



Clival and Paraclival Lesions: A Pictorial Review

Delnaz R. Palsetia¹ Antariksh V. Vijan¹ Foram B. Gala² Amit C. Sahu³ Deepak P. Patkar⁴
Arpita Sahu A.¹

¹ Department of Radiodiagnosis and Imaging, Tata Memorial Hospital, Homi Bhabha National Institute, Mumbai, Maharashtra, India

² Department of Radiodiagnosis and Imaging, Lifescan Imaging Centre & Bai Jerbai Wadia Hospital for Children, Mumbai, Maharashtra, India

³ Department of Interventional Neuro-Radiology, Wockhardt Hospital, Mumbai, Maharashtra, India

⁴ Department of Imaging, Nanavati Superspecialty Hospital, Mumbai, Maharashtra, India

Address for correspondence Arpita Sahu A., MD, Department of Radiodiagnosis and Imaging, Tata Memorial Hospital, Homi Bhabha National Institute, Room no. 119, Main Building, Parel 400012, Mumbai, Maharashtra, India (e-mail: drarpitasahu@gmail.com).

Indian J Radiol Imaging 2023;33:201–217.

Abstract

Keywords

- magnetic resonance imaging
- skull base
- clivus
- abducens nerve
- skull base neoplasms/ complications

The clivus is a midline anatomical structure in the central skull base. It is affected by a wide range of non-neoplastic, benign and malignant pathologies, some of which typically affect the clivus because of its strategic location and embryological origins. Clival lesions may often be asymptomatic with occasional complaints like headache or cranial neuropathy in few. Cross-sectional imaging techniques, namely, computed tomographic scan and magnetic resonance imaging, thus, play a key role in approximating to the final diagnosis and estimating the disease extent. In this article, we highlight the important imaging features of various clival and paraclival pathologies to facilitate effective diagnosis, therapeutic planning, and management.

Introduction

The clivus is the sloping midline bony structure at the skull base formed by the basioccipital and basisphenoid bones that join at the spheno-occipital synchondrosis. Although lesions confined to clivus may not be symptomatic, aggressive lesions may cause cranial nerve symptoms. The abducens nerve runs in close proximity to the clivus, thus diplopia can be a presenting feature.¹

This article describes the various pathologies involving the clivus and paraclival region, including congenital, inflammatory, infective, benign tumors, primary malignant tumors, and metastatic lesions.

Cross-sectional imaging, that is, computed tomographic (CT) scan and magnetic resonance imaging (MRI), are complementary to each other in describing the extent of the lesion and narrowing down the list of diagnoses. CT scan is the modality of choice to look for bony involvement and serves as a guide in evaluating type of intratumoral calcification(matrix), bone destruction or osteosclerosis, and also bone hyperostosis in cases of meningioma. However, the limitation of CT scan is lower soft tissue resolution. On the contrary, MRI provides higher soft tissue resolution, and clival lesions owing to their central location and fatty marrow are best appreciated on sagittal T1-weighted (T1W) nonfat saturated noncontrast sequence.²

article published online
February 11, 2023

DOI <https://doi.org/10.1055/s-0043-1761183>.
ISSN 0971-3026.

© 2023. Indian Radiological Association. All rights reserved.
This is an open access article published by Thieme under the terms of the Creative Commons Attribution-NonDerivative-NonCommercial-License, permitting copying and reproduction so long as the original work is given appropriate credit. Contents may not be used for commercial purposes, or adapted, remixed, transformed or built upon. (<https://creativecommons.org/licenses/by-nc-nd/4.0/>)
Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

Embryology and Anatomy

The term “clivus” is Latin for “slope” and aptly describes this inclined bone of central skull base, sometimes referred to as clivus of Blumenbach.

Anatomic boundaries and relations are as follows (→Fig. 1).

- Superiorly—dorsum sellae; sella & its contents.
- Inferiorly—foramen magnum; jugular tubercle.
- Laterally—petroclival fissure (petro-occipital synchondrosis); petrous temporal bone; petrous internal carotid artery (ICA).
- Anteriorly—sphenoid sinus, nasopharynx.
- Posteriorly—prepontine, premedullary cisterns and contents; brainstem.

The petro-occipital fissure, which begins near cavernous sinus and inferiorly extends upto the jugular foramen, flanks the clivus laterally. The fissure harbors cartilage and dense connective tissue and is the recognized epicenter for many skull base chondrosarcomas.

Neurovascular Relations

The VIth cranial nerve is intimately related to the clivus. After exit from brainstem, the VIth cranial nerve pierces outer layer of dura mater covering the clivus to lie in the interdural Dorello canal. It ascends the clival slope in Dorello canal upto the petrous apex where it passes under the petrosphenoid ligament to enter cavernous sinus. Such a long interdural course makes VIth nerve sensitive to changes of raised intracranial pressure, and VIth nerve palsy resulting from compression against petrosphenoid ligament is a commonly described phenomenon. Additionally, space-occupying clival lesions result in VI nerve palsy by direct mass effect.

The petrous ICA is closely related to the clivus along its lateral margins, with the basilar artery along posterior clival margin in the prepontine cistern. Basilar venous plexus is present on the dural surface of clivus, and may mimic focal lesions if prominent.

Embryologically, clivus is derived mainly from the first and second occipital sclerotomes, with contribution from the proatlas. Clivus is a cartilaginous structure which ossifies from the basisphenoid (post-sphenoid) and basiocciput ossification centers, joined by the spheno-occipital synchondrosis. This synchondrosis fuses by 25 years of age.³ The basi post-sphenoid forms the upper clivus, while the basiocciput contributes to lower clivus. In fetal life, notochord is intimately related to central skull base (particularly the clivus) as its cranial attachment is to the dorsum sellae. In postnatal life, the notochord regresses with few residual notochord remnants along its embryologic path (→Fig. 2). This serves as the embryologic basis to understand various variants of notochordal development (discussed in →Fig. 2).

MR Signal Changes with Age

Since the clivus is a midline structure, it is well visualized on sagittal MR sequences. Bone marrow signal of the clivus depends on the amount of red and yellow marrow and changes with age. It is imperative to be aware of these changes so as not to misinterpret them as pathology. The marrow signal is best assessed on sagittal T1 non-fat-saturated, non-contrast images. As is seen in the rest of the body, the proportion of yellow (fatty) marrow increases with age; hence with age, clivus becomes hyperintense on T1W images. Occasionally, clival marrow heterogeneity may be seen in the form of T1 hypointense foci. Clival signal intensity when isointense or hyperintense relative to pons is usually normal. T1 hypointensity relative to pons / intervertebral discs should be regarded with suspicion. The evolution of marrow signal intensity of the clivus with age is shown in →Fig. 3.^{4,5}

Grading of clival bone marrow signal intensity on T1W images is as follows:

1. Predominantly low signal intensity, occupying more than 50% of the clivus.
2. Low signal intensity portion occupying less than 50% and more than 20% of the clivus.

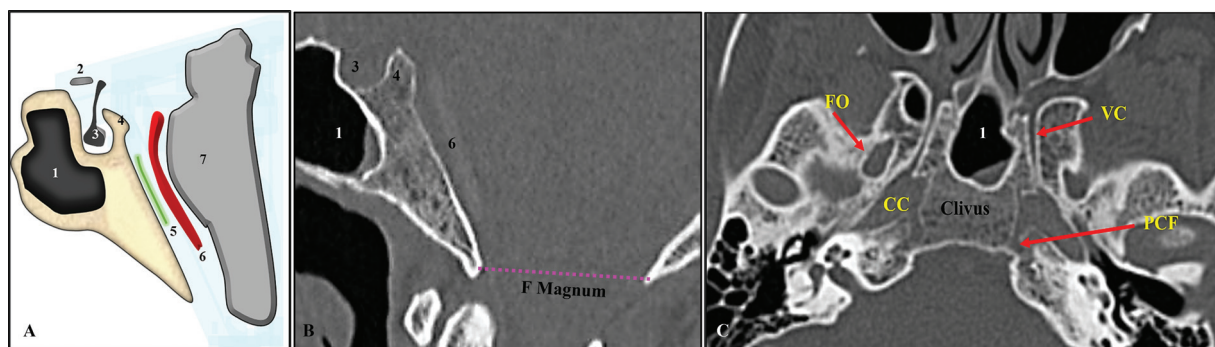


Fig. 1 Graphic illustration. (A) The adult clivus. 1—sphenoid sinus, 2—optic chiasm, 3—pituitary gland in sella turcica, 4—dorsum sellae, 5,6 represent the VI cranial nerve (abducens) and basilar artery, respectively, within the prepontine cistern, 7—pons. Note the fatty marrow within the clivus. Sagittal computed tomographic (CT) image (B) in bone window shows these features on CT. The pink line joins the basion and opisthion marking the foramen magnum. Axial CT image (C) in bone window shows petroclival fissure (PCF)—articulation between petrous temporal bone and basiocciput of clivus. Laterally, the internal carotid artery is seen in carotid canal (CC). Anteriorly, foramen ovale (FO) and Vidian canal (VC) are in proximity.

