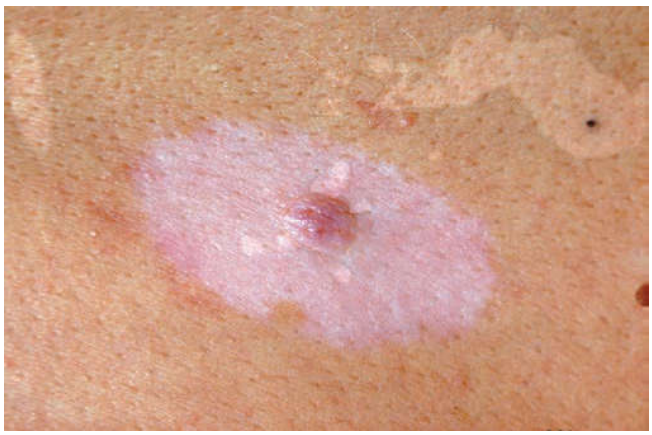

Figure 42.21 Halo naevus (courtesy of St John's Institute for Dermatology, London, UK).

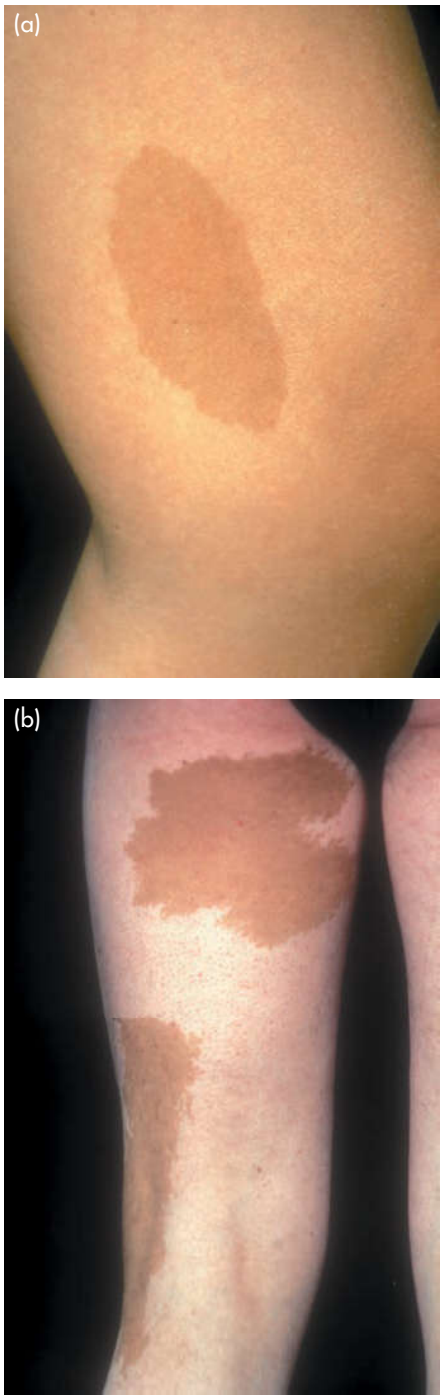

Figure 42.22 Café-au-lait spots. Note the two topographical variants: in (a) the spot has a smooth 'coast of California' border, whereas the upper spot in (b) has an irregular 'coast of Maine' border. Multiple smooth-bordered lesions are commonly associated with syndromes (courtesy of St John's Institute for Dermatology, London, UK).

Pilomatrixoma (calcifying epithelioma of Malherbe)

These are benign tumours of hair matrix cells characterised by basaloid and eosinophilic ghost cells. They commonly calcify and 40 per cent are found in the under-10 age group.

Tricholemmoma (naevus sebaceous of Jadassohn)

Tricholemmoma is a congenital hamartoma with the appearance of a linear verrucous naevus. These are believed to form basal cell carcinomata (BCC) in up to 10 per cent of cases (Figure 42.28).

Adenoma sebaceum (tuberous sclerosis, Bourneville disease)

These are typically red facial papules (angiofibromas) found usually on the nasolabial folds, cheek and chin (Figure 42.29). They form part of the disease process in tuberous sclerosis. These skin lesions usually appear in children less than ten years of age and increase in size and number until adolescence. Cosmetic removal by argon or pulse dye lasers or scalpel is indicated.

Rhinophyma

Rhinophyma is the end-stage sequela of acne rosacea (Figure 42.30). It is a hypertrophy and hyperplasia of the sebaceous glands and tends to affect elderly men (M:F 12:1). Up to 3 per

Josef Jadassohn, 1863–1936, a dermatologist of Breslau, Germany (now Wroclaw, Poland).
Desire M Bourneville, 1840–1909, physician, Le Bicêtre, Paris, France.

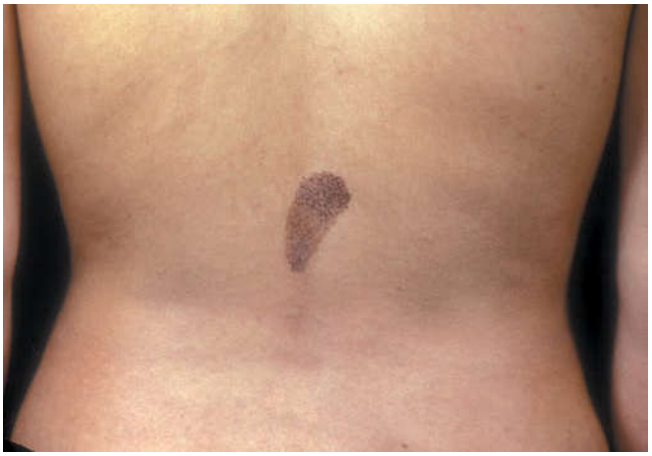

Figure 42.23 Naevus spilus (courtesy of St John's Institute for Dermatology, London, UK).

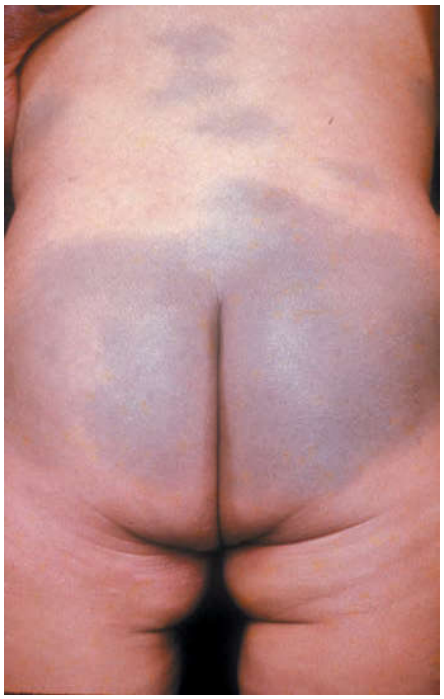

Figure 42.24 Mongolian spot (courtesy of St John's Institute for Dermatology, London, UK).

cent of cases may have an occult BCC. Treatment by dermabrasion or laser resurfacing produces good results.

Sweat glands

Cystadenoma (hydrocystadenomas, hidradenomas)

These are 1–3-cm translucent blue cystic nodules.

Eccrine poroma (papillary syringoma)

These are single raised or pedicled lesions found most often on the palm or sole.

Cylindroma (turban tumour)

A variant of eccrine spiradenoma which can be multiple on the scalp and can coalesce to form a 'turban tumour'.

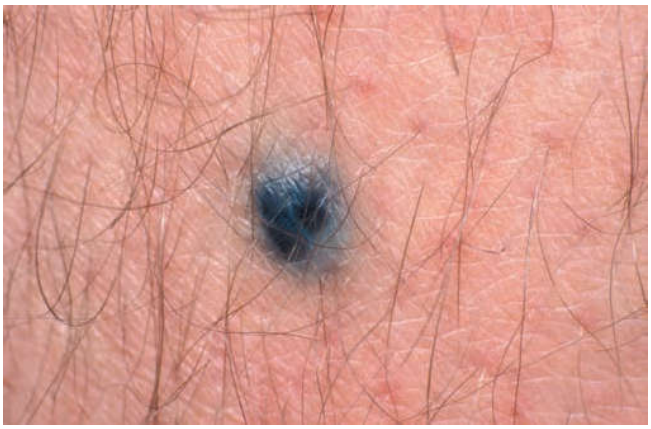

Figure 42.25 Blue naevus (courtesy of St John's Institute for Dermatology, London, UK).

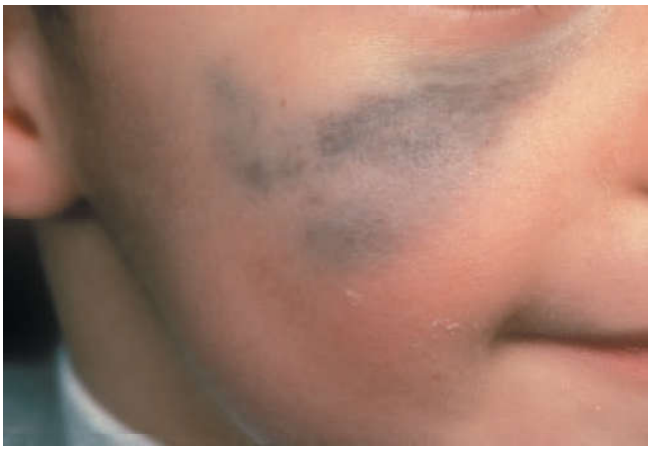

Figure 42.26 Naevus of Ota (courtesy of St John's Institute for Dermatology, London, UK).

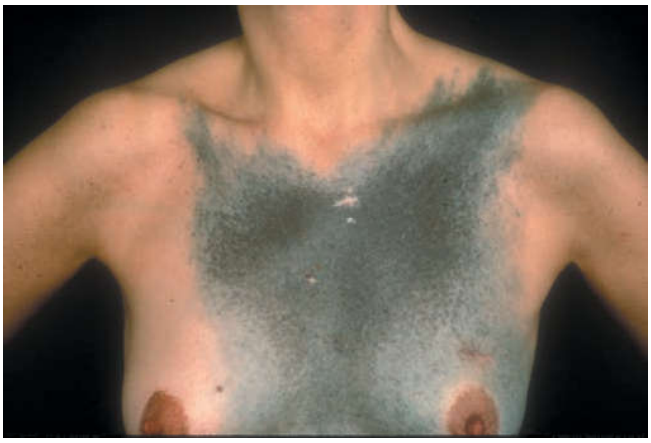

Figure 42.27 Naevus of Ito (courtesy of St John's Institute for Dermatology, London, UK).

