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Original research article

Imaging of skull base tumours

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ARTICLE INFO

Article history:

Received 24 June 2015

Received in revised form

12 November 2015

Accepted 23 December 2015

Available online 2 February 2016

Keywords:

Skull base imaging

Head and neck

Skull base tumours

Oncology

ABSTRACT

The skull base is a highly complex and difficult to access anatomical region, which constitutes a relatively common site for neoplasms. Imaging plays a central role in establishing the differential diagnosis, to determine the anatomic tumour spread and for operative planning. All skull base imaging should be performed using thin-section multiplanar imaging, whereby CT and MRI can be considered complimentary. An interdisciplinary team approach is central to improve the outcome of these challenging tumours.

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1. Background

The skull base is a highly challenging anatomical region with regards to imaging diagnosis and intervention. Interposed between the cranial contents, face and neck, it is difficult to access, but constitutes a relatively common site for neoplasms. With densely packed neural structures and vessels traversing multiple tissue layers, there is limited tolerance to manipulation or mass effect. The blood supply to the skull base is dual via the extracranial and intracranial circulation, which has implications for disease propagation and endovascular therapy. Numerous foraminal openings further complicate the anatomy and provide a conduit for tumour spread into the deep spaces of the head and neck. Rather than a uniform entity, the skull base represents a complex, multicompartamental network of tissues. Not surprisingly, there

is no definitive system for the categorisation of skull base tumours. The main purpose of this review is to guide the radiological differential diagnosis. Benign and malignant skull base lesions frequently arise in a typical location, whereas their imaging features may overlap. Importantly, the clinical presentation and deficit usually depend more on the tumour site than histology. For these reasons, the authors have chosen a subdivision of neoplasms according to their anatomical location. The upcoming sections will discuss anterior, central and posterior skull base tumours, with neoplasms of the petrous temporal region included in the latter section.

2. Technical considerations

Owing to its undulating course and varying thickness, all skull base imaging should be performed using thin-sections

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<http://dx.doi.org/10.1016/j.rpor.2015.12.008>

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in at least two planes, whereby most authors consider CT and MRI complimentary.¹ CT is extremely useful in delineating bone detail and identifying aggressive features of malignancy, particularly erosion. We recommend a CT slice thickness of 0.5–1 mm with a bone algorithm reconstruction including multiplanar reformats. The choice of a small field of view can be helpful to maximise the signal to noise ratio in small structures, for example to delineate the bony labyrinth. Where available, the use of a high field strength (3 T) should be considered, as this produces increased image signal for the resolution of lesion detail and to identify perineural disease.^{2,3} MRI sequences may vary between institutions, but a typical skull base MRI protocol includes T1, T2 and post Gadolinium contrast T1 sequences at a slice thickness ≤ 3 mm. High resolution T2 CISS (constructive interference in steady state) imaging improves the accurate visualisation of the cranial nerves, particularly in the basal cisterns and internal auditory meatus, in which the nerves stand out from surrounding CSF. Thin-section post Gadolinium T1 imaging is the most sensitive technique to demonstrate perineural tumour spread, presence of which usually worsens the prognosis and may preclude cure if unrecognised.^{4,5} Diffusion-weighted imaging at the skull base remains technically challenging due to artefact and distortion from the bone air interface, which can be reduced by employing a non-echo planar sequence. Although reliable threshold values are difficult to define, malignant tumours tend to display lower diffusivity than benign disease with the exception of certain non-neoplastic lesions such as epidermoid cysts and cholesteatoma, which are known for their characteristic diffusion restriction.⁶ Bone marrow invasion may be evident on unenhanced T1 images or STIR (short tau inversion recovery), which can also unmask metastatic lymph node involvement. Post Gadolinium imaging generally aids the delineation of solid tumours including differentiation of peripherally enhancing inflammatory sinus disease from tumour. Some controversy exists regarding the value of fat-suppressed post contrast imaging of the skull base due to a potential risk of artefact from incomplete fat suppression being mistaken for enhancing tumour.⁷ Time-of-flight MR angiography (TOF-MRA) is a flow imaging technique, which gives information about the patency and position of the cranial arteries in relation to tumour. This has relevance for surgical planning, although MRA may not accurately depict blood supply for all lesions. Therefore digital subtraction angiography (DSA) retains an important role in delineating the arterial supply of hypervascular tumours, particularly in candidates for embolisation treatment. This may be performed either as an adjunct prior to radical surgery or as a palliative measure where curative resection is not possible.⁸ Recently PET/CT, specifically 18-F Fluorothymidine imaging, has been suggested as potentially beneficial in head and neck cancer staging.⁹

3. The role of imaging

As the imaging features of some skull base tumours overlap, radiologists may appropriately refrain from giving a definitive tissue diagnosis.¹⁰ It is however important to accurately describe the anatomic spread of the tumour and identify

the structures involved as this will determine the lesion resectability and would help planning the optimal and safest route for biopsy. For example, invasion of vascular structures such as the ICA or cavernous sinus could preclude radical resection. Imaging evidence of tumour hypervascularity may alter the differential diagnosis and increases the biopsy risk. Orbital and optic nerve involvement may prevent curative surgery, especially if bilateral.¹ Meningeal spread and brain invasion, as well as extension to surgically less accessible sites such as the paranasal sinuses or the prevertebral space should always be assessed for. Specialist centre referral onto an established multidisciplinary pathway represents internationally accepted best practice for patients with complex skull base lesions.

