Posterior skull base: review of the anatomy and pathology

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Authors: A. Tanasa¹, D. Garcia Figueredo¹, R. Contreras Chacon¹, S. Bolivar¹, M. Cuadrado², J. F. Madureira Cordeiro¹, J. R. Torino¹, X. Pruna¹; ¹Granollers/ES, ²Parets del Valles/ES  
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Learning objectives

To know and understand the CT and MR anatomy and pathology of the posterior skull base.

To analyze the different pathologies which can affect this region.

To emphasize on what clinicians need to know in a skull base pathology.
Background

Evaluation of posterior skull base lesions is challenging. On the one hand, the skull base is not directly accessible for clinical evaluation and on the other hand, radiological studies are excellent in demonstrating a skull base lesion and its extent.

Only few skull base lesions present with specific, disease-related symptoms that significantly help to narrow the differential diagnosis. Recognition of anatomical mimics and of medically treatable conditions is essential as the majority of skull base lesions are inaccessible for biopsy. Differentiation of malignant from benign lesions is critical for the proper manage.

Different entities which affect the skull base include congenital pathology, traumas, infections, inflammation and tumours.

In addition, determination of the exact origin and extent of a lesion is crucial for radiation therapy and even more for surgical planning purpose.
Findings and procedure details

POSTERIOR SKULL BASE: ANATOMY AND PATHOLOGY

Imaging interpretation of skull base lesions can be challenging because of their infrequency, the complex nature of the skull base and the ability of normal anatomical variations to mimic pathology.

The goals of the image analysis are to distinguish normal anatomical variations from true pathology, recognize medically treatable conditions and differentiate benign from malignant processes.

The anatomy of the skull base is very complex. Knowledge of the location of the different neuronal connections can explain the wide range of clinical symptoms and facilitate the detection of extracranial tumors spread.

The skull base is formed by the ethmoid, sphenoid, occipital, frontal and temporal bones. It is divided into 3 regions: anterior, central and posterior skull base.

The distinction of the central to posterior skull base is ambiguous as the roof of the petrous apex represents part of the central skull base, while the posterior margin borders the posterior skull base.

LIMITS:

- **Anterior**: the clivus and the petrous apex
- **Lateral**: mastoid-superoanteriorly, petrous portions-inferoanteriorly, occipital bone posteriorly
- **Posterior and inferior**: occipital bone

The jugular foramen and the foramen magnum are the largest opening of the posterior skull base. **Fig. 1 on page 14** **Fig. 2 on page 14**

The jugular foramen lies at the posterior end of the petrooccipital fissure and is bordered by the petrous bone of the temporal bone anteriorly and the occipital bone posteriorly. It is subdivided by the jugular spine into two compartments:

- the pars venosa-posterolateral: contains the internal jugular bulb and the cranial nerves X and XI.
• the pars nervosa - anteromedial: contains the cranial nerve IX and the inferior petrosal sinus.

The internal carotid artery enters the skull base anterior to the jugular foramen. Therefore, all three of these cranial nerves lie between the internal jugular vein and the internal carotid artery below the skull base.

The posterior skull base has few additional smaller openings:

• **The porus acusticus** - the medial opening of the internal auditory canal along the posterior surface of the petrous bone: contains the cranial nerves VII and VIII, branches of the anterior-inferior cerebellar artery that supply the inner ear.
• **Opening of the vestibular aqueduct** - courses almost parallel to the posterior margin of the petrous bone and contains the endolymphatic duct.
• **Opening of the cochlear aqueduct** - courses parallel to the internal auditory canal.
• **Hypoglossa foramen** - courses inferomedial to the jugular foramen: contains the cranial nerve XII, the hypoglossal venous plexus and a meningeal branch of the ascending pharyngeal artery.

**Acoustic schwannoma**

Acoustic neuroma is the most common eighth nerve tumor. It represents 8% of all intracranial neoplasms with 90% arising from the vestibular division, usually the superior segment. They usually arise at the porus acoustic s and within the IAC, then growing into the CPA cistern. 15% of them will have cysts. They may also contain calcium, have a fatty xanthomatous change, dense collagen, prominent vascularity, or intratumoral thrombosis.