Premalignant lesions

Actinic/solar keratosis

These are areas of dyskeratosis and cellular atypia, with subepidermal inflammation, but a normal dermoepidermal junction

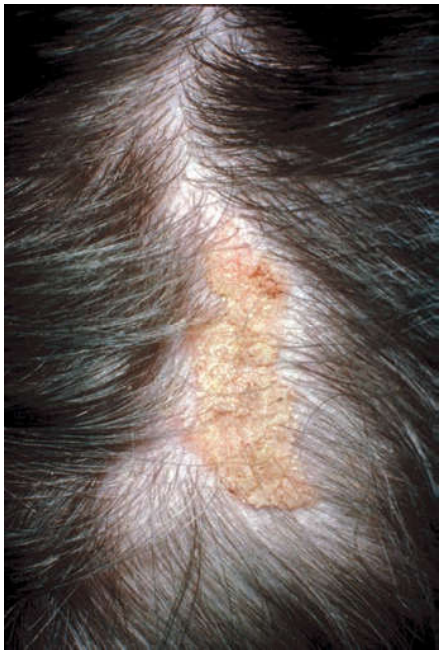

Figure 42.28 Naevus sebaceous of Jadassohn (courtesy of St John's Institute for Dermatology, London, UK).

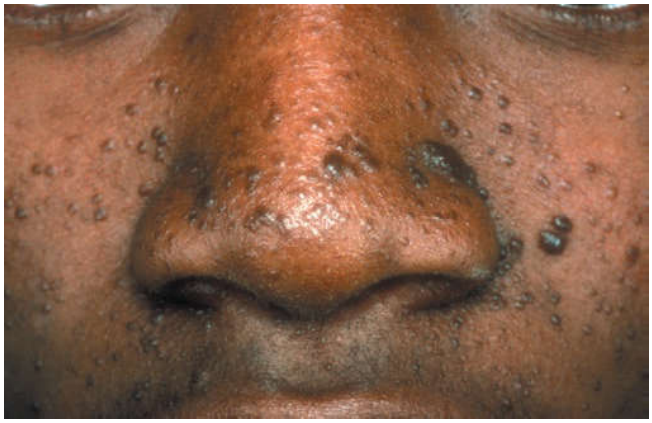

Figure 42.29 Adenoma sebaceum (courtesy of St John's Institute for Dermatology, London, UK).

(Figure 42.31). However, up to 20 per cent go on to form squamous cell carcinomas.

Cutaneous horn

A cutaneous keratin accumulation which by definition has a height greater than its base diameter. Ten per cent will have an underlying squamous cell carcinoma (SCC) (Figure 42.32).

Keratoacanthoma

Classically, this is a symmetrical, cutaneous growth with a central crater filled with a keratin plug (Figure 42.33). It is twice as common in men and is usually found on the face of 50–70 year olds. The aetiology of keratoacanthoma is unclear but may be caused by a papilloma virus in a hair follicle during the growth phase. It has also been associated with smoking and chemical carcinogen exposure.

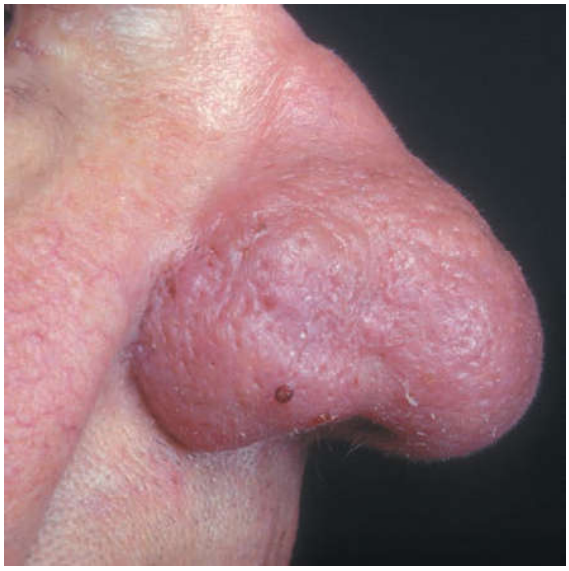

Figure 42.30 Rhinophyma (courtesy of St John's Institute for Dermatology, London, UK).

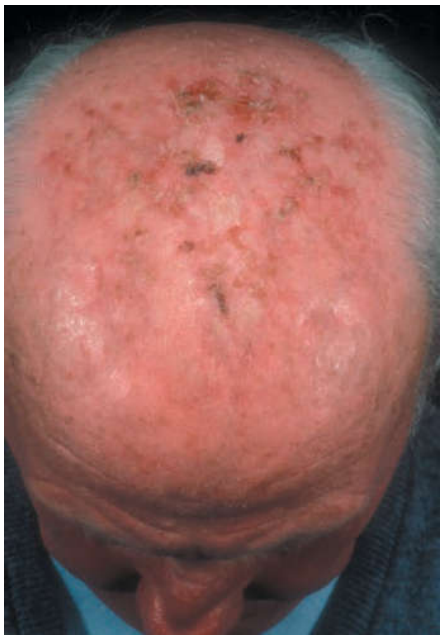

Figure 42.31 Actinic keratosis (courtesy of St John's Institute for Dermatology, London, UK).

Keratoacanthoma can grow to between 1 and 3 cm in a 6-week period and then typically resolve spontaneously over the subsequent six months. Removal of the central keratin plug may speed resolution. Excision is recommended as the differential diagnosis includes anaplastic SCC and the excision scar is often better than that which remains after resolution.

Bowen's disease

Bowen's disease was first described by John T Bowen in 1912. It is a carcinoma *in situ* with between 3 and 11 per cent progressing to SCC (Figure 42.34). It is currently not thought to be a paraneoplastic condition. Chronic solar damage and inorganic

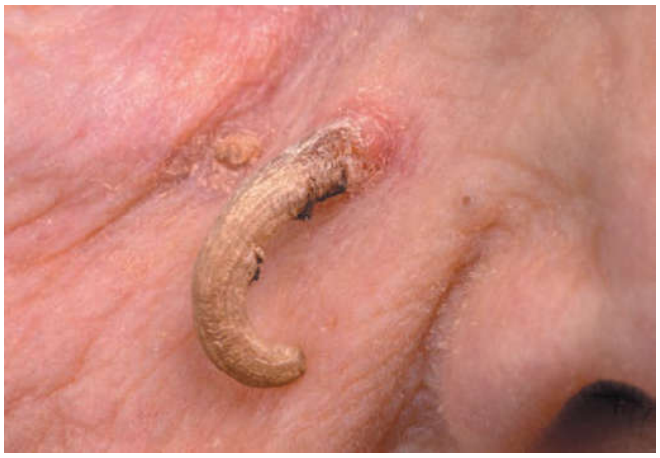

Figure 42.32 Cutaneous horn (courtesy of St John's Institute for Dermatology, London, UK).

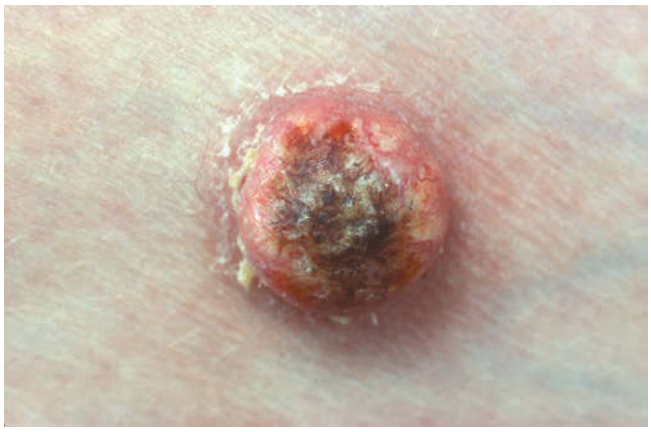

Figure 42.33 Keratocanthoma (courtesy of St John's Institute for Dermatology, London, UK).

arsenic ingestion have been implicated as aetiologic factors in the development of Bowen's disease. The human papilloma virus 16 has also been documented as a cause.

Bowen's disease often presents as a slowly enlarging, erythematous, scaly patch or plaque. It may occur anywhere on the mucocutaneous surface of the body. On the glans penis, it is called erythroplasia of Querat (Figure 42.35).

Topical therapy with 5-fluorouracil or imiquimod are effective treatments. Alternatives include surgical excision with a 4-mm margin or Mohs' micrographic surgery for larger or recurrent lesions.

Extramammary Paget's disease

It is a form of intraepidermal adenocarcinoma, which may occur in the genital, perianal regions or in cutaneous sites rich in apocrine glands such as the axilla (Figure 42.36). Approximately 25 per cent of the cases of extramammary Paget's disease are associated with an underlying *in situ* or invasive neoplasm.

The early skin changes are subtle and may present as an eczematous lesion or intertrigo.

Surgical excision forms the basis of treatment with up to 20 per cent demonstrating invasion on excision.

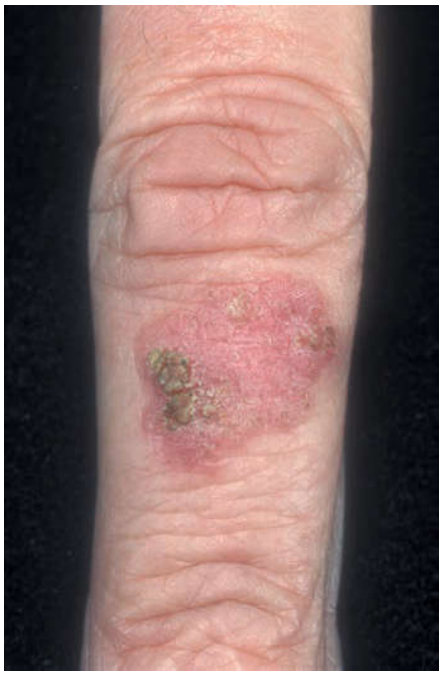

Figure 42.34 Bowen's disease – squamous cell carcinoma *in situ* (courtesy of St John's Institute for Dermatology, London, UK).