4. Anterior skull base

4.1. Anatomical boundaries

The anterior skull base separates the cranial contents from the paranasal sinuses and orbits. It extends from the posterior wall of the frontal sinuses to the anterior clinoid processes and lesser sphenoid wing. Laterally, it is bound by the orbital plates of the frontal bone. In its centre, it contains the cribriform plate of the ethmoid bone, a thin osseous layer which is susceptible to damage. The upcoming section will focus on olfactory groove, sinonasal neoplasms and the orbits.

4.2. Anterior skull base tumours

4.2.1. Olfactory groove

The olfactory groove is a paired furrow in the cribriform plate on either side of the crista galli. It transmits the olfactory nerves and anteriorly contains a small foramen for the nasociliary nerve, a branch of the ophthalmic nerve (V1).

4.2.1.1. Meningioma. Meningioma commonly arises in the olfactory groove, though this tumour may occur anywhere along the skull base. Other typical sites include the planum sphenoidale, parasellar region, clivus and tentorium cerebelli. Meningiomas represent 13–26% of all intracranial masses with a female to male ratio of 2:1 and most often occur beyond the 5th decade.^{11–14} On CT, meningiomas may show associated hyperostosis as a supportive finding and approximately 75% appear hyperattenuating to brain. Classic MRI appearances are those of a T1 and T2 isointense lesion with avid contrast enhancement and an enhancing dural “tail”, but this is not a necessary or pathognomonic feature (Fig. 1a and b). Depending on their size and location, these lesions may be surprisingly indolent, but certain features such as progressive optic nerve compromise can require emergency intervention.¹⁴

4.2.1.2. Olfactory neuroblastoma. Olfactory neuroblastoma (synonym esthesioneuroblastoma) is a neuroendocrine tumour arising from the olfactory mucosa. The incidence of this aggressive neoplasm peaks in the second and sixth decade, although it can occur at any age.¹⁵ Soft tissue in the olfactory niche, which could be confused with a small meningioma or sinus disease, may be the earliest imaging evidence

Fig. 1 – (a) and (b) Coronal and sagittal post Gadolinium T1 images demonstrating an olfactory groove meningioma with a dural tail (arrow). (c) Large right sphenoorbital meningioma in a different patient with proptosis, optic nerve compression and intracranial extension. Note the hyperostotic expansion of the adjacent sphenoid wing (arrow).

Fig. 2 – Coronal post Gadolinium T1 images in two different patients demonstrating subtle enhancing tissue in the right olfactory groove (a, white arrow) and an extensive destructive lesion (b) involving the anterior skull base and paranasal sinuses with evidence of prior surgery. In both patients, the proven histological diagnosis was olfactory neuroblastoma. Patient (a) also has an incidental mucus retention cyst in the left maxillary antrum (black arrow) characterised by minimal rim enhancement.

(Fig. 2a). Olfactory neuroblastoma frequently spreads to the ipsilateral maxillary and ethmoid sinuses; meningeal and olfactory bulb invasion is common in advanced tumours (Fig. 2b). On MRI, olfactory neuroblastomas usually show intermediate T1 signal and mild T2 hyperintensity with avid homogenous enhancement due to hypervascularity. Small peripheral cysts and calcification have been reported as characteristic features, but may be absent.¹⁶ On CT, bone destruction may disclose the malignant nature of olfactory neuroblastoma and represents a distinguishing feature from meningioma.

Fig. 3 – Not a tumour: Sagittal T2 image demonstrating a mass within an anterior skull base defect. This sincipital encephalocele could be mistaken for a tumour, unless the herniation of brain tissue (arrow) is appreciated. (Courtesy of Dr K. Mankad, Great Ormond Street Hospital, London.)

4.2.2. Sinonasal region

Situated immediately below the osseous anterior skull base, the sinonasal space represents a common primary site for benign and malignant neoplasms. The cribriform plate, which forms the roof of the nasal cavity, provides relatively little resistance to spreading cancers.

4.2.2.1. Benign sinonasal masses.

4.2.2.1.1. *Benign sinonasal masses.* Benign sinonasal masses consist of polyps, inflammatory mucosal disease, haemangiomas and inverted papillomas, whereby polyps represent the vast majority (approximately 70%).¹⁷ Benign lesions may require resection if causing obstructive symptoms, but do not usually invade the skull base. A sincipital encephalocele is a congenital malformation, which may on first glance mimic an anterior skull base tumour, but the presence of herniating brain tissue (Fig. 3) should alert to the correct diagnosis.