Clinically, small acoustic neuromas from the vestibular nerve may cause dizziness, imbalance, nausea and tinnitus. The sensorial hearing loss is usually unilateral and progressive, ultimately, leading to deafness.

The MRI appearance of acoustic neuromas without contrast on T1-weighted images is a mass that is isointense to slightly hypointense compared with gray matter. T2-weighted images show slight hypointensity compared with gray matter. PD-weighted appearance is usually hyperintense. These findings allow for detection of most lesions without contrast. Smaller tumors are more difficult to see unless contrast is administered.
Small schwannomas between 3 to 15mm tend to conform to the tubular shape of the IAC. As the acoustic neuromas become larger, in the 1.5 to 3-cm range they assume a more typical ice-cream cone appearance, with the cone being the intracanalicular component and the projecting portion in the CPA cistern being the ice cream. When the lesions become larger, the CPA cistern component tends to be more dominant and the lesions may become large enough to compress the brain stem and cerebral peduncle.

Malignant melanotic schwannomas have been described and have signal change related to melanin content as well as hemorrhagic degeneration.

If bilateral acoustic schwannoma is seen on MRI, neurofibromatosis type II should be suggested.

**Fig. 3 on page 15**

**Meningioma**

Meningiomas are neoplasms that arise from arachnoid cap cells, dural fibroblasts or arachnoid membrane. The latter accounts for lesions in the cerebellopontine angle (CPA) region.

These tumors are typically benign and well circumscribed, and are epidemiologically and histologically similar to meningiomas that occur elsewhere in the cranial vault. They are the second most common intracranial tumours. If multiple (10%), are associated with NF2.

Common skull base locations are the clivus, sphenoid wing and jugular foramen.

Depending on their location, CPA meningiomas may present with cranial neuropathy, cerebellar dysfunction, or other symptoms caused by local mass effect. Rarely, a meningioma that is limited to the IAC may mimic a vestibular schwannoma clinically.

On non-enhanced CT, meningiomas may be isodense or hyperdense and unlike schwannomas may show calcification in up to 25%. The presence of hyperostosis on CT also suggests a diagnosis of meningioma.

On MRI, meningiomas show T1 signal that is roughly similar to the brain, and may be hyperintense or hypointense on T2W1. The hallmark imaging features of meningioma include extra-axial dural-based location, intense enhancement, hyperostosis and
calcification. The dural origin of these tumors typically results in a hemispheric mass with a broad dural margin that forms an obtuse angle against the adjacent bone, compared with the spheroid or nodular mass with acute angle typically seen in schwannomas. The presence of a dural tail, also favors the diagnosis of meningioma.

A CPA meningioma may also extend into the IAC and differentiation between meningioma and schwannoma may be difficult.

**Fig. 4 on page 16**

**Paraganglioma**

Paraganglioma are rare tumors of neural crest origin, that may affect nerve function of the cranial nerves IX, X and XI when arising within the jugular foramen or along the course of the vagus nerve. Usually nonfunctioning, paragangliomas most commonly arise from the carotid bifurcation (carotid body), the cervical carotids space (vagal), the jugular foramen (jugular) and within the middle ear ( tympanic), in decreasing order of frequency. Paragangliomas may be multifocal.

A jugular paraganglioma typically compresses cranial nerves IX to XI within the jugular foramen.

The majority of them are benign, only 13% are malignant, but both of them demonstrate locally aggressive growth pattern making them indistinguishable from each other. It has been suggested that the incidence of pain and hearing loss is higher with the malignant form. The main differentiating feature, however, is the existence of metastatic lesions in the malignant form. Metastatic disease spreads to the bone, lung and cervical lymph nodes and might present even years after the initial diagnosis and treatment.