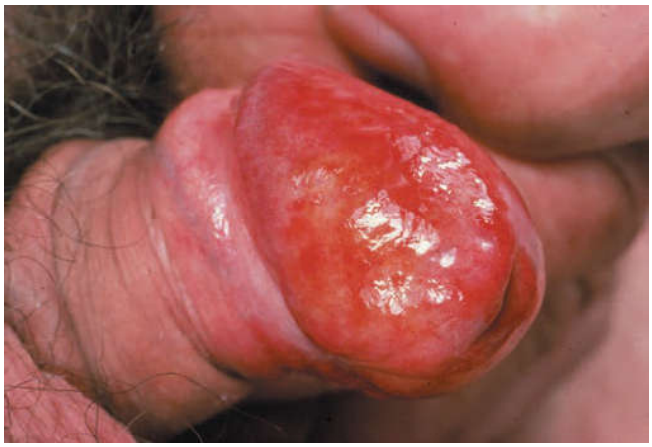

Figure 42.35 Erythroplasia of Queyrat – squamous cell carcinoma *in situ* on the glans penis; also called Paget's disease of the penis (courtesy of St John's Institute for Dermatology, London, UK).

Giant congenital pigmented naevus or giant hairy naevus

The giant congenital pigmented naevus (GCPN) causes a great deal of confusion as its definition and management is contentious. It is a hamartoma of naevo-melanocytes that has a tendency to dermatomal distribution (Figure 42.37). It has a similar histology to compound naevi, but the naevus cells are distributed variably from the epidermis throughout all layers and into the subdermal fat and muscle. There is general agreement that

Frederic E Mohs, 1910–2002, a twentieth century American Surgeon, Physician and General Surgeon, University of Wisconsin, Madison, WI, USA. Developed the Mohs Micrographic Surgical technique in 1938 for cutaneous malignant lesions.

John Templeton Bowen, 1857–1941, Professor of Dermatology, Harvard University Medical School, Boston, MA, USA, described this intradermal precancerous skin lesion in 1912.
August Queyrat, 1856–1933, dermatologist, Paris, France, described this condition in 1911.

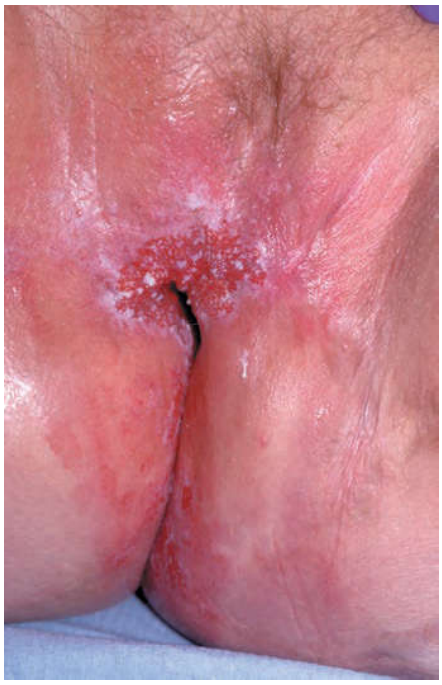

Figure 42.36 Extramammary Paget's disease involving perineum (courtesy of St John's Institute for Dermatology, London, UK).

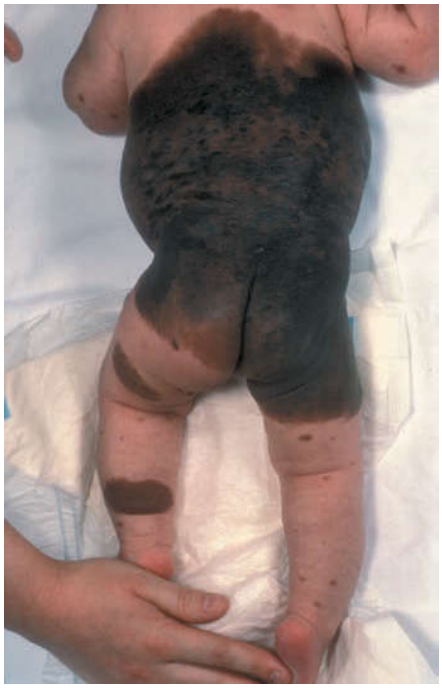

Figure 42.37 Giant congenital pigmented naevus (courtesy of St John's Institute for Dermatology, London, UK).

GCPNs are precursors of melanoma but the magnitude of this risk is unclear largely due to the lack of well-conducted studies and poor classification. A 3–5 per cent lifetime risk of melanoma is quoted. One in three childhood malignant melanomas arise in patients with GCPN but the risk decreases with age with 15 per cent presenting at birth, 62 per cent present by puberty and 99 per cent by 45 years of age.

A multidisciplinary management approach is advocated with initial investigations examining for neurocutaneous melanosis as there may be leptomeningeal involvement. Removal of GCPN should be considered for both aesthetic and oncological reasons.

Dysplastic (atypical) naevus

Dysplastic naevus is an irregular proliferation of atypical melanocytes at the basal layer of epidermis (Figure 42.38). It has variegated pigmentation with irregular borders, measuring more than 5 mm in size. Dysplastic naevus has a familial inheritance and carries a 5–10 per cent risk of forming a superficial spreading melanoma.

Malignant lesions

Basal cell carcinoma

Usually a slow growing, locally invasive malignant tumour of pluripotential epithelial cells arising from basal epidermis and hair follicles, hence affecting the pilosebaceous skin (Summary box 42.4).

Summary box 42.4

Basal cell carcinoma

- Slow growing
- Risk factor – ultraviolet light
- 90 per cent nodular/nodular cystic
- High and low risk basal cell carcinoma

Epidemiology

The strongest predisposing factor to BCC is ultraviolet radiation. The incidence of BCC therefore rises with proximity to the equator, although 33 per cent arise in parts of the body which are not sun exposed. It occurs in the middle aged or elderly with 90 per cent of lesions found on the face above a line from the lobe of the ear to the corner of the mouth. Other predisposing factors include exposure to arsenical compounds, coal tar, aromatic hydrocarbons, ionising radiation and genetic skin cancer syndromes. White-skinned people are almost exclusively

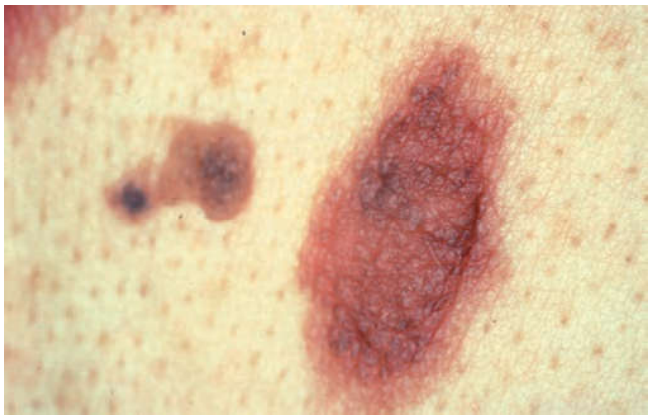

Figure 42.38 Dysplastic naevus (courtesy of St John's Institute for Dermatology, London, UK).

affected. Ninety-five per cent occur between the ages of 40 and 80 years and are more common in men.

Pathogenesis

BCCs have no apparent precursor lesions and their development is proportional to the initial dose of the carcinogen, but not duration of exposure. BCCs rarely metastasise, are hard to culture and resist transplantation. All of this suggests that a multistep mechanism for their development is unlikely, and that mesodermal factors acting as intrinsic promoters coupled with an initiation step is the most likely mechanism.

Macroscopic

BCC can be divided into localised (nodular, nodulocystic, cystic, pigmented and naevoid) and generalised lesions. These lesions can be superficial (multifocal or superficial spreading) or infiltrative (morphoeic, ice pick and cicatrising) (Figure 42.39). Nodular and nodulocystic variants account for 90 per cent of BCC.

Microscopic

Twenty-six histological types have been described. The characteristic finding is of ovoid cells in nests with a single 'palisading' layer. It is only the outer layer of cells that actively divide. This may explain why tumour growth rates are slower than the cell cycle speed would suggest, and why incompletely excised lesions are more aggressive. Morphoeic BCCs synthesise type 4 collagenase and so spread rapidly (Figure 42.40).

Prognosis

There are 'high risk' and 'low risk' BCCs. High risk BCCs are the ones that are large (>2 cm) and located at specific sites (near the eye, nose, ear) and have ill-defined margins. Recurrent tumours and those forming in the presence of immunosuppression are also higher risk.

Management

Treatment can be surgical or non-surgical. Margins should always be assessed and marked under loupe magnification and vary between 2 and 15 mm depending on the macroscopic variant. Where margins are ill-defined, or tissue at a premium (nose, eyes), then either Mohs' micrographic surgery or a two-stage surgical approach with subsequent reconstruction after confirmation of clear margins is advisable. The histological sample must be orientated and marked for pathological examination.

Mohs' micrographic surgery is a method used by dermatological surgeons (dermatologists who have undergone extra training in techniques of cutaneous surgery and histopathology) to excise skin cancer under microscopic control.

It has been demonstrated in suitable skin tumours to minimise recurrence rates and maximise conservation of surrounding normal tissue. This technique is therefore considered the optimal treatment for poorly demarcated, recurrent or incompletely excised BCC (including BCC around the nose, eyes and ears where clearance may be uncertain and significant morbidity is associated with incomplete excision, and where reconstruction with a flap is preferable cosmetically).

Mohs' micrographic surgery (using either frozen section and immunohistochemistry or horizontal paraffin-embedded sections) can also be used for excision of SCC, dermatofibrosarcoma protuberans and lentigo maligna.