4.2.2.1.2. *Juvenile angiofibroma.* This tumour arises in the posterior nasal space from primitive vascular tissue at the sphenopalatine foramen. It occurs in adolescence and is noteworthy because of its relatively characteristic appearances and high bleeding risk on biopsy. The typical imaging finding is that of a T1 hypointense and T2 intermediate signal lesion exhibiting avid enhancement with internal flow voids and extension into the pterygopalatine fossa (Fig. 4) as a hallmark sign. These features, together with the patient's age, can help in differentiating juvenile angiofibromas from other nasopharyngeal lesions.¹⁸ Juvenile angiofibromas may destroy surrounding bone and invade the skull base or orbit. DSA accurately demonstrates the vascular supply of these neoplasms, which most commonly originates from distal internal maxillary artery (IMA) branches, particularly the sphenopalatine, descending palatine, and posterior superior alveolar branches. Blood supply may also be present via the ascending pharyngeal artery. Presurgical embolisation should

Fig. 4 – Axial T1 image: Avidly enhancing juvenile angiofibroma presenting as a posterior nasal space mass with extension into the right pterygopalatine fossa, sphenoid sinus and masticator space.

be considered for these hypervascular tumours to reduce the surgical risk and blood loss. Despite their benign histology, JNA are locally aggressive and tend to recur if not completely resected.

4.2.2.2. *Sinonasal malignancy.* Imaging is unreliable in the differentiation of sinonasal cancer subtypes, but is essential for the identification of malignant features and for disease staging. Bone invasion constitutes a suspicious sign, which may manifest as lysis on CT or loss of the normal bone low signal rim on T1 MRI. Sinonasal tumours can easily spread along the lamina papyracea into the orbit and via the cranial nerves into the central skull base.¹⁰ Solid enhancement distinguishes these tumours from retained secretions (Fig. 5) and mucosal inflammation with the caveat that large tumours can show central necrosis.

4.2.2.2.1. *Squamous cell carcinoma.* Squamous cell carcinoma (SCC) is the most common malignancy of the paranasal sinuses, typically affecting men over the age of 50 years.¹⁵ SCCs account for 80% of sinonasal malignant tumours with 25–58% in the maxillary sinus, 25–35% in the nasal cavity, 10% in the ethmoid air cells and 1% in the frontal sinus.¹⁹ Known risk factors include exposure to Nickel, wood dust, a number of chemicals, alcohol and tobacco. The imaging features of sinonasal SCC are non-specific, however, the presence of unilateral sinus disease, especially if associated with a large soft tissue mass and necrosis should raise concern.²⁰ As CT may be first test performed for ongoing rhinosinusitis, identification of bone invasion becomes particularly important. SCC may exhibit intermediate to low T2 signal as a hallmark of cellularity, which together with solid enhancement represents a distinguishing feature from benign mucosal disease. Meticulous staging is very important, because synchronous head and neck cancers or pulmonary malignancy may be present in approximately 15% of patients.

Fig. 5 – (a) and (b) Axial and coronal post Gadolinium T1 images showing a SCC centred on the left sided nasal space with invasion of the ethmoid air cells, orbit and secondary obstruction of the left maxillary antrum (non-enhancing secretions, arrow).

4.2.2.2.2. Adenoid cystic carcinoma. Adenoid cystic carcinoma is a slow growing but highly aggressive malignancy with the highest local recurrence rate of all sinonasal tumours (75–90%).²⁰ Perineural spread represents a hallmark feature of these neoplasms, which may account for their characteristic delayed recurrence after up to 20 years (for more information on perineural spread see Section 5.2.2.1). Adenoid cystic carcinomas are usually of intermediate T2 signal, but low grade lesions can mimic inflammatory disease by exhibiting T2 hyperintensity.

4.2.2.2.3. Adenocarcinoma. Adenocarcinoma (Fig. 6) represents around 10% of nasal cavity malignant tumours. Similar to SCC, a particular association has been reported between tumour incidence and toxic exposure the hardwood and shoe industry.²¹

4.2.2.2.4. Sinonasal undifferentiated carcinoma. Sinonasal undifferentiated carcinoma is a particularly

Fig. 6 – (a) and (b) Axial CT and post Gadolinium coronal T1 images featuring a destructive frontoethmoidal tumour with a histological diagnosis of adenocarcinoma.

aggressive malignant tumour, often diagnosed in an advanced stage involving the orbit, paranasal sinuses and anterior cranial fossa.¹⁹

4.2.2.2.5. Mucoepidermoid carcinoma. Mucoepidermoid carcinoma, although more common in the parotid gland, may arise from minor salivary glands of the hard palate and typically exhibits T1 shortening on MRI due to proteinaceous content.

