

Seamus S. Napier and Ramzan M. Ullah

13.1 Anatomy

The external nose contains the right and left *nostrils* (or nares), each communicating with the nasal cavities via a slight dilation just inside the nostril called the *nasal vestibule*. Bone from the frontal, maxillary and nasal bones supports the upper one third of the external nose, while cartilage supports the lower two thirds.

Each nasal cavity extends posteriorly from just behind the nasal vestibule, through the opening called the *anterior choana*, to communicate with the nasopharynx via the *posterior choana*. They are separated by the *nasal septum*, which is composed of bone posteriorly and cartilage anteriorly. Each nasal cavity has a roof, a floor, a medial (or septal) wall, and a lateral wall (Fig. 13.1). The *roof* of the nose is closely related to the frontal sinuses, the anterior cranial fossa, the ethmoidal sinuses, and the sphenoidal sinus. The *floor* of the nose is closely related to the anterior maxillary teeth and the vault of the palate,

while the *medial wall* represents the nasal septum. The *lateral wall* of the nose is complex and bears three (occasionally four) horizontal projections called *turbinates* or *conchae*, the superior turbinate being the smallest and the inferior turbinate the largest. The passageway of the nasal cavity below and lateral to each of the turbinates is called the *superior, middle, and inferior meatus*, respectively; above and behind the superior turbinate lies the sphenoidal recess. The paranasal sinuses open onto the lateral wall of the nasal cavity, as does the nasolacrimal duct, so that disease affecting this region of the nose can obstruct the drainage of secretions and present as sinusitis.

Each nasal cavity is divided into functional areas, reflected in the nature of the epithelial lining. The nasal vestibule is lined by skin and contains many short hairs that help to filter particles from the inspired air. The olfactory area, concerned with the sense of smell, is restricted to the upper part of the nasal cavity and is centred on the cribriform plate of the ethmoid bone, the adjacent part of the nasal septum and the superior turbinate. The rest of the nasal cavity is lined by respiratory mucosa, the function of which is to warm and humidify the air and to trap particulate material. The complex architecture of the lateral wall of the nasal cavities facilitates this process by increasing the surface area and the turbulence of the airflow.

The *paranasal sinuses* are extensions of the nasal cavities and represent air-filled spaces in

S.S. Napier (✉)
Histopathology Laboratory, Institute of Pathology,
Royal Victoria Hospital, Belfast Health and Social
Care Trust, Belfast, UK
e-mail: seamus.napier@belfasttrust.hscni.net

R.M. Ullah
Directorate of ENT Surgery, Royal Victoria Hospital,
Belfast Health and Social Care Trust, Belfast, UK
e-mail: ramzan.ullah@belfasttrust.hscni.net

Fig. 13.1 Right lateral wall of nasal cavity (Reproduced, with permission, from Allen and Cameron (2013))