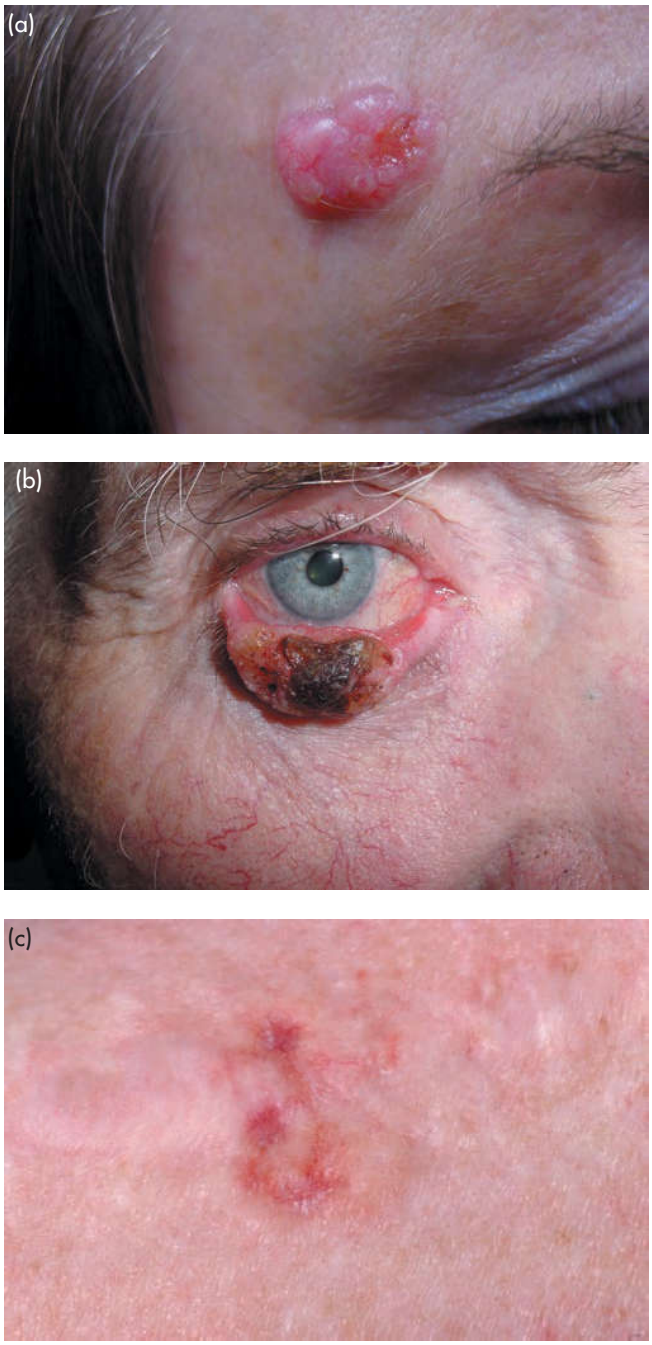

Figure 42.39 (a) A nodulocystic basal carcinoma (BCC). Note the characteristic pearly surface with telangectasia. (b) An ulcerating BCC on the lower eyelid. (c) A recurrent morphoeic BCC ((a) and (b) courtesy of Mr AR Greenbaum; (c) courtesy of St John's Institute for Dermatology, London, UK).

Mohs' micrographic surgery is performed under local anaesthesia (which is one of its limitations) and involves an initial 'saucerising excision' of the primary tumour's gross extent. The sample and the defect are then marked and orientated. A map of the specimen is drawn and characterised using different coloured stains in different quadrants. A histotechnician and a Mohs' surgeon work together, whereby the histotechnician sections the tissue horizontally (including lateral and deep margins

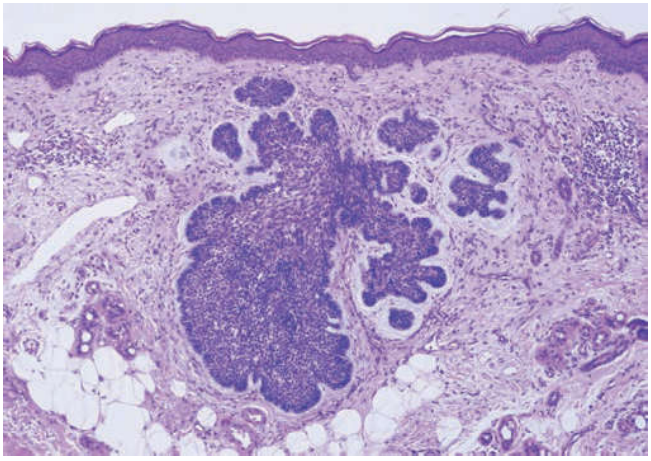

Figure 42.40 A basaloid, epithelial tumour with palisading cells on the periphery of the tumour that sits within a mucinous stroma (courtesy of Dr Catherine di Stefanato, Consultant Dermatopathologist, St John's Institute for Dermatology, London, UK).

in the same slide) and stains it with haematoxylin and eosin. The Mohs' surgeon then examines the slides for the presence of residual tumour and excises more tissue from the relevant parts of the mapped defect as necessary. In theory, the technique offers complete evaluation of the lateral and deep margins of tumour excision and so should thus be dependable. Complete excision rates exceeding 99 per cent are the rule in trained experienced hands.

In the elderly or infirm patients, radiotherapy produces similar recurrence rates to surgery. Superficial tumours can be treated with topical treatments (5-fluorouracil, imquimod) or cryotherapy.

Excision must be complete as there is a 67 per cent recurrence rate if margins are grossly involved and a 33 per cent recurrence rate within two years with microscopic involvement or when reported 'close'. Thus, patients with uncomplicated completely excised lesions can be discharged. Follow up is reserved for patients with tumours in high-risk areas, namely globally sun-damaged skin, syndromes (for example naevoid basal cell carcinoma syndrome) and incomplete excisions in patients who have declined further surgery.

Cutaneous squamous cell carcinoma

SCC is a malignant tumour of keratinising cells of the epidermis or its appendages. It arises from the stratum germinatum of the epidermis and expresses cytokeratins 1 and 10.

Epidemiology

SCC is the second most common form of skin cancer. It is four times less common than BCC and affects the elderly. It is strongly related to cumulative sun exposure and damage, and is twice as common in men and in white-skinned individuals living nearer the equator. SCC is also associated with chronic inflammation (chronic sinus tracts, pre-existing scars, osteomyelitis, burns, vaccination points) and immunosuppression (organ transplant recipients). When an SCC appears in a scar it is known as a Marjolin's ulcer.

The time taken to develop an SCC after radiation exposure is proportional to the wavelength of the radiation. SCC is also caused by chemical carcinogens (arsenicals, tar), and infection with human papilloma virus 5 and 16. There is also evidence that current and previous tobacco use doubles the relative risk of SCC (Summary box 42.5).

Summary box 42.5

Squamous cell carcinoma

- Associated with chronic inflammation
- Invariably ulcerated lesion
- Metastasis in 2 per cent cases

Macroscopic

The appearance of SCC may vary from smooth nodular, verrucous, papillomatous to ulcerating lesions (Figure 42.41). However, all variants will eventually ulcerate as they grow. The ulcers have a characteristic everted edge and are surrounded by inflamed, indurated skin. The differential diagnosis of an SCC are actinic keratosis, BCC, keratoacanthoma, pyoderma gangrenosum and warts.

Microscopic

Characteristic irregular masses of squamous epithelium are noted to proliferate and invade the dermis from the germinal layer (Figure 42.42). This tumour stains positive for cytokeratins 1 and 10. SCC can be histologically graded according to Broders' histological grading. This system describes the proportion of de-differentiated cells (i.e. the ratio of pleomorphic and anaplastic cells:normal cells).

The histopathology report on an SCC should include information on the pathological pattern (e.g. adenoid); the cellular morphology (e.g. spindle); the Broders' grade; the depth of invasion, the presence of any perineural or vascular invasion and the deep and peripheral margin clearance (Table 42.3).

Prognosis

There are several independent prognostic variables for SCC:

1 Invasion:
   a. Depth: the deeper the lesion, the worse the prognosis. For SCC <2 mm, metastasis is highly unlikely; whereas if >6 mm, 15 per cent of SCCs will have metastasised.
   b. Surface size: lesions >2 cm have a worse prognosis than smaller ones.
2 Histological grade: the higher the Broders' grade, the worse the prognosis.
3 Site: SCCs on the lips and ears have higher local recurrence rates than lesions elsewhere and tumours at the extremities fare worse than those on the trunk.
4 Aetiology: SCCs that arise in burn scars, osteomyelitis skin sinuses, chronic ulcers and areas of skin that have been irradiated have a higher metastatic potential.

Albert Compton Broders, 1885–1964, an American pathologist of Minnesota, USA and Chairman of the Department of Surgical Pathology, The Mayo Clinic, Rochester, Minnesota, MN, USA; for one year in 1935 Professor of Surgical Pathology and Director of Cancer Research, University of Virginia, VA, USA. Broders graded rectal cancer in the USA in a manner that Cuthbert Dukes classified them in the UK. A combination of Broders' grading and Dukes' classification gave a more accurate prognosis for rectal carcinoma than either method alone.

Jean-Nicholas Marjolin, 1780–1850, surgeon, Paris, France, described the development of carcinomatous ulcers in scars in 1828.

5 Immunosuppression: SCC will invade further in those with impaired immune response.
6 Tumours with perineural involvement have a worse prognosis and require a wider than usual clearance.

The overall rate of metastasis is 2 per cent for SCC – usually to regional nodes – with a local recurrence rate of 20 per cent.

Management

SCC is a heterogeneous tumour with a malignant behaviour that varies between subtypes. Management must therefore address the need for definitive treatment, the possibility of in-transit metastasis and the tumour's tendency for lymphatic metastasis.

Surgical excision is the only means of providing accurate histology. The margins for primary excision should be tailored to surface size in the first instance. This should ideally be assessed using surgical loupe magnification. A 4-mm clearance margin should be achieved if the SCC measures <2 cm across, and a 1-cm clearance margin if >2 cm.

Ninety-five per cent of local recurrence and regional metastases occur within five years, thus follow up beyond this period is not indicated.

Cutaneous malignant melanoma

Melanoma is a cancer of melanin producing cells and can therefore arise in skin, mucosa, retina and the leptomeninges.

Epidemiology

Cutaneous melanoma is caused largely by exposure to ultraviolet radiation. Its rise in incidence reflects social behaviour and increased recreational activity in the sun among white-skinned races not suited to sun exposure. Although it accounts for less than 5 per cent of skin malignancy, it is responsible for over 75 per cent of skin malignancy related deaths.

Malignant melanoma (MM) accounts for 3 per cent of all malignancy worldwide. It is the most common cancer in young adults (20–39 years) and the most likely cause of cancer-related death.

Distribution between the sexes varies around the world and reflects occupational and recreational exposure to sunlight. Likewise, geographical distribution reflects exposure of white-skinned individuals to sunlight: Auckland in New Zealand currently reports the highest incidence per capita, and before that Brisbane in Australia held that distinction.