4.2.2.2.6. Sarcoma. Approximately 15–20% of head and neck sarcoma occurs in a sinonasal distribution and is morphologically indistinguishable from the carcinomatous malignancies.

4.2.2.2.7. Sinonasal melanoma. Sinonasal melanoma is the commonest mucosal melanoma, although overall it only accounts for 2.5% of all melanomas.²¹ Melanomas often arise from the nasal septum or turbinate remodelling bone in a deceptively benign manner, although erosion occurs eventually (Fig. 7).⁹ These neoplasms may show intrinsic T1 shortening due to its melanin content and enhance with contrast. Desmoplastic melanoma has a striking propensity to

Fig. 7 – (a) Axial CT featuring right sided nasal mucosal thickening with subtle erosion of the middle turbinate. (b) Axial post Gadolinium T1 image obtained 9 months later demonstrating a destructive lesion involving the nasal mucosa and both maxillary antra. The diagnosis in this case was sinonasal melanoma.

invade nerves, presumably due to high expression of the p75 neurotropic receptor, which is also found in a proportion of adenoid cystic carcinoma.¹⁵

4.2.2.2.8. Non-hodgkins lymphomas. Non-hodgkins lymphomas represent approximately 5% of head and neck malignancies and may involve almost any site in this region.²² Among sinonasal lymphomas, lesions arising from the nasal septum or Waldeyer's lymphatic ring are relatively common.¹⁹ Early imaging signs may be confined to non-specific mucosal thickening, with subsequent florid bone destruction (Fig. 8a). In immunocompetent patients with no predisposing factors, the diagnosis is often delayed owing to the relative rarity of the disease and its lack of pathognomic appearances. Sinonasal lymphoma tends to exhibit intermediate signal intensity on all sequences, although T2 hyperintensity is also recognised.²⁰ B-cell lymphoma typically presents as a large soft tissue mass, which is commonly associated with bone remodelling although bone destruction also occurs. On the

Fig. 8 – (a) Coronal CT showing nasal lymphoma with bone destruction and (b) coronal post Gadolinium T1 image in a different patient demonstrating sinonasal lymphoma spreading to the central skull base.

contrary, T-cell lymphoma may result in bone destruction in excess of the soft tissue mass.²⁰ Diffusion restriction is a known feature of lymphoma and a marker of its relative cellularity. Accompanying fever and weight loss in addition to local signs such as nasal obstruction and epistaxis could prompt suspicion, but are hardly specific. Primary lymphoma of the osseous skull base is distinctly uncommon; however, sinonasal lymphoma may invade the anterior skull base directly or reach the central skull base via perineural spread (Fig. 8b).

4.2.3. Orbital region

Due to their close proximity, anterior skull base lesions may involve the orbits, and vice versa. The roof of the orbit forms the floor of the frontal sinus and part of the anterior cranial fossa. Medially, each orbit is separated from the ethmoid air

cells by the lamina papyracea. The orbital apex forms the transition to the central skull base.

4.2.3.1. Skull base lesions with orbital invasion. If sufficiently large, any tumour of the anterior and central skull base may spread to the orbits. Aggressive neoplasms such as **olfactory neuroblastoma** and **malignant sinonasal tumours** have the destructive potential to invade the orbit, especially via the thin lamina papyracea. In a relevant clinical context, the presence of visual symptoms or ophthalmoplegia should prompt urgent imaging. CT imaging provides a good natural contrast between bone, fat, muscles and vitreous, whereby bone erosion and loss of normal fat planes can be subtle markers of involvement. Orbit targeted MRI sequences (e.g. coronal STIR, axial and coronal pre and post contrast T1 imaging at a slice thickness of 3 mm) can be helpful to depict orbital invasion. A common skull base lesion to spread via the orbital apex towards the globe is **meningioma** (Fig. 1c), but these lesions can also arise within the orbit itself from the optic nerve dural layer.

4.2.3.2. Orbital tumours with spread to the skull base. Intracranial extension of an optic nerve sheath **meningioma** or **optic nerve glioma** are both well demonstrated on imaging and can be distinguished by the presence of nerve sheath enhancement versus an intrinsic optic nerve mass. Skull base dissemination including perineural spread may be encountered in malignant **orbital neoplasms** such as **metastatic disease (especially breast and lung)**, **lymphoma** and **uveal melanoma**.²³ There are no pathognomonic features, but if the epicentre of the tumour is clearly located within the orbit, this can help to narrow the differential diagnosis. Certain benign lesions such as cavernous haemangioma less commonly extend beyond the orbital borders. Nerve sheath tumours, particularly ophthalmic nerve Schwannoma may rarely be encountered at the roof of the orbit bordering the anterior cranial fossa. Benign lacrimal gland lesions can erode the roof of the orbit, the most common being pleomorphic adenoma.¹⁰

5. Central skull base

5.1. Anatomical boundaries

The central skull base extends from the anterior clinoid processes and lesser wing of the sphenoid to the petrous ridge and temporal bones, incorporating the middle cranial fossa laterally on each side. In its centre, it contains the sella turcica and part of the clivus. The spheno-occipital synchondrosis marks the posterior border of the central skull base medially and the petroclival synchondrosis laterally. The central skull base contains a large number of important structures including the ICA, cavernous sinus, optic canal, superior orbital fissure, the foramina rotundum, ovale and spinosum and the vidian canal. The upcoming section will focus on tumours of the sella, the parasellar region and the clivus including osseous neoplasms of the central skull base in the latter section.