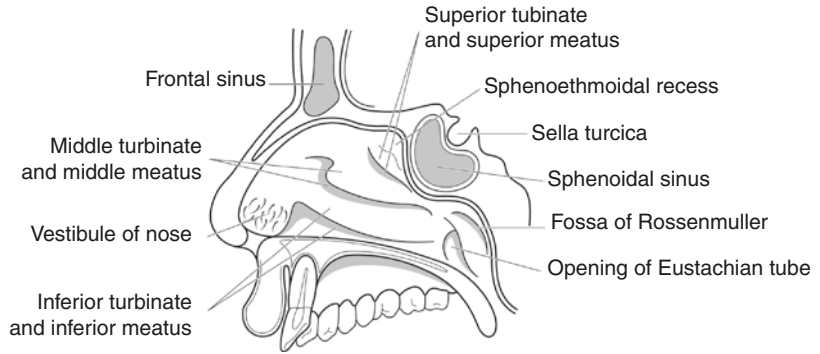
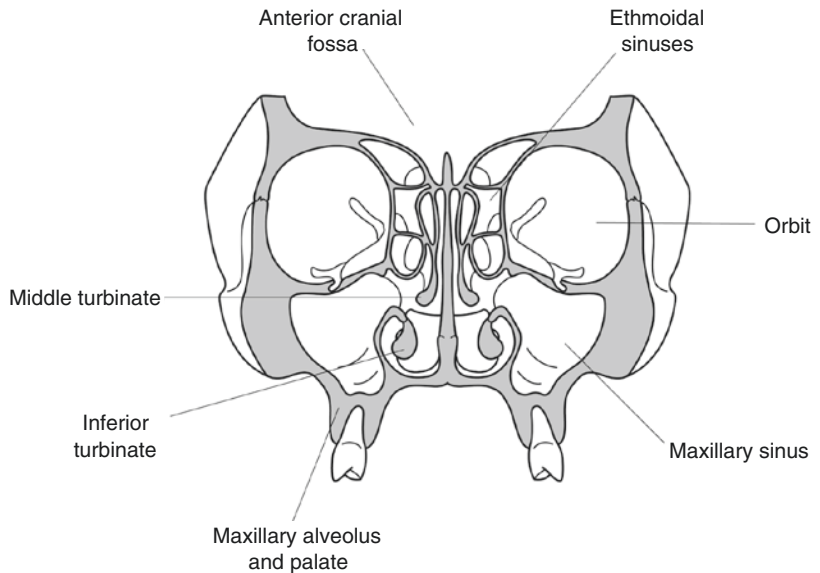


Fig. 13.2 Ethmoidal sinuses and maxillary sinuses. Coronal view of nasal cavities at level of first molar tooth showing maxillary and ethmoidal sinuses (Reproduced, with permission, from Allen and Cameron (2013))



the skull bones lined by respiratory mucosa (Fig. 13.2). They are usually absent or poorly developed at birth but enlarge most during the eruption of the permanent teeth and after puberty. They are located in the frontal, ethmoidal, sphenoidal, and maxillary bones as paired structures about the midline but tend to be considered from a pathophysiological perspective into anterior and posterior groups. The *anterior group* comprises the frontal sinus, the anterior and middle ethmoidal sinuses, and the maxillary sinus, all opening into the middle meatus, while the posterior ethmoidal sinuses and the sphenoidal sinus represent the *posterior group* and drain into the superior meatus and the sphenoethmoidal recess, respectively. The

nasolacrimal duct opens into the inferior meatus anteriorly.

The *frontal sinuses* lie between the outer and inner tables of the frontal bone and are closely related to the anterior cranial fossa. Disease in the frontal sinus can be associated with intracranial complications. The *ethmoidal sinuses* number between 3 and 18, and consist of a labyrinth of thin-walled bony cavities between the upper part of the nasal cavity and the orbits. The pathway for drainage of the frontal sinus passes through the anterior ethmoidal sinus group and may be impeded by disease in this area. The *sphenoidal sinuses* lie within the body of the sphenoid bone posterior to the upper part of the nasal cavity. Adjacent structures such as the optic

chiasma, pituitary gland, internal carotid artery, and cavernous sinus may be affected by disease of the sphenoidal sinuses. The *maxillary sinus* is the largest of the paranasal sinuses and is closely related to the posterior maxillary teeth, the floor of the orbit, the inferior portion of the lateral wall of the nose, and the pterygoid plates of the sphenoid bone.

Lymphovascular drainage:

Lymphatics from the external nose and anterior nasal cavity, together with those from the skin of the mid-portion of the face, drain to Level I lymph nodes in the submandibular region. The rest of the nasal cavity and the paranasal sinuses drain to Level II lymph nodes in the upper part of the deep cervical chain (Fig. 20.1), sometimes via the retropharyngeal nodes.

13.2 Clinical Presentation

Disease affecting the nose presents with unilateral or bilateral nasal obstruction, rhinorrhea (watering of the nose), epistaxis (bleeding), facial pain, facial swelling, epiphora (watering of the eye), proptosis (bulging outward of the eye) or anosmia (loss of the sense of smell). Deafness or otitis media may be due to obstruction of the opening of the Eustachian tube in the nasopharynx by extension of a nasal tumour.