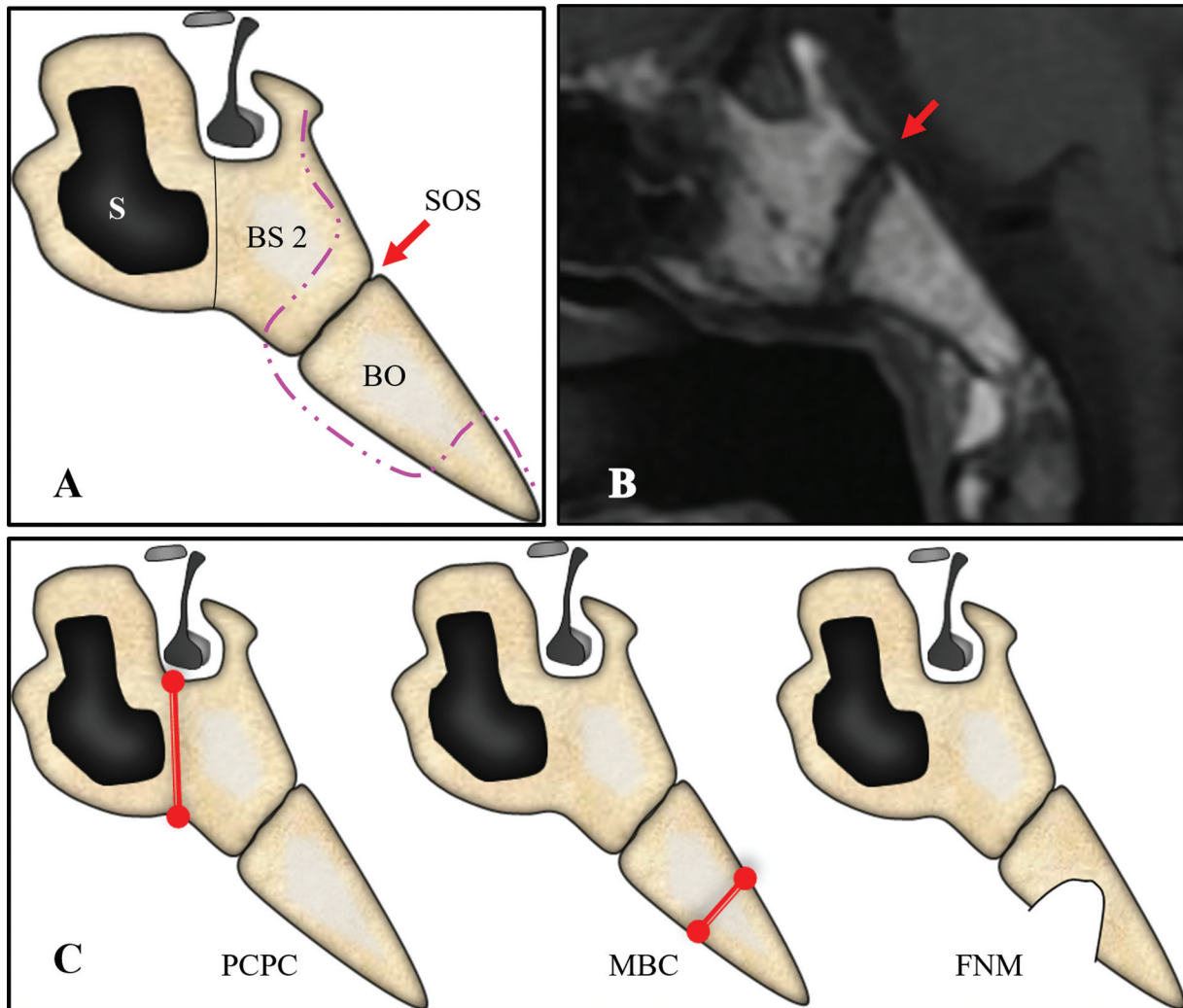


Fig. 2 Graphic illustration (A) showing normal clivus with unfused midline spheno-occipital synchondrosis (SOS) between basi post-sphenoid (BS2) and basiocciput (BO). Pink dotted line represents course of notochord remnants. Sagittal T1-weighted magnetic resonance imaging of a 6-year-old boy (B) demonstrates these features; red arrow—spheno-occipital synchondrosis. Graphic illustrations in image (C) demonstrate characteristic locations of clival congenital/embryological variants. Persistent craniopharyngeal canal (PCPC), is a canal lying anterior to the SOS, connecting anterior floor of sella to nasopharynx. Junction between pre-sphenoid part (S) and basi post-sphenoid marks the site of PCPC (compare with 2A). Median basal canal/canalis medianus basalis (MBC) lies posterior to the SOS and is in the expected location of notochord remnant (see 2A). —Fossa navicularis magna (FNM) is characterized by a divot or saucer shaped defect in the basiocciput portion of the clivus.

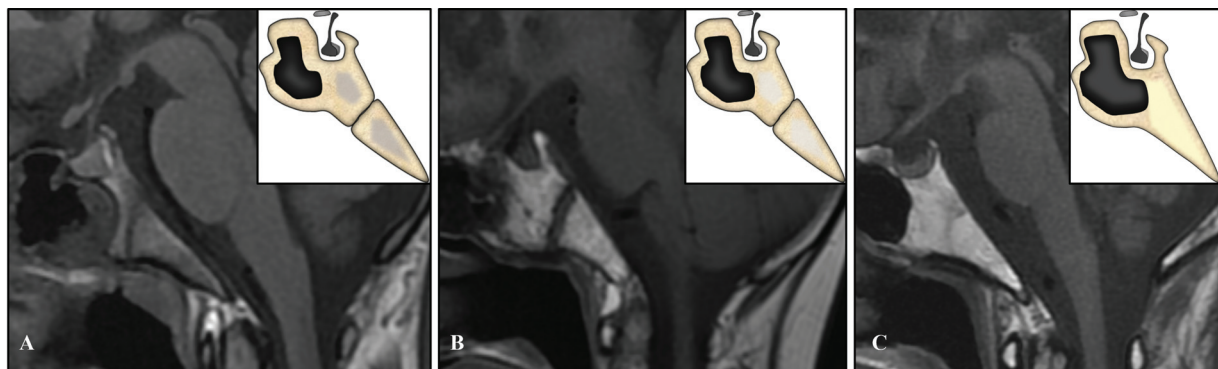
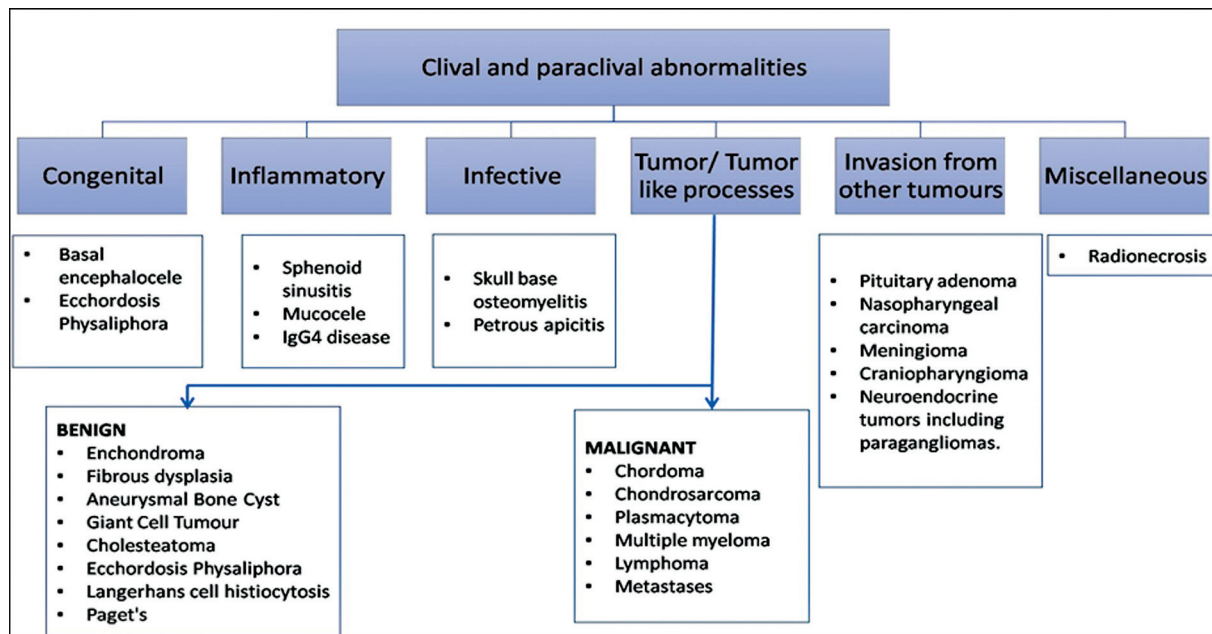


Fig. 3 Evolution of clival marrow signal intensity with age. (A) A 6-year-old child, with unfused spheno-occipital synchondrosis with T1 isointensity, suggesting red marrow. (B) A 14-year-old child with unfused spheno-occipital synchondrosis with homogeneously T1 hyperintense (fatty) marrow. (C) Adult clivus with fused spheno-occipital synchondrosis and homogeneously fatty marrow. Note how clival marrow is not darker than the pons on T1-weighted magnetic resonance imaging in any case.

