Otolaryngology

Rhinology

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Sinonasal Tumours:

Pathology:

About 10% of head and neck cancer is sinonasal. It is about double this in Arabs, the Japanese and Africans. The male: female ratio in series varies between 1:2 and 1:5. Half of sinonasal cancer arises from the upper jaw, a quarter from the ethmoids and a quarter from the nasal cavity. Histologically 50% are squamous cell carcinoma, 15% anaplastic, 10% lymphomas and about 4% adenocarcinomas.Rarely olifactory neuroma is also reported. The association between the ethmoidal adenocarcinoma and hardwood workers is well documented. Workers handling chromate salts and those involved in nickel refining are at an increased risk of nasal malignancy. Chronic nasal pathology, including sepsis and Wegener's granulomatosis, and smoking cigarettes have recently been implicated as providing an increased risk of squamous cell carcinoma. Human Papiloma virus genome has been identified and therefore implicated as an aetiological agent in non-dysplastic and dysplastic inverted papilloma and seuamous cell carcinoma.

Classification:

Both benign and malignant groups can be classified into epithelial, non epithelial, odontogenic, and fibro-osseous tumours.

1. Benign.

Epithelial (papilloma, adenoma and inverted papilloma).

Non-epithelial (fibroma, haemangioma, nasal glioma, Schwannoma, chon-droma, haemangiopericytoma, chordoma, meningioma and osteoma).

2. Malignant.

Epithelial (squamous cell carcinoma, adenocarcinoma, anaplastic carcinoma, transitional cell carcinoma, malignant melanoma, salivary gland malignancy in particular adenoid cystic carcinoma and olfactory neuroblastoma.

Non-epithelial (fibrosarcoma, angiosarcoma, chondrosarcoma, rhabdomyo-sarcoma, and osteogenic sarcomas).

Important benign sinonasal tumours

Inverted papilloma:

originally described by Ringertz, is the most important of this group forming about 5% of all nasal tumours. Macroscopically there is usually a papilliferous exophytic mass. Microscopically there are deep invaginations of epithelium into the stroma, with microcyst formation. The epithelium retains its basement membrane. Probably less than 2% undergo malignant change although there may be a synchronous sinonasal squamous cell carcinoma in up to a further 10%. This emphasizes the need for careful endoscopic follow up.

Osteomas:

most commonly arise from the frontal region where they may expand medially to block the frontal recess predisposing to a secondary mucocele or frontal sinusitis, inferiorly to displace the orbit, superiorly where they may erode the cribriform plate or posteriorly to erode the posterior wall of the frontal sinus and impinge on frontal lobe dura. They consist of hard cortical bone and require excision if symptomatic or enlarging. **Haemangiopericytomas:**

may arise anywhere in the sinonasal region and have a spectrum of aggression, with a propensity to recur many years after apparent cure.

<u>Malignant sinonasal tumours:</u>

Squamous cell carcinoma and adenocarcinoma both usually present at an advanced stage because their presenting symptoms of epistaxis, nasal obstruction and headaches occur only with a significant tumour mass. Further delays may arise because these symptoms are not usually associated with carcinoma by primary care physicians. Cheek swelling occurs only when the tumour has breached the anterior antral wall to impinge on periosteum. It is usually impossible to define a site of origin of the carcinoma due to its diffuse extent. This may include the cheek, orbit, nasal cavity and anterior cranial fossa. Five per cent of subjects will have a metastatic neck lymph node, usually upper deep cervical, on presentation. This indicates a poor prognosis but not necessarily incurability. A quarter of patients will die from distant metastases, most commonly the bronchus. Adenoid cystic carcinoma is particularly difficult to eradicate because of its ability to spread via the branches of the trigeminal and olfactory nerves along perineurium. Malignant melanomas comprise 1% of sinonasal carcinomas and usually arise from the septum or lateral nasal wall, where the prognosis is reasonable.

Frontoethmoidal and antral malignant melanoma have a much worse prognosis. There is no relationship between Clark's classification (penetration of specific skin layers) and prognosis in this region although the latter is associated with Breslow's classification (thickness of the lesion in mm). Olfactory neuroblastomas usually arises from neural crest stem cells, the precursors of olfactory cells, and microscopically resembles other small cell malignancies such as high grade lymphoma and anaplastic carcinoma. All ages are affected and urine vanillylmandelic acid (VMA), is not usually detectable. It always involves the cribriform plate so if resection is contemplated this must be via a craniofacial approach.