13.3 Clinical Investigations

- Direct visualization of the nasal cavities is performed using a speculum for the anterior aspect or a postnasal mirror for the posterior portion. Nasal endoscopy is the preferred method of sampling tissue from the nose and nasal sinuses.
- Plain radiographs of the nose and sinuses may demonstrate bone destruction, soft tissue mass, or fluid levels, although these are more accurately determined by CT and MRI scanning.
- Markedly elevated erythrocyte sedimentation rates (ESR) and titers of “cytoplasmic” anti-neutrophil cytoplasmic antibodies (cANCA) are helpful adjuncts to diagnosis in the so-

called Midline Destructive Diseases, a collection of diseases characterized by progressive destruction of the nose and sinuses. These tests help distinguish principally between granulomatosis with polyangiitis (previously known as Wegener’s granulomatosis) and T-cell/natural killer cell lymphoma.

13.4 Pathological Conditions

13.4.1 Non-neoplastic Conditions

Sinusitis: Acute infections are usually bacterial and often follow the common cold. Empyema or mucocele may result if the draining of the secretions is obstructed. Chronic sinusitis follows acute sinusitis and may be associated with obstruction (e.g., by polyp or tumour) or immune compromise. Maxillary sinusitis may occur alone or may be associated with involvement of frontal and/or ethmoidal sinuses. Most cases respond to antibiotics and topical medications to improve drainage. Functional endoscopic sinus surgery (FESS) is the commonest surgical management of recurrent sinusitis; opening of the osteo-meatal complex under the middle turbinate or partial removal of pneumatized middle turbinates (concha bullosa) or nasal polyps will improve physiological drainage and allow biopsy sampling.

Pain of dental origin can mimic maxillary sinusitis and vice versa. Extraction of upper premolar or molar teeth may damage the floor of the maxillary sinus and result in an oroantral fistula through the socket.

Inflammatory polyps: A frequent complication of long-standing rhinitis, often but not exclusively allergic in origin. Often multiple and bilateral, they are a cause of sinusitis and nasal obstruction. Histologically, there is abundant myxoid or oedematous stroma covered by respiratory epithelium; ulceration and/or squamous metaplasia are common in larger polyps, where they contact the nasal walls. The antrochoanal polyp is an uncommon large single inflammatory polyp that arises in the maxillary sinus and extends into the nasal cavity, presenting at the

posterior choana. Nasal polyps in children are often associated with cystic fibrosis.

Granulomatosis with polyangiitis: Previously known as Wegener's granulomatosis, an uncommon systemic disorder characterized by necrotizing granulomatous inflammation and vasculitis that usually presents in the upper respiratory tract, lungs, and/or kidneys. Symptoms can be nonspecific (malaise, pyrexia) or related to the anatomical sites involved; in the nose it may manifest as sinusitis, rhinorrhea, epistaxis, or nasal obstruction. Rarely are the classical features present in nasal biopsies; diagnosis requires a high index of suspicion and careful clinicopathological correlation. ESR and cANCA titers are useful at confirming the diagnosis, although a negative cANCA does not exclude. A distinctive form of small multinucleate giant cell with clumped smudged nuclei, foci of granular collagen necrosis, and neutrophil microabscesses are characteristic if under-recognized features.

Other non-neoplastic conditions that may affect the nose and sinuses include fungal infections (chronic noninvasive colonization by *Aspergillus*, acute fulminant or angioinvasive aspergillosis, allergic fungal sinusitis), pyogenic granuloma, haemangioma and other vascular malformations, lymphoid hyperplasia, glial heterotopia, and hairy polyp.