5.2. Central skull base tumours

5.2.1. Sella turcica

The sella turcica forms part of the sphenoid body containing the pituitary gland in its central hypophyseal fossa. The anterior border of the sella is defined by the tuberculum sellae and its posterior border by the dorsum sellae and posterior clinoid processes. Tumours in this region include pituitary microadenoma and macroadenoma, meningioma and craniopharyngioma.

5.2.1.1. Pituitary microadenoma. Pituitary microadenoma (<1 cm) may be discovered incidentally or through excess hormone production with prolactinoma being the commonest. Small lesions are best depicted by MRI, which typically demonstrates low T1 and increased T2 signal with reduced Gadolinium enhancement compared to the remainder of the gland (Fig. 9a). Dynamic post contrast imaging can improve the visualisation of altered contrast uptake in microadenomas, where this is not appreciable on conventional post Gadolinium sequences. A typical indication for dynamic imaging would be following a normal or inconclusive conventional pituitary MRI in a patient with proven excess serum hormone levels.

5.2.1.2. Pituitary macroadenoma. Pituitary macroadenoma (>1 cm) often extend into the suprasellar compartment, classically giving rise to a “snowman” or “Fig. 8” morphology (Fig. 9b), potentially compressing the optic nerve and/or the optic chiasm. Pituitary adenomas invade the cavernous sinus in 6–10% of cases, often unilaterally, thereby limiting surgical resectability.²⁴ Unlike meningiomas, pituitary adenomas rarely narrow the cavernous carotid artery.²⁵ Occasionally, pituitary adenomas can grow inferiorly eroding the sella floor or arising in an exclusively intraosseous location in which case an empty sella appearance maybe the relevant imaging clue.

5.2.1.3. Craniopharyngioma. Craniopharyngioma is a tumour arising in the suprasellar midline from remnants of Rathke's pouch, though it can occur anywhere from the hypothalamus to the nasopharynx. Typical imaging features are calcifications and a cystic-solid appearance with T1 signal shortening due to proteinaceous content (Fig. 10).²⁶

5.2.1.4. Other suprasellar tumours. Other suprasellar tumours include germinoma, glioma and hamartoma, which originate from brain parenchyma as opposed to the skull base.

5.2.2. Parasellar tissues and neural foramina

Immediately bordering the sella laterally on each side is the cavernous sinus, containing the cavernous segment of the ICA (internal carotid artery) and from superior to inferior the transiting cranial nerves III, IV, VI, V1 and V2. Anterior to the sella is the optic canal and superior orbital fissure; infero-laterally is the greater sphenoid wing containing the vidian canal (for the Vidian nerve), the foramina rotundum (maxillary nerve, V2), ovale (mandibular nerve, V3) and spinosum (middle meningeal artery, vein and nerve). Through these apertures, tumours originating outside the cranial cavity invade the central skull base. In the following section, perineural spread,

Fig. 9 – (a) Coronal post Gadolinium T1 image demonstrating a pituitary microadenoma (arrow). (b) Pituitary macroadenoma with suprasellar extension and encroachment on the optic chiasm. The lesion extends to the medial surface of the right cavernous ICA.

meningiomas and schwannomas of the parasellar region will be reviewed.

5.2.2.1. Perineural spread. Perineural spread (PNS) represents an insidious and feared form of metastatic disease, which may propagate along any nerve, most commonly via the trigeminal branches V2 and V3 (Fig. 10) into the central skull base. PNS also commonly affects the descending branch of the facial nerve in face and neck cancers. Although the presence of PNS worsens the prognosis, cure may still be achievable, but this depends on accurate estimation of the disease extent. On post Gadolinium imaging, PNS manifests as nerve thickening and pathological, sometimes nodular, enhancement, which can be subtle and is easily missed unless specifically searched for.

Fig. 10 – (a) and (b) Sagittal pre and post Gadolinium T1 images demonstrating a cystic-solid craniopharyngioma with intrinsic T1 shortening and heterogeneous enhancement.