Five per cent of all patients with MM will develop a second

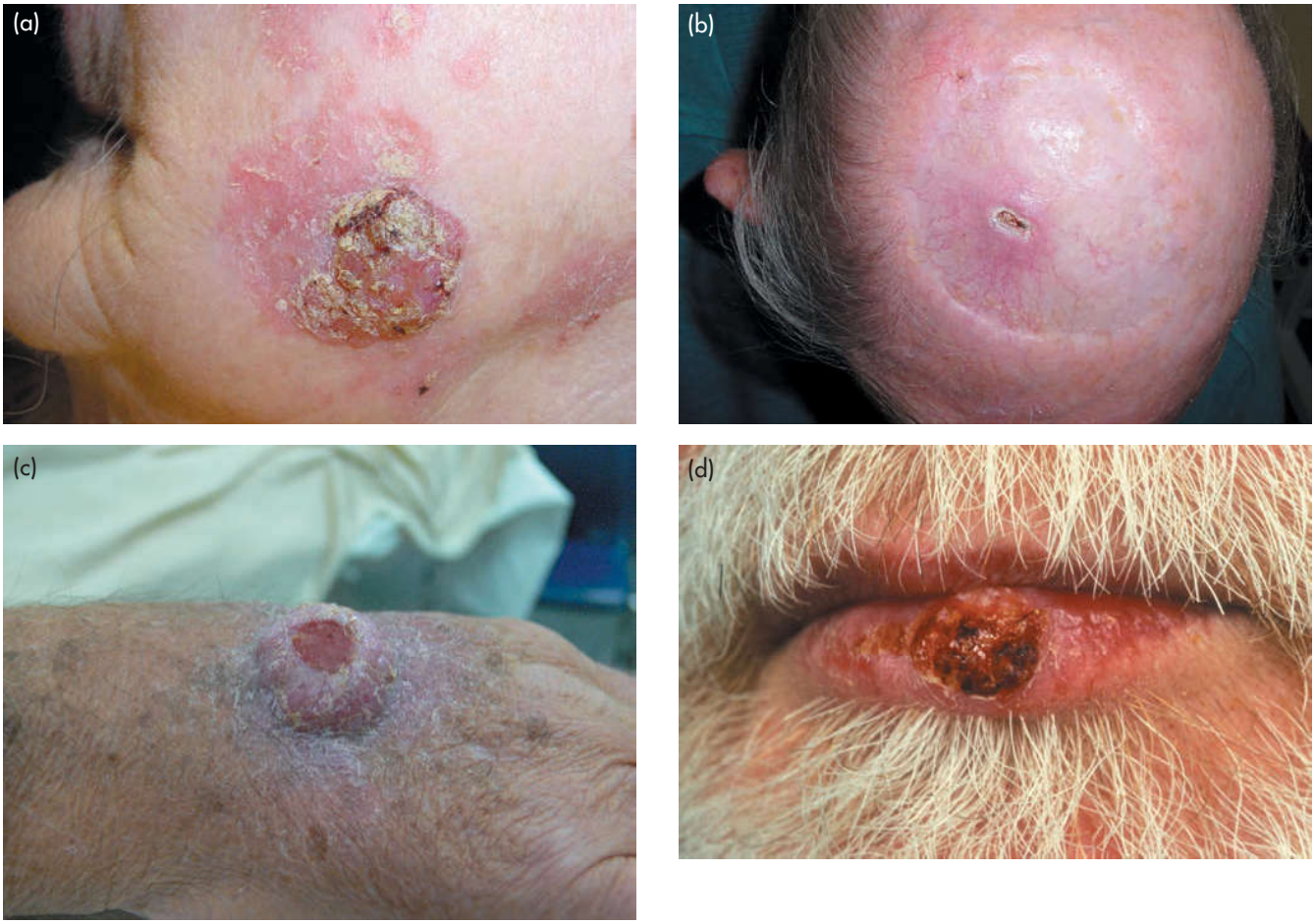

Figure 42.41 (a) A squamous cell carcinoma (SCC) on the face. (b) A recurrent SCC arising in a previously skin-grafted area of the scalp. (c) An SCC arising on the dorsum of the hand in a renal transplant recipient on immunosuppressive therapy. (d) An SCC arising on the lip of a smoker who worked outside on a farm. ((a–c) courtesy of Mr AR Greenbaum; (d) courtesy of St John's Institute for Dermatology, London, UK.)

Table 42.3 TNM classification and staging.

Size	Nodes	Mets	Grade
T_1 = <2 cm	N_0 = no regional nodes	M_0 = no mets	G_1 = low grade
T_2 = 2–5 cm	N_1 = regional nodes	M_1 = distant mets	G_2 = moderately differentiated
T_3 > 5 cm			G_3 = high grade or highly anaplastic
T_4 = muscle or bony invasion			

Stage I = T1, N0, M0; stage II = T2–3, N0, M0; stage III = T4, N0, M0 and any T, N1, M0; stage IV = any T, any N1, M1(+).

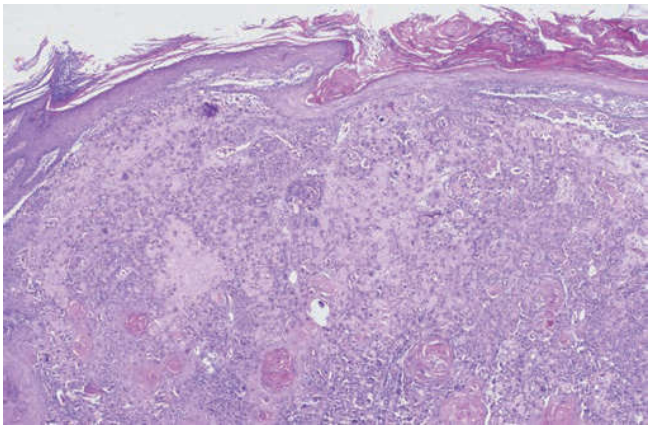

Figure 42.42 An invasive, epidermal keratinising tumour characterised by proliferation of atypical squamous cells with 'horn pearls' (courtesy of Dr Catherine di Stefanato, Consultant Dermatopathologist, St John's Institute for Dermatology, London, UK).

primary melanoma. Seven per cent of MMs present as occult metastases from an unknown primary.

Pathophysiology

UV exposure is the major causal factor for developing MM. Cumulative exposure favours the development of lentigo maligna melanoma (LMM), whereas 'flash fry' exposure – typical of rapidly acquired holiday tans – favours the other morphological variants (Summary box 42.6).

Summary box 42.6

Malignant melanoma

- Rising incidence
- Genetic and acquired risk factors
- Superficial spreading form the most common
- Breslow thickness most important prognostic indicator
- Sentinel node biopsy useful for lymphatic mapping

A small proportion of MMs are genetically mediated, as in xeroderma pigmentosum which increases the relative risk of developing MM to 1000. Immunosuppression secondary to drugs or HIV infection will increase the incidence of MM by 20–30-fold.

The risk factors for developing MM are summarised below:

- Xeroderma pigmentosum (relative risk = 1000)
- Past medical or family history of MM with dysplastic naevi (relative risk = 33–1269)
- Previous melanoma (relative risk = 84)
- High total number of naevi (relative risk = 3.4, if >20 naevi)
- Dysplastic naevi (10 per cent lifetime risk)
- Red hair (relative risk = 3)
- Tendency to freckle (relative risk = 1.9)
- Immune compromised conditions: HIV infection, Hodgkin's disease, cyclosporin A therapy
- Giant congenital pigmented naevus (increased risk)
- History of sunburn – especially in childhood.

Macroscopic

Only 10–20 per cent of MM form in pre-existing naevi, with the remainder arising *de novo* in previously normally pigmented skin. The most likely naevi to form MM are the junctional and compound types.

Macroscopic features in a pre-existing naevus that suggest malignant change are listed in Summary box 42.7.

There are four common macroscopic variants of MM. There are several other notable, but rarer forms.

Summary box 42.7

Macroscopic features in naevi suggestive of malignant melanoma

- Change in size – any adult naevus >6 mm is suspect (for reference a lead pencil diameter is 7 mm) and anything changing to >10 mm is more likely to be malignant than benign
- Shape
- Colour
- Thickness (elevation/nodularity or ulceration)
- Satellite lesions (pigment spreading into surrounding area)
- Tingling/itching/serosanguinous discharge (usually late signs)
- Blood supply: melanomas >1 mm thick have a blood supply which can be found with a hand-held Doppler, so 'Doppler positive' pigmented lesions should be excised

Superficial spreading melanoma

This is the most common type (70 per cent), usually arising in a pre-existent naevus, after several years of slow change, followed by rapid growth in the preceding months before presentation

(Figure 42.43). Typically it is a darker pigmented area in a junctional naevus. Nodularity within superficial spreading melanoma (SSM) heralds the onset of the vertical growth phase.

Nodular melanoma

Nodular melanoma (NM) accounts for 15 per cent of all MM and tends to be more aggressive than SSM, with a shorter clinical onset. These lesions typically arise *de novo* in skin and are more common in men than women, often presenting in middle age and usually on the trunk, head or neck (Figure 42.44). They typically appear as blue/black papules, 1–2 cm in diameter, and because they lack the horizontal growth phase, they tend to be sharply demarcated. Up to 5 per cent are amelanotic.

Lentigo maligna melanoma

Previously also known as Hutchison's melanotic freckle. This variant presents as a slow-growing, variagated brown macule, on the face, neck or hands of the elderly (Figure 42.45). They are positively correlated with prolonged, intense sun exposure, affecting women more than men. They account for between 5 and 10 per cent of MM. LMM are thought to have less metastatic potential than other variants as they take longer to enter a vertical growth phase. Nonetheless, when they have entered the vertical growth phase their metastatic potential is the same as any other melanoma.

Acral lentigious melanoma

Acral lentigious melanoma (ALM) affects the soles of the feet and the palms of the hands. It is rare in white-skinned individuals (2–8 per cent of MM) but is more common in the Afro-Caribbean, Hispanic and Asian populations (35–60 per cent). It usually presents as a flat, irregular macule in later life (Figure 42.46). Twenty-five per cent are amelanotic and may mimic a fungal infection or pyogenic granuloma.

MM under the finger nail is usually SSM rather than ALM. For finger or toe nail lesions it is vital to biopsy the nail matrix rather than just the pigment on the nail plate. A classical feature of a subungual melanoma is Hutchinson's sign. This is nail fold pigmentation which then widens progressively to produce a triangular pigmented macule with associated nail dystrophy. The differential diagnosis is 'benign racial melanonychia', which produces a linear dark streak under a nail in a dark-skinned individual. Malignancy is unlikely if the nail fold is uninvolved.