Table 1 Classification of clival and paraclival abnormalities

3. Predominantly high signal intensity, with low signal portion being less than 20% of the clivus. Normal clivus may or may not show mild enhancement with greater prevalence of enhancement in grade 1 clivus signal intensity.^{4,5}

Clival and Paraclival Pathologies (→ Tables 1-3)

Congenital

Basal encephaloceles are rare midline bony defects of skull base, through which there is herniation of the brain parenchyma and meninges. They can be associated with other congenital anomalies like cleft lip, cleft palate, and hypertelorism.

They are classified anatomically depending on the site of defect as:

- 1) Transsphenoidal or
- 2) Transethmoidal.

They may also be classified as:

- 1) Sphenopharyngeal where there is defect in sphenoid bone and encephalocele herniates into nasopharynx and
- 2) Spheno-orbital where encephalocele herniates through the superior orbital fissure to lie in retrobulbar region resulting in unilateral exophthalmos.

On imaging, transsphenoidal encephalocele is characterized by a vertical midline skull base defect with protrusion of hernia sac into nasopharynx and at times varying degree of naso-oropharyngeal lumen compromise can be seen leading to respiratory and feeding difficulties. MRI will show the

contents of the sac as well as associated abnormalities^{6,7} (→ Fig. 4).

Inflammatory

Mucoceles of sphenoid sinus can erode intracranially involving the clivus, causing neurological symptoms, most commonly diplopia.⁸ Primary clival mucoceles are extremely

Table 2 Categorization of various clival and para clival pathologies

Pathologies involving the clivus directly	Paraclival pathologies
Basal encephalocele	<ul style="list-style-type: none"> • Skull base osteomyelitis • Petrous apicitis
Benign: <ul style="list-style-type: none"> • Enchondroma • Fibrous dysplasia • Aneurysmal bone cyst • Giant cell tumor • Cholesteatoma • Ecchordosis physaliphora • Paget disease 	Head and neck pathologies extending to clivus: <ul style="list-style-type: none"> • Nasopharyngeal carcinoma • Sphenoid sinusitis • Mucocele • IgG4 disease
Malignant: <ul style="list-style-type: none"> • Chordoma • Chondrosarcoma • Plasmacytoma • Langerhans cell histiocytosis • Multiple myeloma • Lymphoma • Metastases 	Intracranial pathologies invading the clivus: <ul style="list-style-type: none"> • Pituitary adenoma • Meningioma • Craniopharyngioma • Neuroendocrine tumors including paragangliomas
Miscellaneous: Radionecrosis	

Table 3 Age-wise categorization of clival and para clival pathologies

Pediatric	Adult	No specific predilection
Basal encephalocele	Paget's disease	Infective and inflammatory processes
Adamantinomatous craniopharyngioma	Chondrosarcoma	Fibrous dysplasia (but < 30 years)
Langerhans cell histiocytosis	Giant cell tumor	Neuroendocrine tumors
Primary skull base lymphoma	Chordoma	Enchondroma
Aneurysmal bone cyst	Plasmacytoma, multiple myeloma, metastases	Radionecrosis
	Meningioma, pituitary adenoma, papillary craniopharyngioma, nasopharyngeal carcinoma	

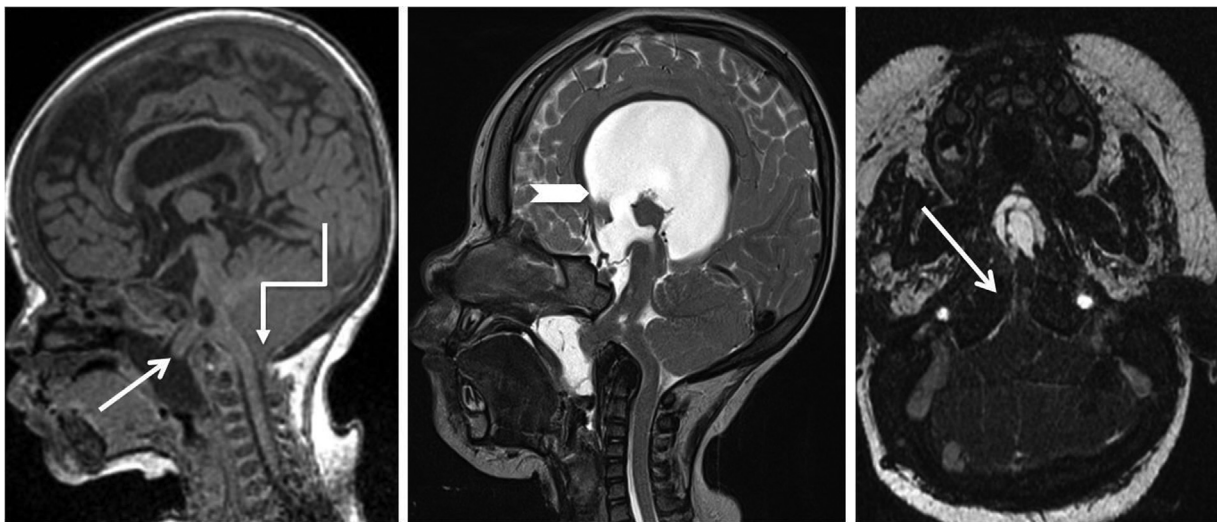


Fig. 4 Basal encephalocele. Sagittal T1-weighted (T1W), T2W, and axial T2W images in an 18-year-old girl shows midline clival defect (arrow) with herniation of midbrain into the nasopharynx. (arrow). Note small posterior fossa with peg like tonsils herniating into upper cervical canal (elbow arrow). Effacement of aqueduct results in supratentorial hydrocephalus (arrowhead).

rare. In some individuals, the clivus is pneumatized in the course of development and may communicate with the sphenoid sinus through an ostium. Sphenoid sinusitis (► **Fig. 5**) in such cases causes obstruction of the clival opening, hence causing retention of the secretions by the clival mucosa. On imaging, mucocèles are locally expansile lesions with thinning and erosions of sinus walls.⁹ Signal characteristic on MRI depends on the content and appears homogeneously hyperintense on T1- and T2-weighted (T2W) sequences with no post-contrast enhancement when rich in protein. Sphenoid sinus mucocèle can be isolated or may be secondary to malignant lesions.^{10,11}

Infective

Skull base osteomyelitis (SBO) is a serious, life-threatening condition seen most commonly in elderly diabetics or immunocompromised patients. The two most common bacterial causes are *Staphylococcus aureus* and *Pseudomonas aeruginosa*. *Aspergillus fumigatus* is the most common

fungal cause. SBO usually presents with severe otalgia, fever, aural fullness, and foul purulent otorrhea. In atypical central skull base osteomyelitis or clival osteomyelitis, there are no otologic symptoms or external auditory canal pathology that are otherwise seen in SBO.¹²

CT will show soft tissue masses and collection associated with bony erosion at the central skull base (► **Fig. 6–I, II**). Typical MRI findings (► **Fig. 6–I, II**) include hypointense signal on T1W and hyperintense signal in the clivus on T2W images with enhancement on post-contrast imaging. However, fungal infections appear classically hypointense on T2W images. There may be associated dural enhancement along the skull base. An important differential is malignancy, which would be T2 intermediate and show solid enhancement, unlike osteomyelitis. Erythrocyte sedimentation rate could aid in differentiating the two entities, as it is not elevated in malignancy.¹³

Tuberculous osteitis (► **Fig. 7**) of the clivus is rare.¹⁴ Presence of tubercular infection elsewhere in the body, for example, lungs, may help in diagnosing the skull base

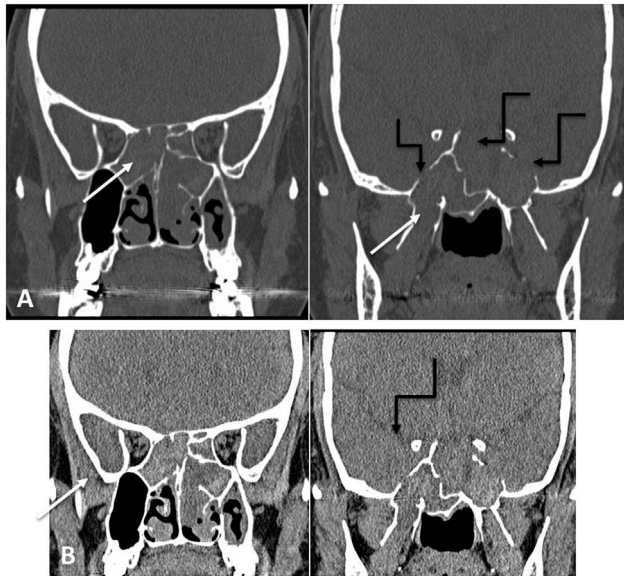


Fig. 5 Allergic fungal sinusitis: Noncontrast computed tomographic images of paranasal sinuses in coronal planes show polypoidal mucosal thickening with hyperdense contents causing opacification of sphenoid and ethmoid sinuses (arrow). Erosion of sinus walls is also seen (elbow arrow).

involvement. Clival destruction can be seen on plain radiographs but is better delineated on CT. Soft tissue will be better seen on MRI.¹⁵