Vagal paraganglioma usually displace the internal carotid artery anteriorly whereas carotid body tumors displace it posteriorly. Vagal paragangliomas may extend superiorly into the jugular foramen, simulating a jugular paraganglioma. Carotid body, jugular and vagal paragangliomas may cause arterial or venous compression or encasement.

The primary diagnostic considerations with a jugular foramen mass are paraganglioma, schwannoma or neurofibroma, and metastatic disease.

Schwanoma and neurofibromas usually appear as a sharply demarcated tumor centered in a smoothly enlarged jugular foramen, with sharply rounded bony margins. Highly vascular jugular paragangliomas usually display intense contrast enhancement, flow
voids within the tumor are often visible, and bony infiltration or destruction is typically present. A "salt- and -pepper" appearance may be apparent on T2-weighted MR images.

Jugular paragangliomas produce irregular erosion of the enlarged jugular foramen margins, the earliest signs being loss of the carotid-jugular spine.

**CT:** glomus jugular tumor causes destruction of the walls of the jugular foramen and adjacent petrous bone giving the "mouth eaten" appearance.

**MRI:** hypervascular mass with flow voids and extensive enhancement.

It tends to grow into the middle ear cavity and posterior fossa.

**Fig. 5 on page 17**

**Perineural spread**

Perineural spread is common in head and neck malignancies. It can be seen in squamous cell carcinoma, malignant melanoma, lymphoma, basal cell carcinoma, adenocarcinoma, mucoepidermoid tumor, rhabdomyosarcoma, chondrosarcoma, malignant mixed tumor, and esthesioneuroblastoma.

Perineural spread is usually associated with cavernous sinus, cranial nerve infiltration, skull base invasion, and has bad prognosis.

Perineural spread occurs commonly along cranial nerves V, III, IV, and VI through foramen rotundum, foramen ovale, superior and inferior orbital fissures. Perineural spread in parotid malignancies occurs along the facial nerve via stylomastoid foramen. Perineural spread along the cranial nerves IX, X, XI, and XII with intracranial extension occurs along the jugular foramen and hypoglossal canal.

**Fig. 6 on page 18**

**Metastases**

Metastasis to the skull-base particularly affects patients with carcinoma of the breast, lung, kidney and prostate.
Clinically, the key feature is progressive ipsilateral involvement of cranial nerves.

MRI is nowadays the most useful examination to establish the diagnosis but plain films, CT scans with bone windows and isotope bone scans remain helpful to demonstrate bone erosion.

Median survival is about 2.5 years.

CT:
1. Infiltrative soft tissue mass with bone erosion.
2. More frequent: Lytic mass with scalloped, poorly marginated, non-sclerotic margins.
3. May be sclerotic (e.g. prostate) or expansive (thyroid and kidney)
4. Multiple calcifications are often visible.

MRI:
1. Iso-hypointense on T1 weighted images heterogeneous.
2. Moderate signal intensity on T2 weighted images (in high cellular tumors)
3. Moderate homogeneous contrast enhancement with central inhomogeneity.

**Fig. 7 on page 19**

**Hemangioma**

Although hemangiomas are common, primary hemangiomas arising in the skull are infrequent.

Hemangiomas which originate primarily in bone are to be distinguished from vascular neoplasm of the soft tissue which secondarily invade the bone by direct extension. Apart from the vertebral column in which the majority occurs, the skull seem to be most frequently involved than other bones.

Primary hemangiomas are always benign. The lesions tend to remain stationary over long periods of time. Normally they are solitary with well circumscribed margins.

**CT:** Expansile lesion sharply marginated, with thin peripheral, sclerotic rim en 30% that may deform overlying soft tissues. Enhance after the administration of intravenous contrast.
**MRI:** T1 - isointense with brain. Small lesions may appear hyperintense because of the fatty tissue. Large hemangiomas typically are hypointense secondary to presence of thickened trabeculae.

T2 - often hyperintense because of the slow flow or pooling with blood.

Enhance after the administration of intravenous contrast.

**Fig. 8 on page 20**

**Histiocytosis**

Langerhans cell histiocytosis (LCH) which originates from immature Langerhans cells is a clonal myeloproliferative disease with a variety of clinical presentations and prognosis.