Miscellaneous

- Amelanotic melanoma (often arising in the gastrointestinal tract and presenting with obstruction, intussusception or as a metastasis from an unknown primary).
- Desmoplastic – mostly found on the head and neck region. It has a propensity for perineural infiltration and often recurs locally if not widely excised. May be amelanotic clinically.

Microscopic

Malignant change occurs in the melanocytes in the basal epidermis, while *in situ*, atypical melanocytes are limited to the dermoepidermal junction and show no evidence of dermal involvement. During the horizontal growth phase, cells spread along the dermoepidermal junction and although they may breach the dermis, their migration is predominantly radial. During the vertical growth phase, the dermis may be invaded.

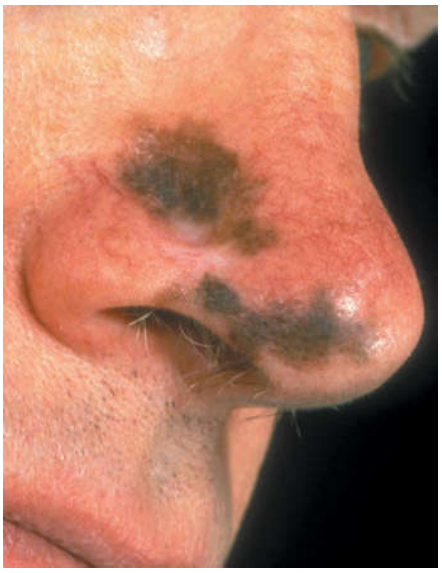

Figure 42.43 Superficial spreading melanoma (courtesy of St John's Institute for Dermatology, London, UK).

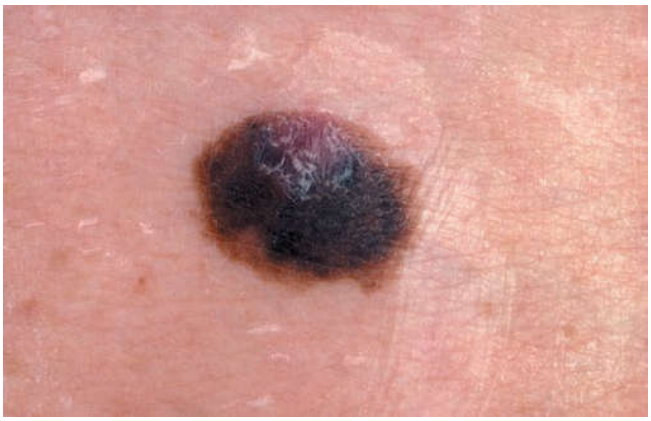

Figure 42.44 Nodular melanoma (courtesy of St John's Institute for Dermatology, London, UK).

The greater the depth of invasion, the greater is the metastatic potential of the tumour.

Management

History and clinical examination should be directed at discovering the primary lesion and identification of local, regional or distant spread. Clinical photography is useful when observation is chosen rather than excision biopsy for definitive histopathological diagnosis. An excision biopsy with a 2–5 mm margin of skin and a cuff of subdermal fat is acceptable. Incision biopsy is occasionally indicated – for instance in large lesions on the face where an excision biopsy of the whole lesion would be disfiguring.

Biopsy and pathological examination provide the first step towards staging melanoma. The Breslow thickness of a melanoma (measured to the nearest 0.1 mm from the granular layer to the

Alexander Breslow, 1928–1980, American pathologist. Pathologist, George Washington University, Washington DC, USA, first reported in 1970 that the prognosis depends upon the thickness of the tumour.

Sir Jonathan Hutchinson, 1828–1913, surgeon, St Bartholomew's Hospital, London, UK.

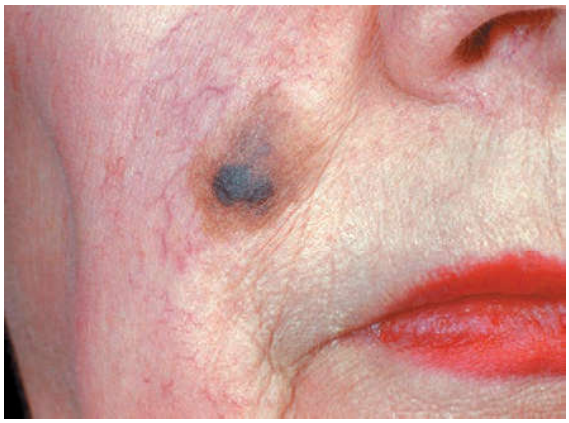

Figure 42.45 Lentigo maligna melanoma (courtesy of St John's Institute for Dermatology, London, UK).

base of tumour) is the most important prognostic indicator in the absence of lymph node metastases. The AJCC staging system then takes lymph node and distant metastases into account (see Table 42.4).

Investigations

Guidelines for staging are controversial. One approach is to aim investigations towards detecting occult disease so as to upstage patients and then treat them accurately and appropriately. Thus, offering sentinel node biopsy to patients with clinical stage II disease is prudent, investigations for stage III disease should be directed to individual clinical presentation.

Local treatment

The treatment for melanoma is surgery. Lentigo maligna (melanoma *in situ*) should be excised completely in most clinical situations because of the risk of it entering the vertical growth phase to become LMM. A complete excision requires no further treatment.

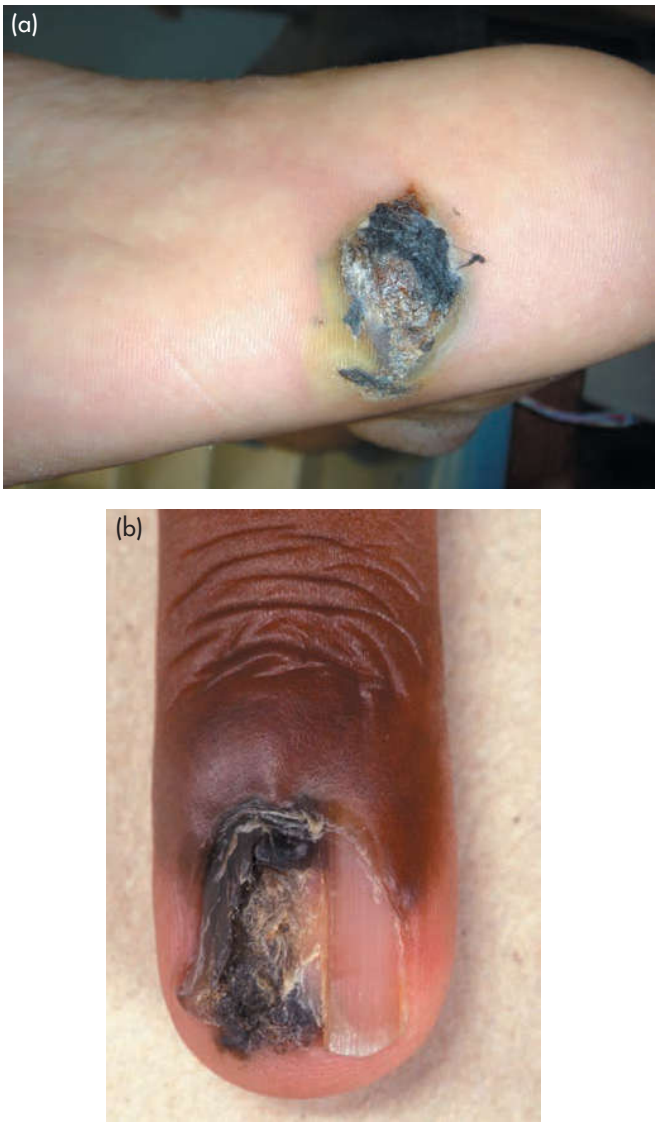

Figure 42.46 (a) Acral lentiginous melanoma on the sole of the foot (courtesy of Mr AR Greenbaum). (b) Subungual melanoma – probably a superficial spreading melanoma. Note the swelling proximal to the nail fold. (c) Benign racial melanonychia. ((b) and (c) Courtesy of St John's Institute for Dermatology, London, UK.)

For melanoma <1 mm deep, wide local excision with a 1 cm margin is sufficient. For deeper lesions, a 2 cm margin is recommended as there is no evidence that wider margins make a difference.

Regional lymph nodes

The likelihood of metastatic spread to regional lymph nodes is proportional to the Breslow thickness of the melanoma. Management of regional lymph nodes has been a contentious topic for well over a century. Some advocate simultaneous elective lymph node clearance at the time of wide excision of the primary melanoma. Ideally, one would like to be able to select for treatment those patients with the highest risk of metastatic spread.

Sentinel node biopsy (SNB) is an investigation based on the fact that lymphatic metastasis proceeds in an orderly fashion and can be predicted by mapping the lymphatic drainage from a primary tumour to the first or 'sentinel' node in the regional lymphatic basin. There is negligible benefit in performing SNB in patients whose primary melanoma is thinner than 1 mm. When SNB is performed according to consensus standards, it is predictive of the regional nodal status in 99 per cent of cases. Seventy-five per cent of patients with metastases in the sentinel nodes will have no other involved regional nodes, so while the current standard approach is to proceed to completion lymphadenectomy, this will overtreat a significant number of sentinel node-positive patients. This might appear a better option than potentially undertreating 20–30 per cent of patients with positive sentinel nodes. Evidence of objective survival benefit from SNB is currently unavailable, but several large prospective clinical trials are in progress to investigate this and meanwhile, SNB remains part of the AJCC staging system.

Current treatment for biopsy positive nodal disease is block dissection of regional lymph nodes to remove all the lymph nodes in that regional basin.

Adjuvant therapy

None is of proven benefit, with clinical trials currently looking at vaccine and interferon treatments. Recent promising research has focused on mutations in the *BRAF* gene, which is found in about 60 per cent of malignant melanoma. Drugs which block the products of the faulty gene are currently showing very promising results in clinical trials with evidence that it shrinks metastatic deposits in patients with stage 4 disease.

Prognosis

The Breslow thickness of the primary tumour offers the best correlation with survival in stage I disease. The higher the mitotic index, the poorer is the prognosis of the primary tumour. This has greater significance than the presence or absence of ulceration.

The presence of lymph node metastases is the single most important prognostic index in melanoma; outweighing both tumour and host factors. The number of affected nodes and the presence of extranodal extension are also significant outcome predictors. Once regional nodes are clinically involved, 70–85 per cent of patients will have occult distant metastases.

Merkel cell (dermal mechanoreceptor) tumour

This is an aggressive malignant tumour of Merkel cells (Figure 42.47). It usually affects the elderly and is four times more common in women than men. Treatment is with wide local excision aiming for a 25–30-mm margin, followed by radiotherapy.