13.4.2 Neoplastic Conditions

Benign tumours: Sinonasal papillomas are uncommon but are the most frequent benign neoplasms, subdivided into fungiform, inverted, and cylindrical cell types. Occur twice as often in males as in females and affect adults aged between 30 years and 60 years. They are usually unilateral lesions but may be multiple or multifocal. *Inverted papillomas* are the commonest form, found on the lateral nasal wall and sinuses. They have an endophytic growth pattern and are composed of thick non-keratinizing, "transitional" epithelium within oedematous stroma. *Fungiform papillomas* are exophytic lesions composed of transitional epithelium supported by fibrovascular stroma, found exclusively on the nasal sep-

tum. *Cylindrical cell papillomas* are rare. They are similar in distribution and appearance to inverted papillomas but are composed of tall columnar (cylindrical) oncocyctic cells.

Other benign neoplasms include pleomorphic adenoma, solitary fibrous tumour, glomangiopericytoma (previously haemangiopericytoma), nasopharyngeal (juvenile) angiofibroma, sinus osteoma, meningioma, teratoma, and paraganglioma.

Sinonasal cancer: The maxillary sinus is the commonest site for sinonasal malignancy and is usually either squamous cell carcinoma or adenocarcinoma in type. The nasal cavity is the second commonest site and is affected by a broad spectrum of lesions, but tumours of the sphenoidal and frontal sinuses are rare. Risk factors include tobacco use, exposure to hard and soft wood dusts, nickel, and irradiation.

Squamous cell carcinoma: The vast majority of malignant tumours of the mucosal lining of the nasal cavities and sinuses are classified as squamous cell carcinoma. The maxillary or ethmoid sinuses are the commonest sites but the nasal vestibule or septum can be affected. Many tumours have a "transitional cell" pattern, similar to that seen in inverted papillomas but exhibiting pleomorphism and necrosis; a broad "pushing" front can make diagnosis of invasion difficult on small biopsy samples. The term "non-keratinizing squamous cell carcinoma" can be used, but a spectrum of changes including the presence of single cell infiltration and/or abundant keratinization may be seen, sometimes making distinction from the usual type of squamous cell carcinoma impossible.

Salivary gland-type adenocarcinoma: The second commonest type of malignant tumour with adenoid cystic carcinoma as the pattern most often encountered.

Intestinal-type sinonasal adenocarcinoma: Adenocarcinoma exhibiting the differentiation pattern of large or small intestinal mucosa, with but occasionally without cytological atypia. Strongly associated with hardwood dusts (males, ethmoidal sinuses) but may occur sporadically (females, maxillary sinus). Commonest pattern mimics colonic adenocarcinoma—metastasis

needs to be excluded. Mucinous tumours with signet ring cells are rare.

Malignant lymphoma: All types of non-Hodgkin's lymphoma may affect the sinonasal region either as a site of origin or as part of disseminated disease; diffuse large B-cell lymphoma is the commonest. T-cell and natural killer cell lymphomas often demonstrate a striking tendency for vascular involvement, sometimes with bizarre acute ischaemic changes, such as tooth exfoliation and bone necrosis. The tumour cells may be small, large or intermediate in size; the admixture of other inflammatory cells masks the neoplastic component by mimicking an inflammatory condition such as infection or granulomatosis with polyangiitis (previously known as Wegener's granulomatosis).

Others: Low-grade sinonasal adenocarcinoma, olfactory neuroblastoma, malignant melanoma, small cell neuroendocrine carcinoma, sinonasal undifferentiated carcinoma, rhabdomyosarcoma, chondrosarcoma and chordoma are all uncommon.

Prognosis: Outcome depends on the histological type of tumour as well as the extent of spread. Most lesions are advanced at presentation although lymph node metastasis with carcinomas is relatively infrequent. Local recurrence is a common problem in spite of radical surgery and radiotherapy. Melanomas, small cell neuroendocrine carcinomas, and sinonasal undifferentiated carcinomas are particularly aggressive but 5-year survival is the norm with adenoid cystic carcinomas. In intestinal type sinonasal adenocarcinoma, grading based on the degree of differentiation is important, in that low-grade lesions do well while high-grade lesions do badly. Around 20% 5-year survival is customary. The usefulness of grading olfactory neuroblastomas is less clear.

detailed examination and large biopsy samples are best obtained with this technique under general anaesthesia, as it avoids contamination with tumour and compromising later definitive surgical procedures. Benign tumours such as nasal papillomas may be resected using endoscopic laser surgery but tend to be delivered as small fragments.