Petrous apicitis is caused by medial extension of acute otitis media into a pneumatized petrous apex. Patients with petrous apicitis present with an acute febrile illness and the classic Gradenigo triad (suppurative otitis media, pain in distribution of trigeminal nerve and abducens nerve palsy). Petrous apicitis can cause meningitis, cerebral abscess, and dural venous sinus thrombosis.¹⁶ Conventional radiograph may show bone erosion and asymmetric clouding of the petrous tip. MRI features of petrous apicitis are hyperinten-

sity on T2W images, hypointensity on T1W images with enhancement. There will be enhancement of the adjacent dura mater if there is associated meningitis. Ring enhancement and diffusion restriction suggest abscess formation. On CT, petrous apicitis seen as opacification of petrous air cells in the early stage of the disease and bone destruction in later stages.¹⁷

Benign Tumors

Enchondroma is a benign chondroid tumor that is common in the phalanges, metacarpals, and metatarsals. Intracranial enchondromas or chondromas are rare benign cartilaginous tumors. They mainly involve the skull base. In the base of the skull, these lesions usually arise from cartilaginous rests, especially around the basal synchondroses. They are usually solitary, but can also be seen as a part of Ollier's disease¹⁸ and Maffucci syndrome. CT may show a hyperdense lesion involving the clivus with multiple areas of calcification. On MRI, enchondromas appear as well-circumscribed lesions without accompanying edema with heterogeneous intermediate- to low-signal intensity on T1W images and mixed intensity on T2W images (→**Fig. 8**). Complications are transformation to chondrosarcoma and increased risk of glial tumors. Bone destruction and soft tissue extension indicate malignant transformation.¹⁹

Clival involvement by fibrous dysplasia (FD) is extremely rare. FD is a developmental disorder caused by abnormal proliferation of fibroblasts resulting in replacement of normal cancellous bone by structurally weak, immature osseous tissue. Most of the clival FDs are monostotic. CT is the imaging tool of choice for the evaluation of FD. The FD involving the cranial base tends to be a sclerotic variety, where the inner table gets thickened. Ground-glass density, ballooning, and expansion of the affected bone and thinning of the cortex are the hallmarks of FD. MRI reveals

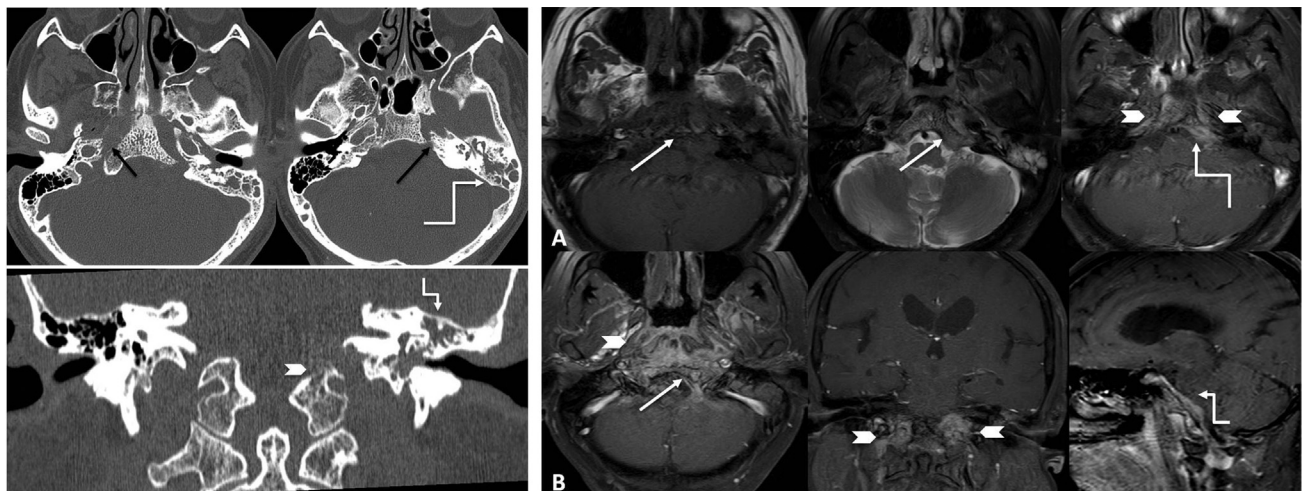


Fig. 6 (I) Skull base osteomyelitis: Axial (A) T1-weighted (T1W), T2W, and T1 + C fat-saturated images in a 62-year-old diabetic male with headache show abnormal signal involving clivus (arrow), associated with enhancing soft tissue in both petroclival regions and in prevertebral region (arrowhead). Note dural enhancement on left side of clivus (elbow arrow) and left otomastoiditis. (II) Skull base osteomyelitis: CT images of same patient as in 4(I) showing erosion of clivus (arrow) with widening of petroclival joint, also there is erosion of left hypoglossal canal (arrowhead) and left otomastoiditis (elbow arrow).



Fig. 7 Tuberculous osteomyelitis: Sagittal (A) and axial (B) T1-weighted (T1W), T2W, and post-contrast T1 fat-saturated images in a 23-year-old male with restricted neck movements shows abnormal signal involving clivus, atlas, and odontoid process associated with large peripherally enhancing collection/abscess in prevertebral region (arrow). Note cord compression at cervicomedullary junction due to soft tissue (elbow arrow).

hypointense lesion on T1W images and variable (usually low) signal intensity lesion on T2W images with heterogeneous post-contrast enhancement (►Fig. 9).²⁰

Aneurysmal bone cysts (ABCs) are well-differentiated benign osteolytic tumors, usually seen below 20 years of age. Common location is metaphysis of long bones; however, the clivus can be involved as well (about 2–6% of all ABCs). Plain radiograph usually shows a complex cystic lesion. Erosion and thinning of the cortex will be better appreciated on CT. MR shows classical multiple fluid levels and multiple internal septations (►Fig. 10). Fluid levels within ABCs are indicative of hemorrhage with sedimentation. Internal septae are better delineated on post-contrast imaging. An associated low signal intensity rim may be seen around the lesion due to thickened but intact periosteum.^{21,22}

Giant cell tumors (GCTs) are benign aggressive lesions that typically arise in the epiphysis of long bones. Although cranial involvement is rare (less than 1% of all GCTs), GCTs preferentially involve the sphenoid and temporal bones. Clival GCTs are rare and may present with symptoms of

raised intracranial pressures, optic pathway compression, or cranial nerve palsy (most commonly 6th nerve).²³ On CT, an expansile enhancing mass can be seen causing erosion of the clivus. Imaging characteristics on MR include a characteristic T2 hypointense soft tissue lesion showing homogenous post-contrast enhancement (►Fig. 11).²⁴

Echordosis physaliphora (EP) is a notochordal remnant appearing at the dorsal wall of the clivus, in midline closer to foramen magnum than pituitary fossa.²⁵ A stalk like clival connection may be visualized on high-resolution heavily T2W sequence like CISS/FIESTA. It is benign and has to be differentiated from chordoma. EP is usually intradural, while chordomas mostly arise extradurally and lead to expansile bone destruction which is well appreciated on CT scan. Although chordomas are generally associated with symptoms, an EP is usually asymptomatic; however, there can be exceptions. CT scan shows well-defined hypodense lesion in clivus with scalloped, sclerotic margins. On MRI, EP is hypointense on T1 and hyperintense on T2W images with no enhancement. It is a “touch me not” lesion (►Fig. 12).²⁶

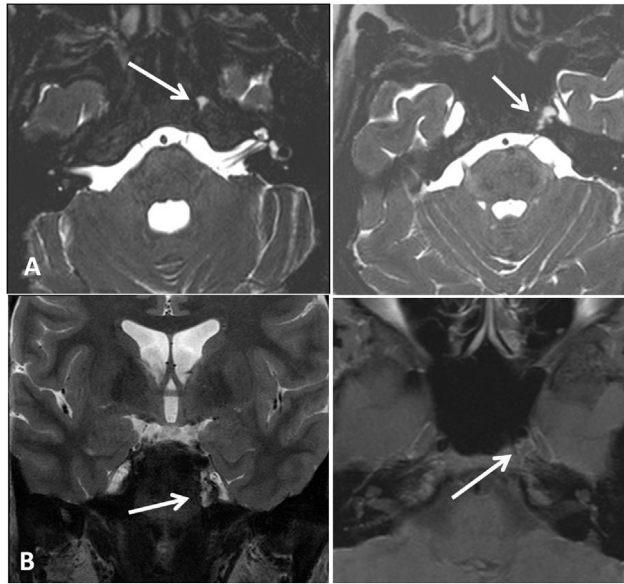


Fig. 8 Chondroid lesion: Axial constructive interference in steady state (CISS) images (A) and coronal T2W and axial T1-post-contrast images (B) in a 23-year-old male with left side painless diplopia show a lobulated extra-axial lesion arising from petrosphenoid synchondrosis, compressing left 6th cranial nerve. The lesion is hyperintense on T2-weighted images and shows heterogeneous (peripheral septal) post-contrast enhancement (arrow), classic of a chondroid lesion.