VASCULAR LESIONS

Congenital: haemangiomata and vascular malformations

These can be subclassified biologically into vascular tumours or vascular malformations based on their endothelial characteristics; or radiologically into haemangiomata, vascular and lymphatic malformations based on their vascular dynamics.

Haemangiomata

These are benign endothelial tumours that affect three girls for every boy. Thirty per cent have a herald patch at birth, which then grows rapidly in the first year of life, then slowly involutes over several years with 70 per cent having resolved by seven years of age. Large haemangiomata can trap platelets leading to thrombocytopenia (Kasabach–Merritt syndrome).

Vascular malformations

Vascular malformations affect boys and girls equally and are associated with numerous syndromes. They are invariably present at birth but may be missed if deep to the skin. Vascular malformations subsequently grow in proportion to the child's growth (other than in response to sepsis or hormonal stimulation). Stasis can lead to a localised, consumptive coagulopathy in large venous malformations. Low-flow malformations may cause skeletal hypoplasia, while high flow malformations can cause hypertrophy.

Common vascular birthmarks

Salmon patch

This is a haemangioma that presents as a pinkish macule usually at the nape of neck (Figure 42.48). It is caused by an area of persistent fetal dermal circulation, which usually disappears at one year.

Capillary haemangioma (strawberry naevus)

This is the most common birth mark occurring most commonly on the head and neck (Figure 42.49). Ninety per cent appear at birth, and as a consequence of intravascular thrombosis, fibrosis and mast cell infiltration, 10 per cent resolve each subsequent year, with 70 per cent resolved by seven years of age.

White skin is affected most commonly and girls are affected three times more than boys.

Capillary vascular malformations ('port-wine' stains)

Capillary vascular malformations ('port-wine' stains (PWS)) are 20 times less common than capillary haemangiomata and result from defective maturation of cutaneous sympathetic innervation during embryogenesis leading to localised intradermal capillary vasodilatation (Figure 42.50). They appear at birth as flat, smooth, intensely purple stained areas, most frequently on

Katharine K Merritt, b. 1886, American paediatrician, Department of Paediatrics, the Babies Hospital, Columbia University College of Physicians and Surgeons, New York, NY, USA. Kasabach and Merritt described the condition as a joint paper in 1940.

Haig H Kasabach, 1898–1943, radiologist, the Presbyterian Hospital, New York, NY, USA.

Table 42.4 American Joint Committee on Cancer 2001 staging.

Stage	Primary tumour	Lymph node	Metastases
0	*In situ*	No nodal involvement	No distant metastases
IA	<1 mm, no ulceration		
IB	<1 mm, with ulceration		
	>1 but <2 mm, no ulceration		
IIA	>1 but <2 mm, with ulceration		
	>2 but <4 mm, no ulceration		
IIB	>2 but <4 mm, with ulceration		
	>4 mm, no ulceration		
IIC	>4 mm, with ulceration		
IIIA	Any Breslow, no ulceration	Micrometastasis	
IIIB	Any Breslow, with ulceration	Micrometastasis	
	Any Breslow, no ulceration	≤3 palpable nodes	
	Any Breslow, ± ulceration	In transit metastases/satellites	
IIIC	Any Breslow, with ulceration	≤3 palpable nodes	
	Any Breslow, ± ulceration	≥4 palpable or matted nodes or nodes + in transit metastases	
			M1: skin, subcutaneous or distant
			M2 : lung
			M3 all other sites/or any site +↑[LDH]

LDH, concentration of lactate dehydrogenase.

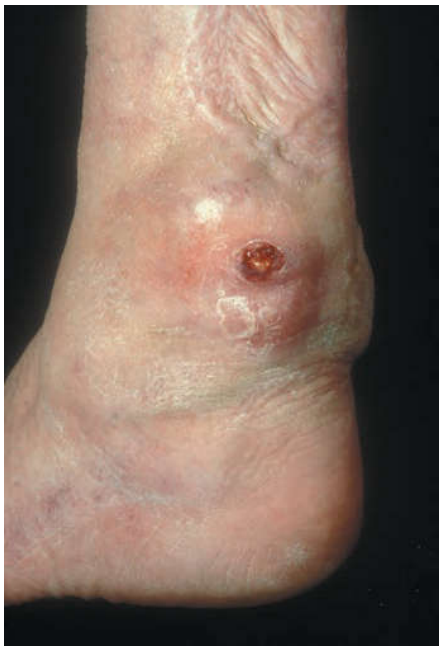

Figure 42.47 Merkel cell tumour (courtesy of St John's Institute for Dermatology, London, UK).

the head and neck, often within the maxillary and mandibular dermatomes of the trigeminal nerve.

Treatment with intense pulsed light and pulse dye laser are successful. PWS may be associated with various syndromes listed below:

- Sturge–Weber syndrome: PWS affecting trigeminal dermatomes; associated with epilepsy, glaucoma secondary to ipsilateral, leptomeningeal angiomatosis, cortical atrophy and visual field defects.
- Klippel–Trenaunay syndrome: PWS on a limb with associated bone and soft tissue hypertrophy and lateral varicose veins when the lower limb is involved. It is called Parkes Weber syndrome if there is an associated arteriovenous malformation.
- Proteus syndrome: PWS and regional gigantism in association with lymphatic (lymphaticovenous) malformation. Hypertrophy is always asymmetrical.

Acquired

Campbell de Morgan spots

These are arteriovenous fistulas at the dermal capillary level in sun exposed skin of older patients (Figure 42.51).

Spider naevi

These are angiomata that appear (and may disappear) spontaneously at puberty or in two-thirds of pregnant women, usually

William Allen Sturge, 1850–1919, physician, The Royal Free Hospital, London, UK.
Frederick Parkes Weber, 1863–1962, physician, The German Hospital, Dalston, London, UK.
Maurice Klippel, 1858–1942, neurologist, La Salpêtrière, Paris, France.
Paul Trenaunay, b. 1875, a French neurologist, Klippel and Trenaunay described the condition as a joint paper in 1900.
Proteus was a minor sea god of Greek mythology, who had the power of prophesy and was able to assume different shapes in order to avoid answering questions.
Campbell Greig de Morgan, 1811–1876, surgeon, the Middlesex Hospital, London UK.

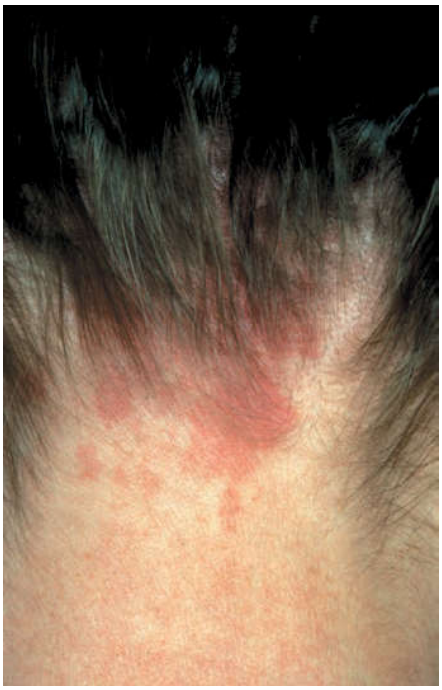

Figure 42.48 Salmon patch (courtesy of St John's Institute for Dermatology, London, UK).

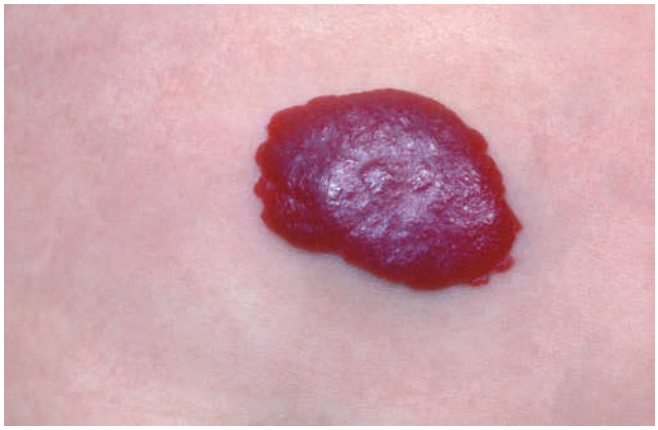

Figure 42.49 Capillary haemangioma (courtesy of St John's Institute for Dermatology, London, UK).

disappearing in the puerperium (Figure 42.52). Spider naevi are also associated with chronic liver disease. They can be treated with intense pulsed light or pulse dye laser.

Pyogenic granuloma

These share many histological characteristics of haemangiomas and are probably a subtype thereof (Figure 42.53).

Most are small (0.5–1.5 cm), raised, pedunculated, soft red nodular lesions showing superficial ulceration and a tendency to bleed after trivial trauma. They should be excised with a minimal margin.

Glomus tumour

These arise from subcutaneous arteriovenous shunts (Sucquet–Hoyer canals) especially in the corium of the nail bed. Typically,

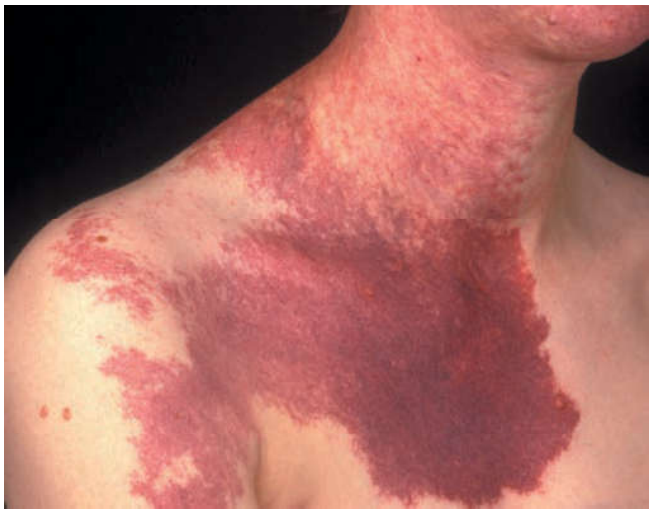

Figure 42.50 'Port-wine' stain (courtesy of St John's Institute for Dermatology, London, UK).