13.5.2 Resection Specimens

Endoscopic resection is being used increasingly for the management of localized malignancy and can be combined with traditional open surgical approaches in extensive disease. *Excision specimens of nasal septum* are easily delivered intact via a lateral rhinotomy incision. *Medial maxillectomy* is the commonest surgical procedure for low-grade tumours of the lateral aspect of the nasal cavity and/or maxillary, ethmoid, and frontal sinuses. Resection specimens tend to be fragmented because of the fragile nature of the bone; in these cases, precise interpretation of surgical margins requires orientation of the tissue samples by the surgeon. Alternatively, separate biopsy samples of critical or suspicious areas may be taken after clearance of tumour and submitted separately. *Palatal fenestration* is recommended for low maxillary sinus tumours involving the oral cavity; definite or possible involvement of the posterior wall of the maxillary sinus requires *maxillectomy*. Prosthetic rehabilitation with an obturator constructed around an upper denture provides optimal functional and aesthetic results and allows good visualization of the wound postoperatively facilitating re-biopsy of suspicious areas.

Craniofacial resection describes a surgical approach through both the anterior skull and the mid-face performed for tumours of the frontal or ethmoid sinus that extend into the anterior cranial fossa. Total ethmoidectomy, nasal exenteration, maxillectomy, and orbital exenteration can be performed if necessary.

Involvement of the orbital floor or medial wall is an important nodal point in the management of sinonasal tumours. Breach of the bony wall or involvement of periosteum by tumour may necessitate clearance or exenteration of the orbit.

13.5 Surgical Pathology Specimens: Clinical Aspects

13.5.1 Biopsy Specimens

Rigid or fibre-optic endoscopy is the usual method of sampling lesions in the nose and paranasal sinuses. When malignant disease is suspected,

Concomitant neck dissections are usually not indicated unless there is proven metastatic disease.

13.6 Surgical Pathology Specimens: Laboratory Aspects

13.6.1 Biopsy Specimens

Usually as small samples from open biopsies or core needle specimens, free-floating in formalin. Measure in three dimensions or length of core and submit in total. Specimens containing bone require decalcification and can be recognized by their tendency to sink rapidly in the fixative.

13.6.2 Resection Specimens

13.6.2.1 Septal Excision, Medial Maxillectomy, and Craniofacial Resection Specimens

Most *septal excision* and *medial maxillectomy specimens* are for the less extensive or less locally aggressive neoplastic diseases, such as inverted nasal papilloma, olfactory neuroblastoma, and even malignant melanoma. They are usually received as multiple fragments of mucosa with underlying bone and/or cartilage. In medial maxillectomy specimens, at least the inferior turbinate is included, but, depending on tumour location, all turbinates may be represented.

Most *craniofacial resection specimens* are for extensive or locally aggressive neoplastic diseases of the frontal or ethmoid sinuses, where a curative outcome is expected. As such, they will represent composites of septal excision, medial maxillectomy, maxillectomy, and skull-base excisions. They are usually received intact or as two or three large fragments. They are handled as if they represented an extended medial maxillectomy specimen. Invasion into dura is an ominous finding.

Samples of critical or clinically suspicious margins taken at clearance of tumour should be submitted separately and handled as biopsy specimens.

Initial procedure:

If intact, orientate the specimen and ink its margins.

Otherwise, if a larger specimen is submitted, ink its margins. A line of ink drawn across medium-sized fragments can aid orientation after microscopic examination and assist assessment of margins.

Slice the larger pieces into 0.4 cm thick slices transversely, using a band saw or equivalent.

Measurements:

If intact, dimensions (cm)

If fragmented, number of fragments, total weight (g), and dimensions of largest specimen (cm)

- Tumour
- Size (cm)
- Number of fragments consisting of tumour
- Distance to closest surgical margins (cm)

Description:

- Tumour
Size, shape, and colour
Presence of necrosis
If fragmented, number of fragments containing tumour
- Adjacent mucosa
Colour and consistency
Presence of other lesions
Other
- Lymph nodes, neck dissection
Maxillectomy specimens
See Maxilla, Mandible, and Teeth (Chap. 15).