Paget disease (osteitis deformans) is a chronic skeletal disorder characterized by abnormal and excessive remodeling of bone. It demonstrates three phases:

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

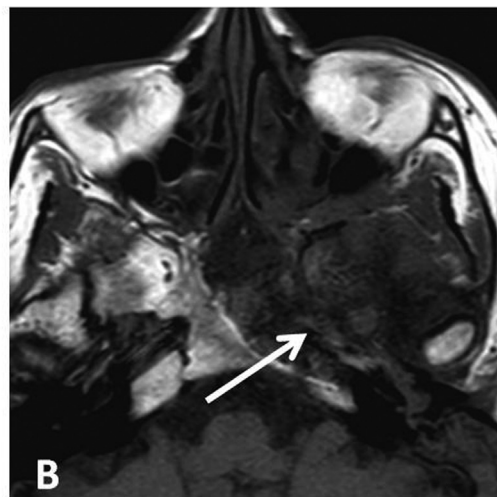
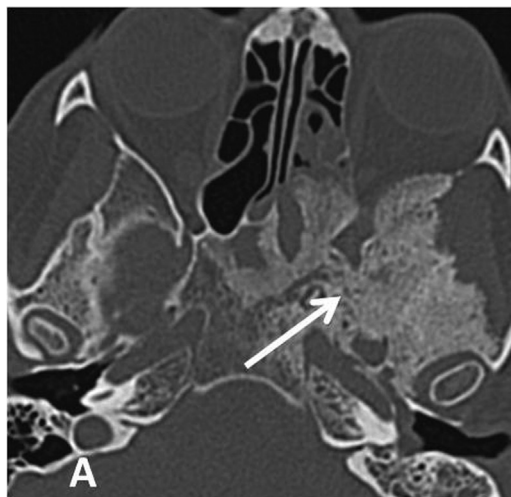


Fig. 9 Fibrous dysplasia. Axial computed tomographic image in bone window (A) shows bony expansion with ground-glass matrix and sclerotic change involving the left half of clivus, with foraminal involvement. Corresponding axial T1-weighted magnetic resonance imaging (B) shows low T1 signal in the expanded bone (arrows), consistent with skull base fibrous dysplasia.

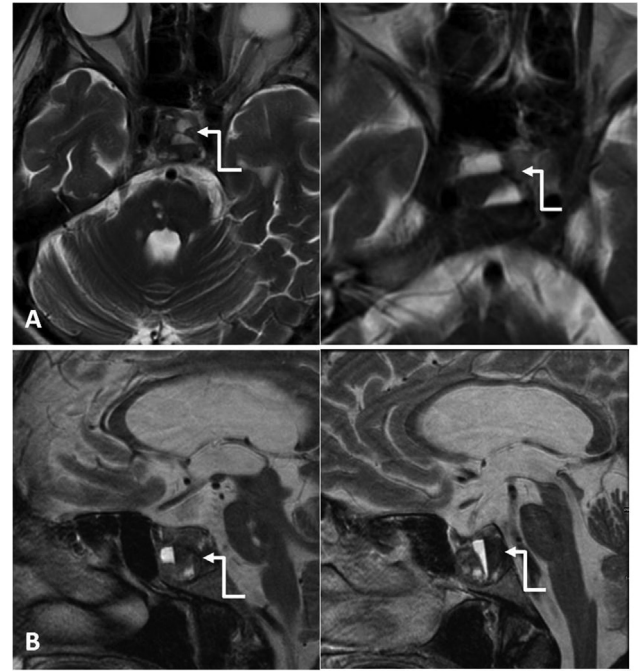


Fig. 10 Aneurysmal bone cyst: Axial T2-weighted (T2W) images (A) and sagittal T2W (B) magnetic resonance imaging showing cystic lesion with fluid-fluid levels (elbow arrows) involving the clivus. Fine-needle aspiration cytology revealed hemorrhagic aspirate, and a diagnosis of aneurysmal bone cyst was established on pathognomonic imaging findings.

When skull base is involved, there is associated platybasia, basilar invagination, and verteobasilar insufficiency. MRI features depend on the phase of the disease: hyperintense on T2, hypointense on T1 in the lytic phase, hyperintense on both T1W and T2W sequences in mixed phase and hypointense on both T1W and T2W sequences in blastic phase. Bone thickening and sclerosis will be seen on CT in the blastic phase.^{20,27}

Cholesteatoma refers to epithelium lined cysts filled with keratinized desquamated debris, typically used in

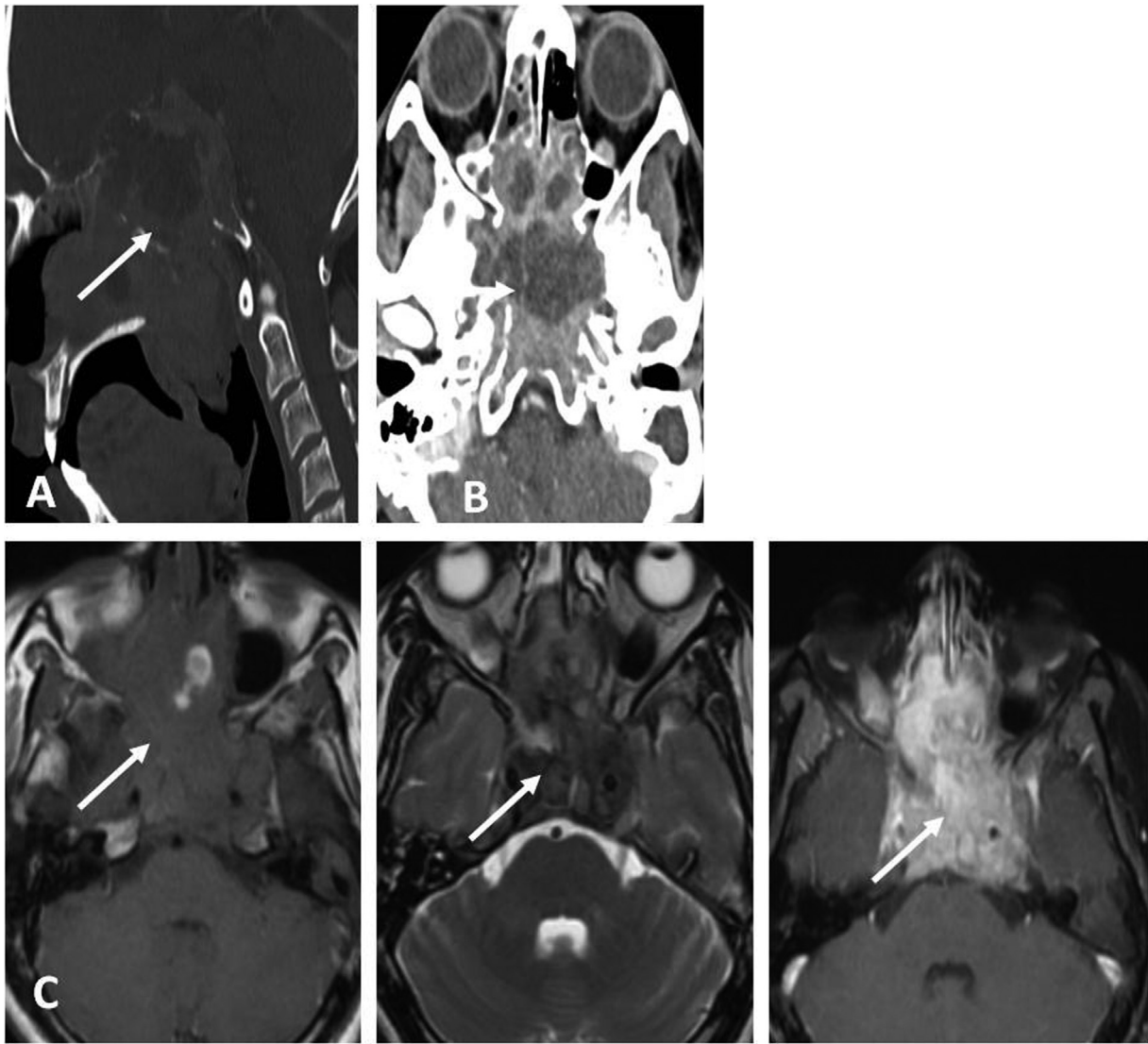


Fig. 11 Giant cell tumor. Sagittal bone algorithm computed tomography (CT) reformatted (A) and axial post-contrast CT (B) images showing an expansile lytic lesion (arrows) with no matrix involving the clivus. Axial T1-weighted (T1W), T2W, and post-contrast T1W (C) images show a large lobulated T2 intermediate to hypointense mass (classic T2 signal of giant cell tumor [GCT]) showing enhancement and bony expansion. Histopathology revealed GCT.