Although the lesions can be asymptomatic, it can present with swelling, pain, or pathological fractures in the site of involvement.

Bony involvement is the most frequent. It is estimated that 15-61% of patients with Langerhans’ cell histiocytosis have otologic involvement. Although more frequent in children with multisystemic disease, otologic symptoms with no other clinical findings may be the initial form of presentation in 5-25% of patients.

Temporal bone lesions may present with the signs and symptoms of an ear infection, which may result in a delay of diagnosis and treatment. The lack of response to antibiotics, combined with appropriate imaging studies, usually expands the clinical differential diagnosis to include Langerhans cell histiocytosis, among other lesions (such as rhabdomyosarcoma).

**CT:** - Lytic lesions with soft tissue masses that enhances after the administration of intravenous contrast.

**MRI:** T1- soft tissue mass at site of bony lysis, with strongly enhancement after the administration of intravenous contrast.

T2- mastoid lesions: soft tissue masses show slight hyperintensity.

Classically, local treatment of Langerhans’ cell histiocytosis of the temporal bone included mastoidectomy and radiotherapy, although the current tendency is to use local injection of
steroids. Treatment of the multisystemic form includes chemotherapy, steroid injections, and immunotherapy.

The clinical course of Langerhans' cell histiocytosis is unpredictable.

**Fig. 9 on page 21**

**Paget disease**

Paget disease (osteitis deformans) is a chronic skeletal disorder characterized by abnormal and excessive remodeling of bone. Abnormal osseous resorption and apposition in Paget disease produce variable clinical and imaging manifestations. Although the disease may be asymptomatic, it can be painful or deforming and associated with various and debilitating musculoskeletal complications.

The primary event in Paget disease is intense focal resorption followed by disorderly bone formation.

Three major phases are recognized:

1. The lytic phase (incipient active), in which osteoclastic resorption predominates;
2. The mixed phase (active), in which there is both osteoclastic and osteoblastic hyperplasia with predominant osteoblastic activity
3. The blastic phase (late inactive), in which osteoblastic activity gradually declines.

Involvement of the base of the skull, in particular, may cause platybasia, basilar invagination, and hydrocephalus.

The major techniques available for imaging patients with Paget disease are radiography, bone scintigraphy, CT, and MRI.

**RX:**

Radiography is traditionally considered the initial imaging examination for diagnosing Paget disease of bone.

- The lytic, or incipient active, phase of the disease is characterized by intense osteoclastic activity that is exhibited on radiographs as osteolysis. In the skull, advancing osteolysis is noted as large areas of radiolucency
usually in the frontal and occipital bones and is designated "osteoporosis circumscripta". Skull lesions are most prominent in the inner calvarial tables and usually cross the suture lines.

- The mixed (middle) phase of Paget disease bears characteristics of both the lytic (initial) and the blastic (late) phases. In the skull, abnormal bone deposition assumes a characteristic "cotton-wooll "appearance with globular to fluffy foci of variable density.
- The blastic, or late inactive, phase of Paget disease is characterized by diminished osteoblastic activity and manifests as osteosclerosis.

**CT** facilitates the diagnosis of pagetic abnormalities in bone like those detected by radiography. Because CT images generally provide superior cortical and trabecular detail in a cross-sectional display, CT conspicuously exhibits the classic findings of Paget disease that include osteolysis, trabecular coarsening, cortical thickening and osseous expansion.

**Fig. 10 on page 22**

**Chordoma**

Skull base chordomas are benign locally invasive neoplasms originating from embryonic remnants of notochord in the basisphenoid and may grow to involve multiple areas of the cranial base, and occasionally erode into the intradural space to encompass neurovascular structures and compress the brainstem.

The relationship of the tumor to the internal carotid, vertebral, and basilar arteries, cavernous sinus, and brainstem determines the operability.

The skull base chordomas can extend ventrally to the anterior cranial fossa or caudally involving the upper cervical spine. It may involve the petrophenoclival junction, petrous apex, occipital condyle, and jugular foramen.