Staging

The UICC staging is as follows: Tx Primary tumour cannot be assessed. T0 No evidence of primary disease. Tis Carcinoma in situ.

Maxillary sinus:

T1 Tumour limited to the antral mucosa with no erosion or destruction of bone.

T2 Tumour causing bone erosion or destruction, except for the posterior wall, including extension into the hard palate and/or middle nasal meatus. T3 Tumour invades any of the following: bone of posterior wall of maxillary sinus, subcutaneous tissues, skin of cheek, floor of medial wall of orbit.

T4 Tumour invades orbital contents beyond the floor or medial wall including apex and/or any of the following: cribriform plate, base of skull, nasopharynx, sphenoid sinus, frontal sinus.

Ethmoid sinus:

T1 Tumour confined to ethmoid with or without bone erosion.

T2 Tumour extends into nasal cavity.

T3 Tumour extends to anterior orbit and/or maxillary sinus.

T4 Tumour with intracranial extension, orbital extension including apex, involving sphenoid and/or frontal sinus and/or skin of nose.

Clinical features:

1. Nasal cavity tumours. Epistaxis, nasal obstruction and a mass visible on nasendoscopy.

2. Frontal sinus. Features are similar to frontal osteomas except the history is shorter and more rapidly progressive.

3. Ethmoidal. Epistaxis, nasal obstruction and if the lamina papyracea is breached, proptosis, epiphora and diplopia. Nasendoscopy may reveal tumour extruding from the middle meatus.

4. Antral. Epistaxis, nasal obstruction, cheek swelling, headache if blocking the osteomeatal complex and atypical facial pain (suggesting involvement of the pterygo-palatine fossa or the infra orbital nerve). Oroantral fistula, ill-fitting dentures, trismus and ethmoidal symptoms occur with advanced disease.

Investigations:

A high definition CT scan on both bone and soft tissue windows is the ideal to show soft tissue and bone involvement. A T2 weighted or STIR sequence MRI scan may distinguish tumour from inflammation, retained secretions and fat.

Treatment:

Inverted papilloma:

The accurate identification of disease extent with CT and MRI imaging has allowed the advent of endoscopic resection for inverted papilloma. Recent studies have demonstrated recurrence rates no higher than with external approach surgery. There is significantly less morbidity by the endoscopic approach. The technique allows direct visualization of tumour and its extent can be accurately identified during surgery. It prevents an external scar, there is less blood loss and the hospital stay is shorter. Tumour limited to the anterior ethmoids or isolated middle turbinate or middle meatal lesion are ideal for endoscopic resection. Recent work has also shown its application for tumours involving the posterior ethmoids and anterior wall of sphenoid. In essence provided the endoscope can visualize the distal extent of tumour during surgery then endoscopic resection is a reasonable and perhaps preferred alternative to external approach surgery. Post-operatively endoscopic inspection of the surgical cavity allows accurate monitoring of patient progress and early identification of recurrence.

Carcinoma:

There are three main surgical options for carcinoma:

(a) Lateral rhinotomy for tumour limited to the lateral nasal wall, nasal cavity and ethmoid. An upper limb extension will allow tumour limited to the frontal ethmoidal region to be accessed.

(b) Total maxillectomy. For antral carcinoma.

(c) Craniofacial resection is indicated when the cribriform plate is involved or breached.

In general an orbital exenteration is indicated only if tumour breaches periostium to involve orbital fat. Adjuvant radiotherapy may be indicated depending on tissue margins. Prognosis by stage is difficult for reasons already outlined. Overall a five-year survival of 40-50% would be reasonable.

Follow-up and aftercare:

Ideally an orthodontist should take an impression of the maxillectomy cavity at operation in order to make a temporary obturator. Further review allows a fine tuning of the prosthesis to provide a light, comfortable, well fitting and easily removable obturator. Those who have a lateral rhinotomy or craniofacial cavity often have excessive crusting in the early post-operative phase. Glycerine and glucose nose drops and regular douching with saline will minimize it Follow-up which involves nose endoscopy to inspect the surgical cavity created or direct inspection of a maxillectomy cavity after obturator removal, laryngopharyngeal examination to exclude a second primary and neck examination to look for metastases should be monthly for the first post-operative year, bimonthly for the second, quarterly for the third and six-monthly until five years post surgery. Some surgeons thereafter review annually for a further five years.