Loss of normal CSF signal within Meckel's cave also represents a suspicious feature. It should be emphasised here, that normal structures (e.g. venous plexus), can give rise to perineural enhancement, hence enhancement as such does not equate to malignant invasion.²⁷ Multiplanar reconstructions, side comparison and a review alongside normal studies are useful especially in challenging cases. Clinical symptoms of PNS are often vague including neuropathic pain or dysaesthesia, however, close to 50% of patients remain asymptomatic.²⁶ Due to its high incidence, squamous cell carcinoma is the most common neoplasm spreading along cranial nerves.¹⁰ Sinonasal PNS is commonly associated with adenoid cystic carcinoma (in up to 28%), melanoma, lymphoma and sarcoma.²⁸

5.2.2.2. Meningioma. Meningioma is especially common in the parasellar region, whereby "en plaque" lesions can be

Fig. 11 – Coronal post Gadolinium T1 image showing SCC perineural spread along the left mandibular nerve (V3) characterised by nerve thickening with avid perineural enhancement.

missed if small and are difficult to completely resect. For lesions in proximity to the cavernous sinus, narrowing of the cavernous ICA is commonly seen with meningioma.

5.2.2.3. Schwannoma. Schwannoma may occur in the central skull base, most frequently involving the trigeminal nerve (V) (Fig. 11). The most common type of schwannoma is vestibular schwannoma, which will be discussed in Section 6.2.3.1.

5.2.3. Clivus and osseous central skull base

The clivus represents a bone slope composed of sphenoid bone superiorly and occipital bone inferiorly with the sphenoccipital synchondrosis joining the two and marking the border to the posterior skull base. Neighbouring structures of the clivus are the ICA within the foramen lacerum laterally, the basilar artery and pons dorsal to the clivus surface. This section will discuss chordoma, chondrosarcoma, bone metastases and osteosarcoma.

5.2.3.1. Chordoma. Chordoma is a histologically benign but locally invasive neoplasm arising from embryonic remnants of the primitive notochord. Chordomas present as expansile clival tumours, often exhibiting an aggressive pattern of bone destruction with replacement of the clivus bone marrow and erosion of its cortical margins.²⁹ On MRI, they are typically T1 isointense to hypointense and T2 isointense to hyperintense lobulated masses (Fig. 12) showing heterogeneous Gadolinium enhancement. In approximately 25–40%, chordomas destroy the clivus and spread to the middle or posterior cranial fossa. Chordomas occur in adulthood at any age, often presenting with headache and cranial nerve palsies. Local recurrence is common with an overall 5 year survival rate of 60%, however, distant metastases are unusual.³⁰

5.2.3.2. Chondrosarcomas. Chondrosarcomas are rare tumours representing approximately 6% of all skull base

Fig. 12 – (a) and (b) Axial T2-weighted and coronal T1-weighted image through Meckel's cave featuring a trigeminal Schwannoma.

lesions. The head and neck represents a relatively common primary site contributing 10% of all chondrosarcomas with a predilection for the central skull base.^{20,31} Chondrosarcomas tend to occur towards the sixth decade and are uncommon below 40 years of age. The degree of bone destruction increases with tumour grade, whereby well differentiated tumours more often exhibit a classic chondroid matrix, characterised by punctuate or curvilinear mineralisation on CT (Fig. 12). Typical MRI appearances are low T1, high T2 signal with relatively avid contrast enhancement. The imaging features of chondrosarcoma often overlap with those of chordoma, making a biopsy necessary unless the clivus is clearly spared, which only happens in chondrosarcoma.

5.2.3.3. Metastases. Metastases are the most common neoplasm affecting the osseous skull base.¹ Even though they may occur anywhere, the clivus, petrous apex and sphenoid triangle are most frequently affected due to their increased marrow content (Fig. 13).¹⁹ Common primary sites include lung, breast, kidney and prostate, whereby imaging evidence of multifocal disease and a relevant past medical history can be useful diagnostic indicators.

Fig. 13 – (a) and (b) Axial T2 and unenhanced sagittal T1 image demonstrating a well-defined clival chordoma.

5.2.3.4. Osteosarcoma. Osteosarcoma is a very rare tumour of the skull base with an estimated 100 cases of reported to date.³¹ These neoplasms occur with an age peak in the third decade, slightly later than long-bone osteosarcoma. Risk factors for secondary osteosarcoma include Paget's disease, Li Fraumeni syndrome and fibrous dysplasia. Osteosarcoma may occur following cranial radiotherapy in the periphery of the radiation field, very commonly in the jaw presenting with dental pain and loose teeth.³¹ On CT, osteosarcoma may demonstrate a mixture of lysis and sclerosis (Fig. 14a) with cloudy or clustered mineralisation.^{26,32} When present, a Codman triangle or “sunburst” pattern of periosteal reaction is characteristic. Depending on mineral content, osteosarcoma may return variable MR signal, most commonly low T1 and intermediate T2 signal. These lesions tend to enhance less avidly than chondrosarcoma.