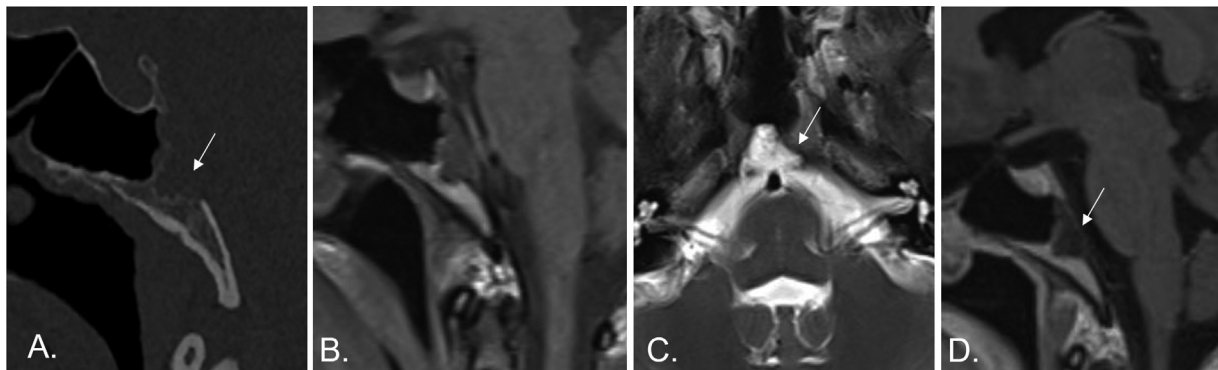


Fig. 12 Echordosis physaliphora: Sagittal computed tomographic (CT) image in bone window (A), sagittal T1 noncontrast (B), sagittal T2-weighted imaging (T2WI) (C) and sagittal T1 + C images (D) in a 63-year-old male show an incidental finding of a well-defined lesion (arrows) in clivus which is hypointense on T1W and homogeneously hyperintense on T2W images, without any post-contrast enhancement (D). These features of well-defined nonenhancing T2 bright lesion were classic for echordosis physaliphora and multidisciplinary tumor board decision to “wait and watch” was taken.

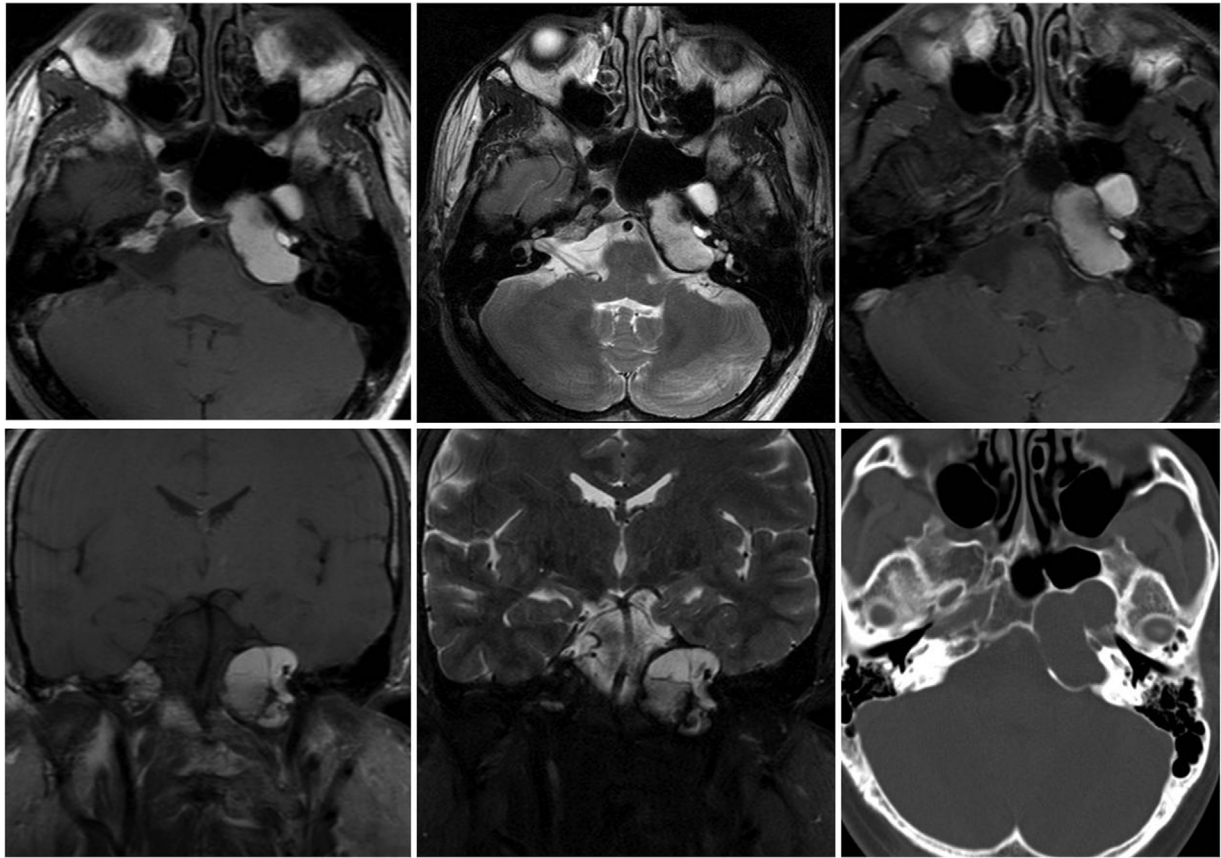


Fig. 13 Congenital cholesteatoma. Axial T1-weighted (T1W), T2W, and post-contrast (A) and coronal T1W, T2W images (B) in a 33-year-old male showing well-defined expansile cystic lesion involving left petrous apex and extending into clivus. It is causing thinning of petrous bone and indents on left cerebellopontine cistern and internal auditory canal. The lesion is hyperintense on T1W and T2W images with no enhancement. Imaging differential diagnosis is cholesterol granuloma.

association with middle ear disease.²⁸ Extraotic locations include the posterior fossa at the cerebellopontine angle in the intradural area. Extradural tumors are most often found in the petrous bone and very rarely in the clivus. Other than erosion of the clivus on CT, cholesteatomas are usually hypo to isointense on T1W images and hyperintense on T2W MRIs with no enhancement (→**Fig. 13**). They appear to have high signal on DWI, which is its characteristic feature. Due to a high propensity to recur, total surgical excision is the preferred method of management.

Langerhans cell histiocytosis a rare but possible cause of a clival mass in the pediatric population. It typically shows destructive, lytic punched out bone lesions. The lesions are hyperintense on T2 and hypointense on T1W images with homogeneously or heterogeneously enhancing destructive soft tissue mass and variable local extension. Diffusion restriction may be seen due to the increased cellularity of the lesion. Once clival Langerhans Cell Histiocytosis (LCH) is detected, synchronous lytic lesions in other areas of the skeleton should be looked for.^{29,30}

Primary Malignant Tumors

Chordomas are malignant tumors arising from embryonic remnants of the primitive notochord, usually seen in the age group of 40 to 70 years, but can also occur in the age group of

20 to 40 years. They are slow growing tumors. They can arise in the clivus from the basisphenoid, basiocciput or the sphenococcipital synchondrosis. Histopathologically, they can be classified as typical and chondroid. On CT, chordomas are seen as a centrally located, expansile soft-tissue mass arising from the clivus with associated extensive bone destruction.³¹ The chondroid variant is more likely to demonstrate intratumoral calcifications. There is moderate to marked enhancement. Low-attenuation areas sometimes seen within the soft-tissue mass represent the myxoid and gelatinous material seen at gross examination. On MRI, they are hypointense on T1 and very hyperintense on T2W images, which is a characteristic finding. It shows moderate to marked heterogeneous honeycomb pattern of enhancement. The mucinous component will show high signal intensity on T1W images. Chondroid chordomas may not be as bright as typical chordomas on T2W MR images, because the gelatinous matrix is replaced by cartilaginous component (→**Figs. 14 and 15**). This is important for prognosis due to the better survival of patients with chondroid chordoma. MRI also shows if there is displacement or partial encasement of intracranial arteries. Thumb sign or thumbing of the pons is seen in midline sagittal images as indentation of the pons.³²

Skull base chondrosarcoma (→**Fig. 16**) arises from the cartilage or bones with endochondral ossification and