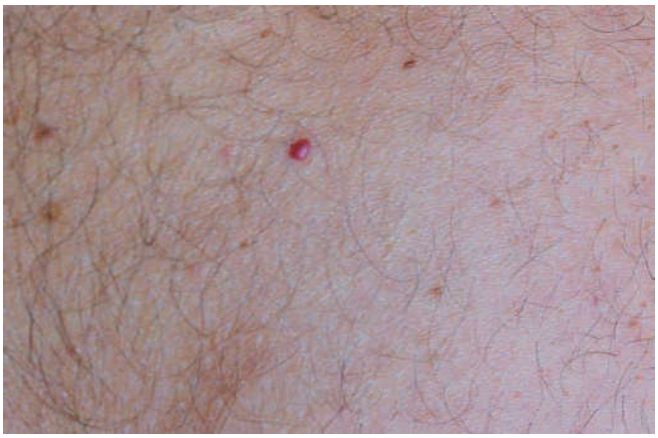

Figure 42.51 Campbell de Morgan spot (courtesy of Mr AR Greenbaum).

it is a small, purple nodule measuring a few millimetres in size which is disproportionately painful in response to insignificant stimuli, including cold exposure (Figure 42.54). Subungual varieties may be invisible causing paroxysmal digital pain.

Angiosarcoma ('malignant angioendothelioma')

Angiosarcoma is a rare, highly malignant tumour arising from the endothelial cells (Figure 42.55). The lymphangiosarcoma variant arises from lymphatic endothelium and can develop in lymphoedematous tissue, particularly an extremity. Proliferation is rapid with early systemic spread.

Kaposi's sarcoma

This is a malignant, proliferative tumour of vascular endothelial cells, which was first described in elderly Jewish men, but is now most commonly associated with immune compromise after transplantation or HIV infection (Figure 42.56). There appears to be a causal link with infection by human herpes virus 8. Kaposi's sarcoma usually starts as a red brown, indurated, plaque-like skin lesion that becomes nodular and then ulcerates. Treatment is with radiotherapy.

JP Sucquet, 1840–1870, an anatomist of Paris, France.
Heinrich Hoyer, 1834–1907, Professor of Histology, Embryology and Anatomy, the Central Medical School, The Polish University, Warsaw, Poland.

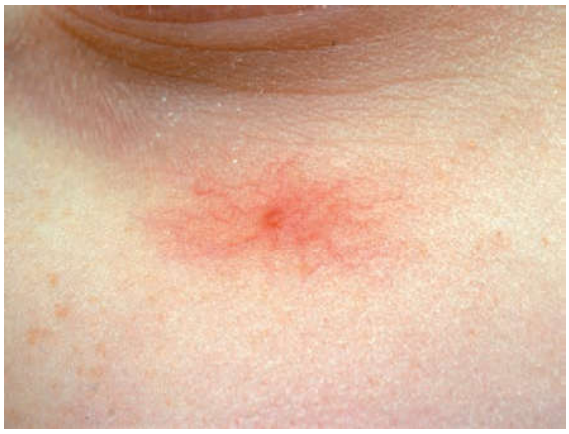

Figure 42.52 Spider naevus (courtesy of St John's Institute for Dermatology, London, UK).

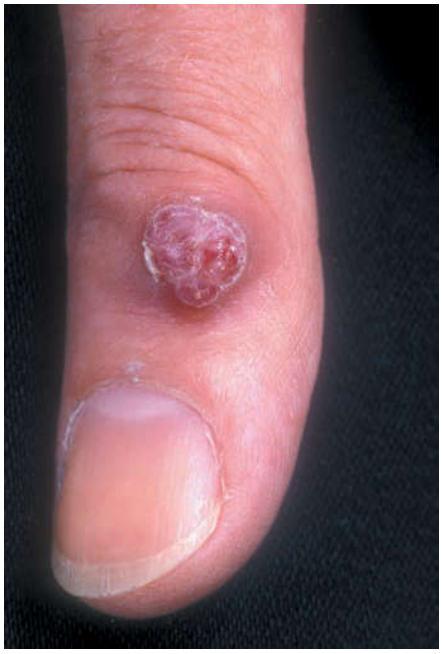

Figure 42.53 Pyogenic granuloma (courtesy of St John's Institute for Dermatology, London, UK).

WOUNDS: CONGENITAL

Cutis aplasia congenita

A rare condition characterised by the congenital absence of epidermis, dermis and, in some cases, subcutaneous tissues, with underlying bony defects in 20 per cent. Lesions may occur on any body surface, but localised scalp agenesis is most frequent.

Treatment depends on the severity of the presentation, but usually involves plastic surgery.

Parry–Romberg disease

An uncommon and poorly understood progressive, hemi-facial atrophy of skin, soft tissue and bone. Its incidence is unknown and its inheritance uncertain, but it affects women more commonly than men.

It commonly starts in a patient's late 20s, but can present in

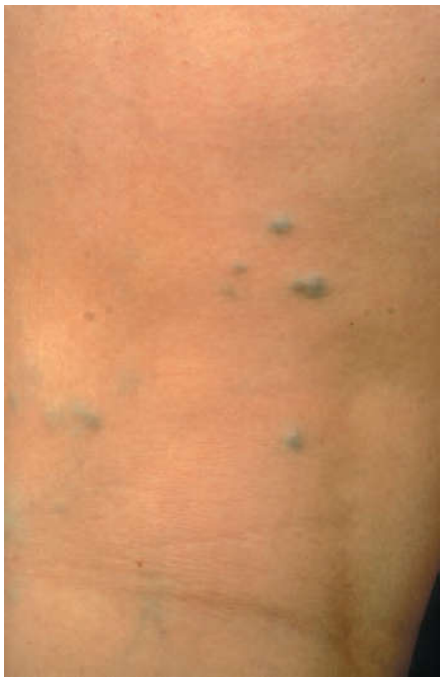

Figure 42.54 Glomus tumour (courtesy of St John's Institute for Dermatology, London, UK).

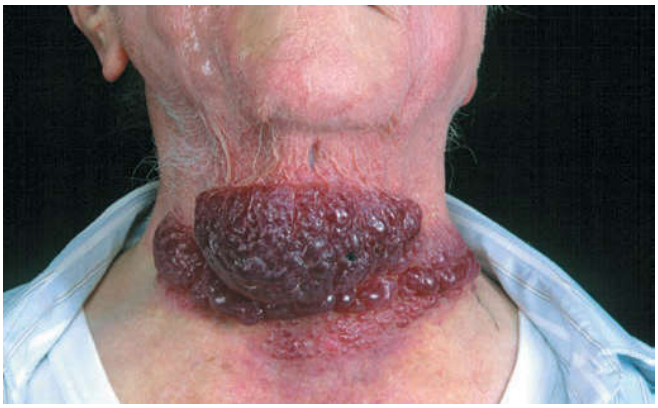

Figure 42.55 Angiosarcoma (courtesy of St John's Institute for Dermatology, London, UK).

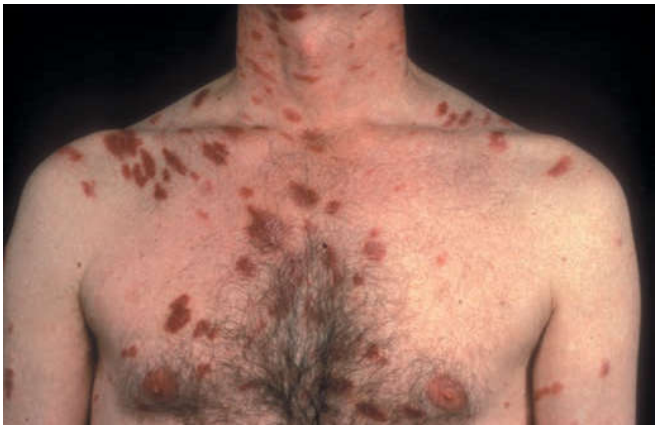

Figure 42.56 Kaposi's sarcoma (courtesy of St John's Institute for Dermatology, London, UK).

Caleb Hillier Parry, 1755–1822, physician, The General Hospital, Bath, UK.
Moritiz Heinrich Romberg, 1795–1873, a German neurologist who was Director of the University Hospital, Berlin, Germany.

childhood, when the resulting deformity is worse because it is magnified by differential growth elsewhere. The most common presentation is confined to lipodystrophy, but mixed atrophy of skin, fat, muscle, cartilage and bone combined result in the classic 'Coup de Sabre' deformity.

The condition is self-limiting, usually by 5–10 years after onset. Once the condition is stable, plastic surgical techniques can be employed alone or in combination to reconstruct an aesthetic contour.

Spina bifida

Failure of closure of the caudal neuropore during the fourth week *in utero* results in incomplete development of some or all of the structural elements posterior to the spinal cord. This can occur anywhere, but is most common in lumbar vertebrae and presents as gross variants: spina bifida occulta in which there is a bony defect without neural protrusion and spina bifida cystica, in which there is herniation of the meninges (meningocoele); spinal cord (myelocoele) or, most commonly, both (meningomyelocoele) and therefore, asymptomatic. Management ideally involves a multidisciplinary approach and is directed towards protecting the spinal cord, preventing cerebrospinal fluid contamination and secondary hydrocephalus and meningitis.

WOUNDS: ACQUIRED

Pressure sores

These begin with tissue necrosis at a pressure point (next to a bony prominence) and develop into a cone shape volume of necrotic loss (with the cone's tip superficial). As many as 10 per cent of acute hospital inpatients will suffer some degree of pressure sores. The majority affect the elderly and patients with spinal injury or decreased sensibility; 80 per cent of paraplegics will get a pressure sore and 8 per cent die as a result.

The pathogenesis of pressure sores revolves around unrelieved pressure: an increase in local tissue pressure above that of perfusion pressure produces ischaemic necrosis that is directly proportional to the duration and degree of pressure and inversely proportional to the area over which it is applied. Muscle and fat are more susceptible to pressure than skin.

In a patient who has no predisposing factors (who developed a sore while unable to move, but normally can move), management is aimed at debridement and repair of the defect on the assumption that recurrence will not occur once normal function and sensibility returns. In the paraplegic, recurrence is likely, so management should involve a multidisciplinary approach with surgery used sparingly once all other predisposing factors have been addressed. Primary treatment involves relieving pressure (special mattress; nursing care; relief of muscle spasm and contractures); optimising nutrition; correcting anaemia; preventing infection and dressings. Surgery involves thorough debridement to promote healing and plastic surgery to reconstruct the defect.

FURTHER READING

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Coup de Sabre is French for a 'sabre cut', a vertical cut across the shoulder from the front to the back.