5.2.3.5. Fibrous dysplasia. An important non-neoplastic bone condition, which may affect the skull base is **fibrous dysplasia**. FD is caused by an activating mutation in the GNAS gene

Fig. 14 – (a)–(c) Axial CT, axial T2 and sagittal post Gadolinium T1 image showing chondroid matrix, T2 hyperintensity and heterogenous enhancement within a chondrosarcoma.

and can occur within a single bone ('monostotic' FD) or involve multiple sites ('polyostotic' FD), with the most severe form of polyostotic FD occurring in the McCune-Albright syndrome. This expansile bone disease could on first glance mimic a malignant tumour, but does not show aggressive features

or growth. On CT, fibrous dysplasia exhibits a characteristic ground-glass like fibrous matrix (Fig. 14b), sometimes mixed with areas of sclerosis and lucency and a variably defined border to surrounding normal bone. On MRI, fibrous dysplasia typically returns reduced T1 and T2 signal. Whilst the condition is entirely benign, it should be remembered that secondary osteosarcoma can arise on a background of long-standing fibrous dysplasia.

6. Posterior skull base

6.1. Anatomical boundaries

Centrally, the posterior skull base extends from the sphenoccipital synchondrosis to the occipital inner table and laterally from the petrous ridge to the temporal and occipital inner table. Important apertures of the posterior skull base are the internal auditory meatus, jugular foramen, hypoglossal canal and foramen magnum. In the upcoming sections, tumours of the petrous temporal region, cerebellopontine angle, vestibular aqueduct and foramen magnum will be discussed.

6.2. Posterior skull base tumours

6.2.1. Petrous temporal region

The petrous ridge marks the anatomical border between the middle and posterior cranial fossa, but tumours do not respect this division and can involve either compartment or both.

6.2.1.1. Glomus tympanicum. Glomus tympanicum is the most common middle ear neoplasm second only to vestibular schwannoma.³⁴ These lesions may occur at any age, but are most common between 40 and 70 years. Glomus tumours are paragangliomas consisting of chromaffin cells separated by vascular channels, giving rise to their characteristic “salt and pepper” appearance on MR imaging caused by a combination of avid enhancement and low signal flow voids. When assessing glomus tumours, it is useful to combine CT imaging to delineate petrous bone detail and thin-section MRI, which is superior at identifying soft tissue enhancement. Glomus tympanicum classically arises at the cochlear promontory (Fig. 14) manifesting on otoscopy as a small red mass within the anterior inferior quadrant of the tympanic membrane. Alternative locations along the medial wall of the middle ear cavity may be encountered, along the tympanic branch of the glossopharyngeal nerve (IX, Jacobson’s nerve) and the auricular branch of the vagus nerve (X, Arnold’s nerve).³⁴ The typical clinical presentation is with pulsatile tinnitus, although this is not a specific symptom.

6.2.1.2. Glomus jugulare. Glomus jugulare arises from glomus bodies related to the jugular bulb (Fig. 15). Its imaging features match those of glomus tympanicum; in fact, with large tumours it can become difficult to distinguish the two entities. Specific imaging signs of glomus jugulare include widening of the jugular foramen, frequently with evidence of erosion on CT. Another typical feature of glomus jugulare is erosion of floor of the tympanic cavity, which is

Fig. 15 – (a) Axial unenhanced T1 image demonstrating loss of the normal skull base bone marrow signal (arrows), which was subsequently proven to represent metastatic infiltration from breast cancer. (b) Normal appearances (N) for comparison.

unusual for glomus tympanicum.³⁴ Clinical symptoms include impaired hearing with pulsatile tinnitus, vestibular dysfunction and compromise of cranial nerves within the jugular foramen, with the glossopharyngeal and vagus nerves being especially vulnerable to compression.³⁶ Larger glomus jugulare tumours can extend cranially into the hypotympanum and mimic glomus tympanicum by being visible on otoscopy. Whenever the diagnosis of a glomus tumour is being queried, it is important to consider vascular mimics, which could cause catastrophic consequences on biopsy, such as an ectopic course of the internal carotid artery (ICA), a high riding internal jugular vein with bone dehiscence or an ICA aneurysm. Although glomus tumours are relatively common middle ear lesions, their differential is very wide ranging from acute inflammation to cholesteatoma and non-glomus tumours.³⁷

Fig. 16 – (a) Axial CT demonstrating a lytic-sclerotic osteosarcoma of the left maxilla (featuring clips relating to previous surgery) with diffuse infiltration of the ipsilateral sphenoid base and clivus. (b) Not a tumour: Expansile ground glass matrix (arrow) of fibrous dysplasia; note the absence of bone destruction.

6.2.2. Vestibular aqueduct

The vestibular aqueduct represents a bony canal within the petrous temporal bone containing the endolymphatic duct, which connects the membranous labyrinth of the inner ear to the posterior temporal endolymphatic sac.