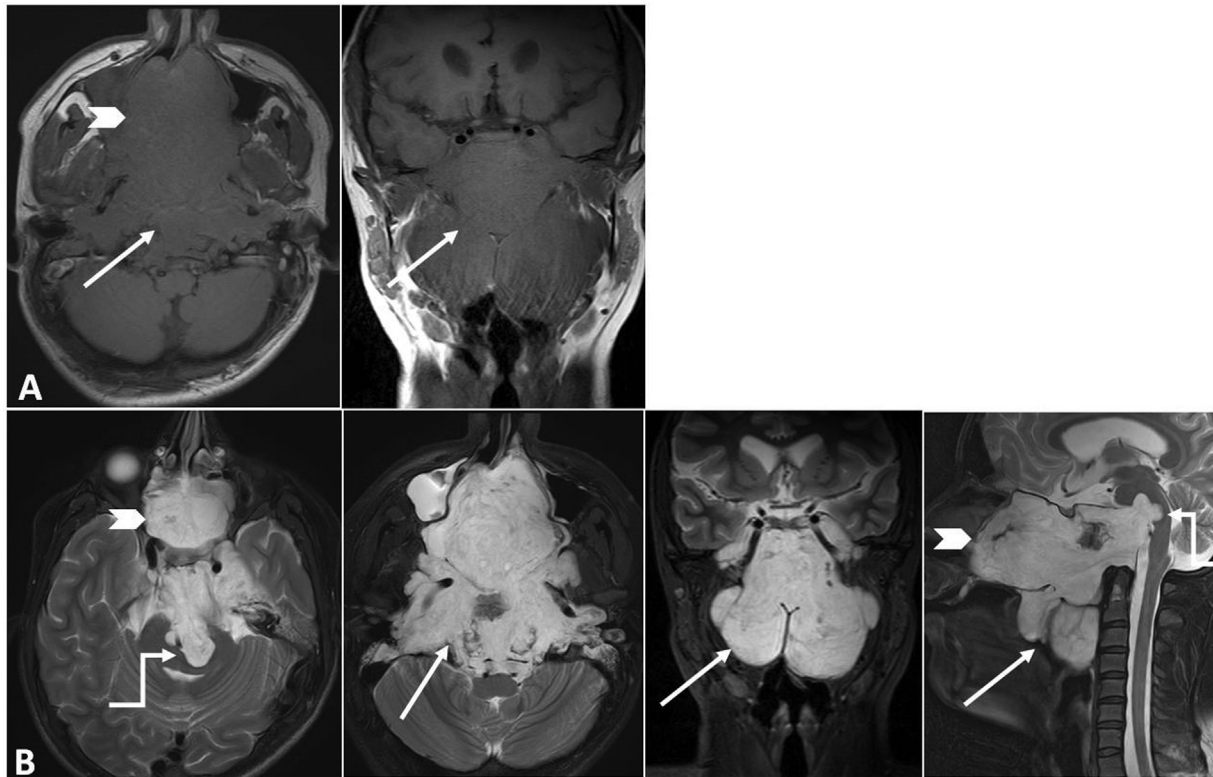


Fig. 14 Clival chordoma. Axial and coronal T1-weighted (T1W) (A); axial, coronal, and sagittal T2W (B) images in a 16-year-old boy with complaints of nasal obstruction showing a large midline intensely T2 hyperintense clival lesion (arrow), with posterior projection indenting the pons, so-called thumb sign (elbow arrow).



Fig. 15 Clival chordoma. Axial T2-weighted (T2W) (A) and sagittal T2W and post-contrast T1W (B) images showing an enhancing T2 heterogeneously hyperintense expansile lesion (arrow) with hypointense thin peripheral sclerotic rim (arrowhead). Histopathology revealed chordoma.

primitive mesenchymal cells of the meninges. It can occur at all ages from the teens into the 90s. Skull base chondrosarcoma most commonly occurs along the petro-occipital fissure. They are usually well-differentiated, grade 1 chondrosarcomas. It is seen as a well-defined osteolytic lesion with characteristic ring and arc calcifications on CT. It is iso to hypointense on noncontrast T1W and hyperintense on T2W MR images with intense heterogeneous enhancement. In contrast with chordoma, chondrosarcomas are off midline.³³ The chondroid matrix may also help to differentiate chondrosarcoma from classical chordoma.

Plasma cell tumors affecting the skull base are generally a part of underlying multiple myeloma (MM). Plasmacytomas are monoclonal proliferations of plasma cells that may arise within soft tissue or bone. Solitary plasmacytoma of the skull base is rare and can progress to MM rapidly.³⁴ Solitary clival plasmacytomas are asymptomatic until symptoms related to compression of cranial nerves, especially sixth cranial nerve and headaches develop.³⁵ Positron emission tomography with fluorodeoxyglucose can aid in the detection of other sites of medullary and extramedullary disease. On MRI, intracranial plasmacytomas are iso- to hyperintense on T1W images and iso- to hypointense on T2W images, differentiating them from other tumors. They show post-contrast enhancement (►Fig. 17). On CT scan, the lesions are osseodestructive and avidly enhance.

MM represents malignant proliferation of monoclonal plasma cells. Intracranial lesions of MM commonly involve the cranial vault, the skull base and the orbit, but the clivus is

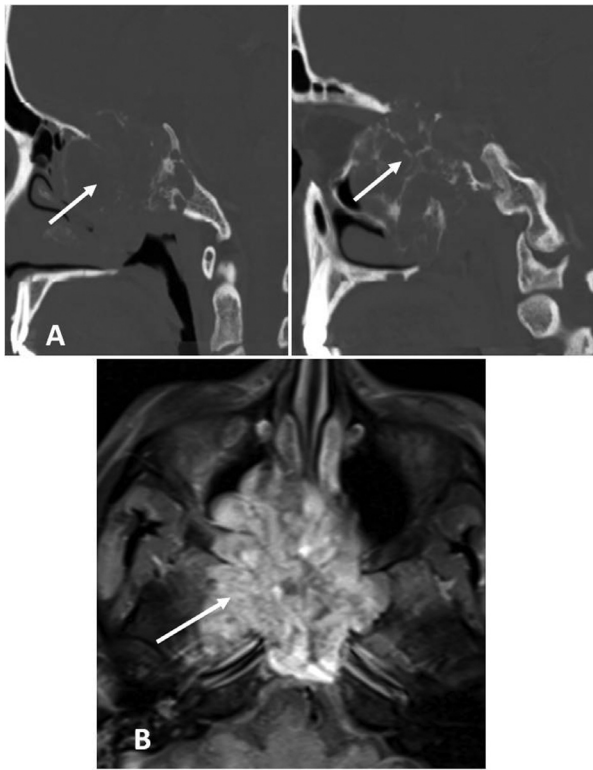


Fig. 16 Clival chondrosarcoma. Sagittal reformatted bone window computed tomographic (CT) image (A) and axial post-contrast magnetic resonance imaging T1-weighted (B) images in same patient showing a large expansile lobulated mass (arrows) in the basiphoid region, showing peripheral lobular enhancement. The CT cuts shows the characteristic ring and arcs calcifications. Histopathology revealed chondrosarcoma.

less commonly affected. Imaging features are similar to that of solitary clival plasmacytoma.³⁶

Primary skull base lymphoma is a rare form of extranodal lymphoma. Diffuse large B-cell lymphoma is the most common subtype. Lymphoma of central skull base is another uncommon tumor in pediatric patients. It is usually a hyperdense soft tissue mass on plain CT. On MRI, it presents as an isointense or hypointense mass on both T1W and T2W images with homogeneous enhancement. It typically shows restricted diffusion owing to high cellularity. The presence of enlarged lymph nodes in the neck would further favor lymphoma.³⁷

Metastases

Clival metastases are extremely rare. Clival metastases can occur in the context of a known history of a distant malignancy or they may be the presenting symptom of a distant malignancy. The primary tumor can be intra-abdominal, thoracic, pelvic or in the head and neck region. They are usually destructive bone lesions with isointense or hypointense signal on T1W images, hyperintense on T2W imaging and enhancement with gadolinium. They can be hyperintense on T1W images, resembling plasmacytomas (►Fig. 18).³⁸

Intracranial Tumors Invading Clivus

Invasive Pituitary adenoma: These are tumors arising from anterior lobe of pituitary gland and invading the clivus/middle skull base (►Figs. 19 and 20). The lesions may show intratumoral cysts/ hemorrhage, also known as

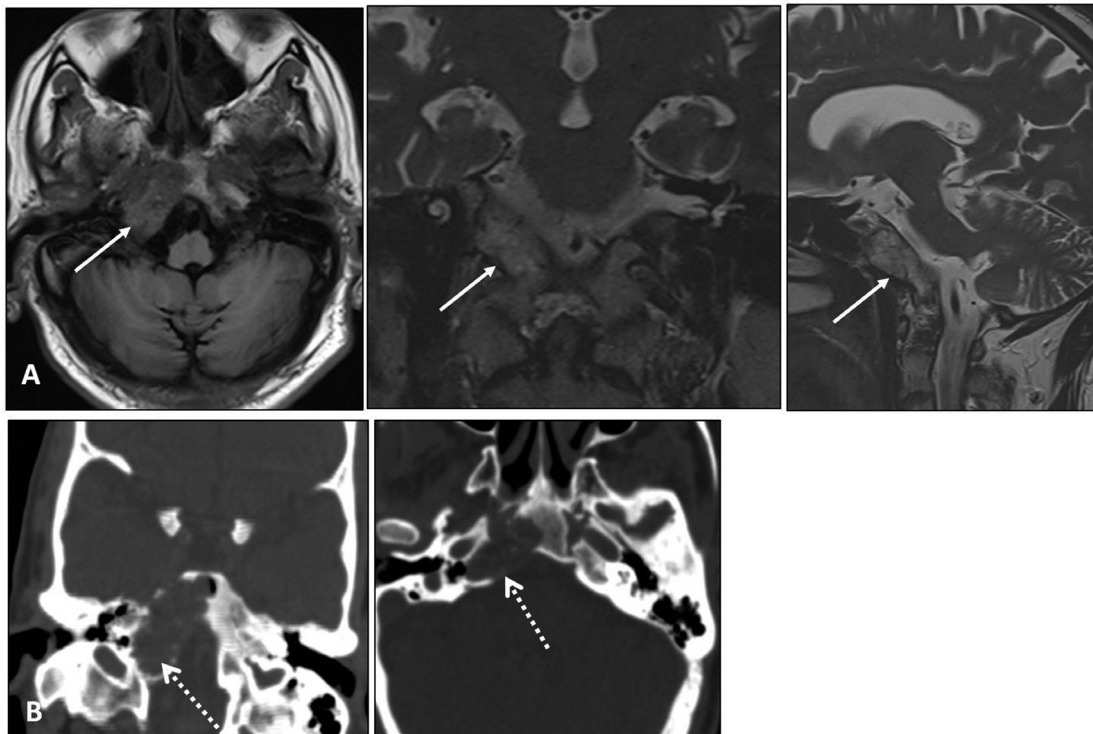


Fig. 17 Clival plasmacytoma. Axial T1W, coronal and sagittal T2W MR (A) and CT images (B) in a 54Y/M show abnormal marrow signal involving right half of the clivus and petrous apex (arrows). On CT scan, it appears as expansile lytic lesion (dotted arrows). Histopathology revealed Plasmacytoma.