Most of the clival chordomas are completely extradural.

However, some invade the dura mater or extend into the intradural space, causing mass effect on the brainstem.

The classic appearance of clival chordoma with high-resolution CT is that of a centrally located, well-circumscribed, expansile soft-tissue mass arising from the clivus with associated extensive lytic bone destruction. Intratumoral calcifications indicate bone destruction rather than dystrophic tumoral calcifications.
There is moderate to marked enhancement following administration of iodinated contrast material.

**Chondrosarcoma**

Chondrosarcoma are a rare malignant cartilaginous tumors of variable aggressiveness. Their aggressiveness depend upon the tumor subtype and grade. The classic variant is the most common and represents a low grade chondrosarcoma.

The most common symptoms are headache and diplopia related to cranial nerve VI palsy. Usually occur in younger patients with a peak incidence in the second and third decades and often are off midline (petroclival) in location. Their paramedian location predisposes to lower cranial nerve deficits in particular hearing loss.

**CT**: A destructive mass centered in the petrous apex with rings and arcs in it reflecting the chondroid nature of this tumor.

Soft tissue component, isodense to the adjacent brain parenchyma, with variable enhancement following intravenous contrast administration.

**MRI**: T1- low to intermediate signal intensity

T2- high signal intensity

Variable enhancement pattern.
Fig. 1

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## Fig. 2

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Fig. 3

51 years old, Acoustic trauma, hearing loss and tinnitus, after corticoid therapy, improves the hearing but persists the tinnitus, MRI

Right Vestibular schwannoma
Fig. 4

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52 years old, Bilateral tinnitus >> MRE nodular lesion in the left jugular foramen, pars nervosa, below the cranial nerve VII and VIII hypointense on T₁, hyperintense on T₂ and with heterogeneous enhancement.

N. IX schwannoma

Fig. 5

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Fig. 6

73 years old.

Operated parotid neoplasm and radiotherapy posteriorly who consults for right facial paralysis. CT: pachymeningeal thickening of the right cerebellopontine angle with remodeling of the internal auditory canal and expansive lytic lesion in the right petrous apex in relation to perineural infiltration of N. VII secondary to parotid neoplasm.

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Fig. 7

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Fig. 8

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Fig. 9

14 years old,
Hearing loss and sporadic otorrhea predominantly right, of 2 years of evolution.

CT: Left: coalescent otomastoiditis with left sigmoid sinus venous thrombosis and Right: chronic otomastoiditis

RIGHT BIOPSY: hystiocitosis

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Fig. 10

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Patient 89, who suffers a fall and went to the emergency with headache and right facial paralysis.

Cranial CT was performed to rule out subdural hematoma. Soft tissue lesion right lateral region that affects right temporomandibular articulation with erosion of the right mandibular condyle. We didn’t administer endovenous contrast because of renal failure.

At biopsy: BENIGN MESENCHYMAL TUMOR
Fig. 12

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Conclusion

CT and MR studies provide very useful information of the skull base anatomy and of the extension of many pathologic processes contributing to a better therapeutic planning of every patient.
Personal information

A. Tanasa, Department of Radiology, FHAG - Granollers's Hospital. Granollers, Spain.

D. Garcia, Department of Radiology, FHAG - Granollers's Hospital. Granollers, Spain.

R. Contreras, Department of Radiology, FHAG - Granollers’s Hospital. Granollers, Spain.

S. Bolivar, Department of Radiology, FHAG - Granollers's Hospital. Granollers, Spain.

M. Cuadrado, Department of Radiology, FHAG - Granollers's Hospital. Granollers, Spain.

J. F. Madureira Cordeiro, Department of Radiology, FHAG - Granollers’s Hospital. Granollers, Spain.

J. R. Torino, Department of Radiology, FHAG - Granollers's Hospital. Granollers, Spain.

X. Pruna, Department of Radiology, FHAG - Granollers's Hospital. Granollers, Spain.
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