Blocks for histology:

In cases of neoplastic disease, the histology should represent the tumour, its relationship to the adjacent mucosa, and underlying bone or cartilage. Focal abnormalities of mucosa need to be sampled.

Three blocks of tumour to illustrate the interface with adjacent normal tissues.

Three blocks of adjacent mucosa.

Closest deep surgical margin. Samples of other lesions, e.g., nodules or polyps.

In intact specimens, sample the mucosal margins before sawing the bone and submit separately (reduces contamination of the margins). Cut with a sharp blade firmly down to bone and use a flat blunt instrument to dissect mucosa free from the bone.

In intact specimens, saw the bone into 0.5 cm slices in the transverse plane (vertical plane in vivo).

If fragmented, bread-slice larger specimens and submit as labeled blocks. Microscopic analysis may allow reconstruction and useful assessment of margins.

Histopathology Report:

Final reports of sinonasal specimens should include details on:

- The specimen type and side
- If fragmented, the number of fragments and the size of the largest
- The type, subtype, and grade of tumour present
- Sinonasal papilloma variants
- Squamous cell carcinoma and variants
- Low-grade adenocarcinoma
- Intestinal-type adenocarcinoma
- Lymphoma
- The macroscopic size of tumour
- The presence or absence of invasion of bone
- The distance of tumour from the nearest margin
- The presence or absence of vascular invasion
- The presence or absence of dural invasion (if craniofacial resection)
- Other pathology such as radiation injury

Extent of local tumour spread: TNM 8: for carcinoma

Maxillary sinus

pT1	Mucosa
pT2	Bone erosion/destruction, hard palate, middle nasal meatus
pT3	Posterior bony wall maxillary sinus, subcutaneous tissues, floor/medial wall of orbit, pterygoid fossa, ethmoid sinus

pT4a	Anterior orbit, cheek skin, pterygoid plates, infratemporal fossa, cribriform plate, sphenoid/frontal sinus
pT4b	Orbital apex, dura, brain, middle cranial fossa, cranial nerves other than V2, nasopharynx, clivus

Nasal cavity and ethmoid sinus

pT1	One subsite
pT2	Two subsites or adjacent nasoethmoidal site
pT3	Medial wall/floor orbit, maxillary sinus, palate, cribriform plate
pT4a	Anterior orbit, skin of nose/cheek, anterior cranial fossa (minimal), infratemporal fossa, pterygoid plates, sphenoid/frontal sinus
pT4b	Orbital apex, dura, brain, middle cranial fossa, cranial nerves other than V2, nasopharynx, clivus

Malignant melanoma

pT3	Involvement of mucosa only
pT4a	Deep soft tissue/cartilage/bone or overlying skin
pT4b	Brain/dura/skull base/lower cranial nerves, masticator space, prevertebral space, carotid artery, mediastinal structures

As malignant melanomas of the upper aerodigestive tract are generally aggressive lesions, Stage I and II disease (i.e., pT1 N0 and pT2 N0) have been omitted.

All sites except nasopharynx: regional lymph nodes (cervical). Selective and modified/radical lymphadenectomy will ordinarily include 10 or 15 or more lymph nodes, respectively.

pN0	No regional node metastasis
pN1	Metastasis in an ipsilateral single node ≤3 cm without extranodal extension
pN2	Metastasis in:
	(a) Ipsilateral single node ≤3 cm with extranodal extension, or, >3–6 cm without extranodal extension
	(b) Ipsilateral multiple nodes ≤6 cm without extranodal extension
pN3	(c) Bilateral or contralateral node(s) ≤6 cm without extranodal extension
	(a) Metastasis in a lymph node >6 cm, or (b) Extranodal extension with any of; >3 cm, multiple ipsilateral, contralateral, bilateral

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