6.2.2.1. Endolymphatic sac tumour. Endolymphatic sac tumour (ELST) is a rare, locally aggressive bone tumour arising from endolymphatic epithelium in the posterior petrous temporal bone.³⁸ Invasion of the mastoid, semicircular canals, cerebellopontine angle and cranial nerves is commonly present at diagnosis. Proximity to the vestibular aqueduct is a key feature of ELST, as these tumours do not arise within the aqueduct but at its posterior margin corresponding to the expected position of the endolymphatic sac. Among other neoplasms, this tumour frequently occurs in individuals with von Hippel-Lindau syndrome (Fig. 16).

Fig. 17 – (a) and (b) Axial CT and post Gadolinium T1 image showing typical features of a small glomus tympanicum arising on the cochlear promontory (arrow).

6.2.3. Cerebellopontine angle

The cerebellopontine (CP) angle represents the junction between cerebellum and pons, from which the facial (VII) and vestibulochochlear (VIII) nerves emerge. At a close distance superiorly, the trigeminal nerve (V) traverses the pontine cistern and can become compressed by large CP angle tumours.

6.2.3.1. Vestibular schwannoma. Vestibular schwannoma (VS) is the most frequent CP angle neoplasm accounting for 85% of all CP angle masses and approximately 6–10% of all intracranial tumours.^{39,40} Most VS lesions are slow-growing with an estimated progression rate of 1–2 mm/year.⁴¹ The classic MRI morphology is that of an “ice cream on cone” lesion with its base at the CP angle and its tip (cone) widening the internal auditory meatus (Fig. 17), small lesions however, may be purely intracanalicular. VS is often T1 isointense and T2 hyperintense. Small tumours are often comprised of dense cellular Antoni A regions, which demonstrate avid contrast enhancement. Regions of loosely packed cells, sometimes with cystic features, are designated Antoni type B and may display

Fig. 18 – (a) Axial post Gadolinium T1 image demonstrating a left sided glomus jugulare. (b) Coronal T2 image in a different patient showing a right sided GJ with flow voids causing a “salt and pepper” appearance and (c) corresponding digital subtraction angiogram obtained during right external carotid artery injection showing marked hypervascularity of the lesion.

Fig. 19 – Axial T2 image demonstrating a small endolymphatic sac tumour (arrow) in a patient with multiple cerebellar lesions representing haemangioblastomas (arrow heads) and an underlying diagnosis of von-Hippel Lindau syndrome.

heterogenous enhancement. The distance from the lateral extent of the intracanalicular lesion component to the fundus of the internal auditory meatus is an important predictor of hearing outcome and can influence the surgical approach.⁴²

6.2.3.2. Meningioma. Meningioma is the second most common CP angle tumour and can often be confidently identified, especially when large. However, in the absence of typical features, the distinction from schwannoma can be challenging. Yamamoto et al.⁴³ recently reported a statistically significant difference in tumour blood flow using pseudocontinuous arterial spin labelling MRI with higher values demonstrated in meningioma compared to schwannoma and a potential application in patients unable to receive contrast (Figs. 18–20).⁴³

6.2.3.3. Other differentials. Other differentials for a CP angle mass include an **epidermoid cyst**, which can be identified through its characteristic restricted diffusion and lack of contrast uptake. Occasionally **metastases** arise in the internal auditory meatus. These do not have specific features in the early stages and may be challenging to diagnose, particularly if the primary tumour is occult.

6.2.4. Foramen magnum

The foramen magnum is an osseous ring formed by the occipital bone and represents the most caudal opening in the skull base, transmitting the medulla oblongata and the vertebral arteries. The hypoglossal canal opens into the anterior wall of the foramen magnum on each side.

6.2.4.1. Foramen magnum tumours. Tumours at the foramen magnum fall into the aforementioned categories, mostly consisting of meningioma, nerve sheath tumours (schwannomas and neurofibromas) and chordoma.^{44,45} Lesions confined to the hypoglossal canal can cause cranial nerve XII deficit even when small. In contrast, lesions involving the

Fig. 20 – (a) and (b) Axial post Gadolinium T1 and T2 images demonstrating the typical “ice cream on a cone” appearance of VS.

foramen magnum are often of a substantial size before they cause symptoms. Clinical symptoms of “foramen magnum syndrome” include sensory symptoms such as tingling in the hands, neck pain and motor deficits progressing to quadriplegia.⁴⁵ The surgical resection of foramen magnum lesions is demanding and may benefit from angiographic imaging for operative planning.

7. Conclusion

Skull base tumours are a heterogeneous group of neoplasms, with often overlapping imaging features. Treatment planning is dependent on excellent imaging, typically provided by the combination of thin-section CT and MRI with multiplanar reformats to precisely locate the lesion, assess the integrity of the adjacent structures and suggest an appropriate diagnosis. An interdisciplinary team approach with an intimate understanding of the clinical, neuroradiological, surgical and

radio-oncological aspects is central to improve the outcome of these challenging tumours.

Conflict of interest

None declared.

Financial disclosure

None declared.

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