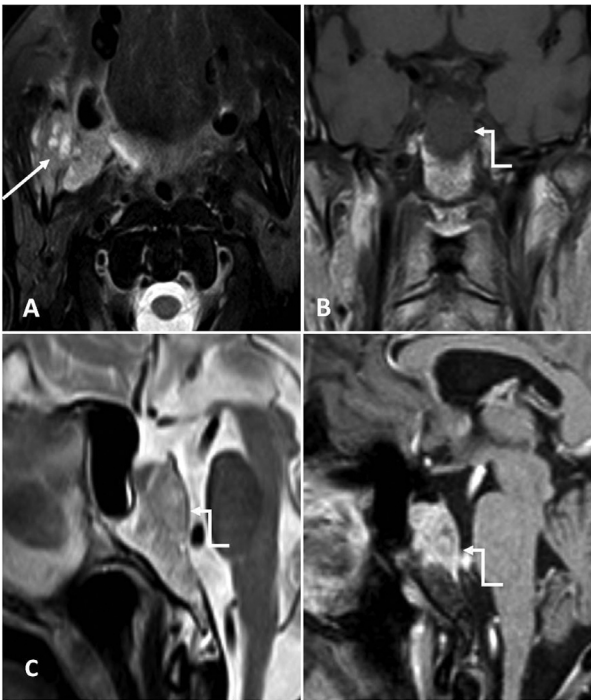


Fig. 18 Clival metastasis. Axial short-tau inversion recovery magnetic resonance (STIR MR) image (A) showing primary mass involving the right buccal mucosa (arrow). Coronal T1-weighted (T1W) (B), sagittal T2W, and T1 post-contrast MR images (C) show an expansile lesion in basisphenoid, which is T1 hypointense, T2 heterogeneously hyperintense showing intense enhancement (elbow arrows).

pituitary apoplexy. This is a tumor of older adults and typically presents with visual fields defects or symptoms related to hormonal activity. Normal pituitary gland will not be seen separately.³⁹

Nasopharyngeal carcinoma, when large, can invade the clivus (► **Fig. 21**). Involvement of the clivus is a bad prognostic factor in patients with nasopharyngeal carcinoma. Patients usually present between 40 and 60 years of age. It is more common in Asian population. Tumors usually arise from the fossa of Rosenmüller and shows mild enhancement.

Meningiomas are the most common benign intracranial neoplasms in adults. Since they are slow growing, they are detected late when they cause symptoms which in case of clival invasion would be cranial nerve palsies. On non-contrast enhanced CT, they are hyperdense and show homogeneous enhancement on post-contrast imaging. They are hypo to isointense on T1 and T2W sequences with intense homogenous enhancement with a dural tail (► **Fig. 22**).

Craniopharyngioma is a suprasellar mass arising from Rathke's pouch. It is of two types,

- 1) Adamantinomatous, which is predominantly cystic and seen in childhood; and
- 2) Papillary, which is a solid mass seen in adults.

Neuroendocrine tumors and paragangliomas are few other lesions affecting the clivus (► **Figs. 23** and **24**). Glomus jugulare is a paraganglioma arising in the jugular fossa. On CT, a characteristic "moth-eaten" pattern of destruction of the temporal bone is seen. The tumor shows intense enhancement. It appears T1 hypointense, T2 hyperintense with classical "salt and pepper" pattern on both of these MR sequences. The "pepper" component is due to areas of signal void, while the "salt" component is caused by slow flow or subacute hemorrhage giving rise to hyperintense foci. This appearance is seen in tumors larger than 1 cm.⁴⁰ Conventional angiography will show intense tumor blush. Indium-111 labeled octreotide scan will detect this tumor due to the presence of somatostatin receptors.

Miscellaneous

Osteoradionecrosis (ORN) is a known complication of radiation therapy for head and neck malignancies, and it involves the destruction of bony structures. ORN of the skull base most commonly involves the sphenoid bone, followed by the clivus. It is difficult to differentiate tumor recurrence from ORN or the two pathologic changes exist at the same time, therefore histological correlation is important (► **Table 4**).⁴¹

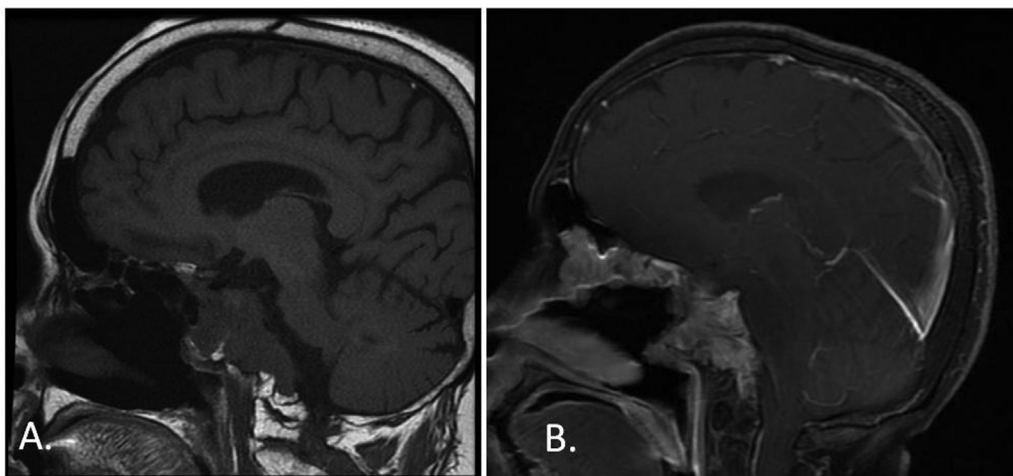


Fig. 19 Pituitary adenoma: Sagittal (A) and coronal (B) images in a 26-year-old female with elevated prolactin levels shows a pituitary mass (arrow) extending into left cavernous sinus and also eroding sella turcica and extending upto clivus histopathology revealed pituitary adenoma.

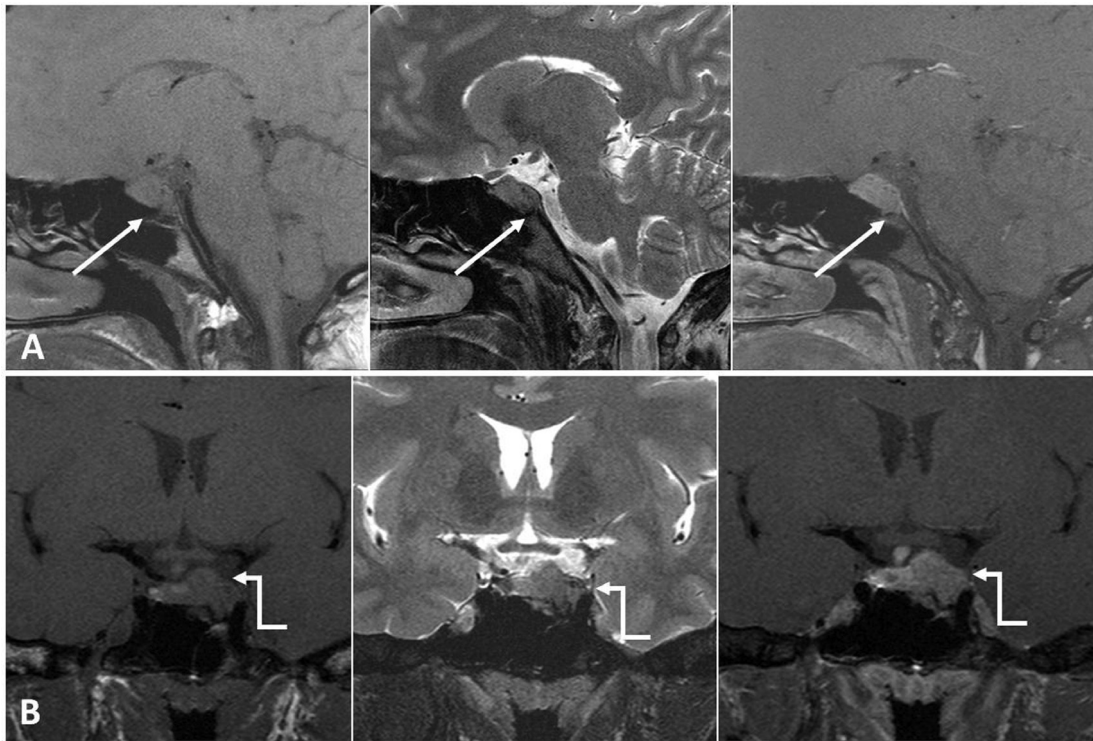


Fig. 20 Invasive pituitary adenoma. T1 and post-contrast images (A, B) of a partially resected pituitary macroadenoma with fat packing, now presenting as a mass in dorsum sellae and invading the entire clivus. The lesion shows homogeneous post-contrast enhancement. Histopathology revealed pituitary adenoma (arrow).

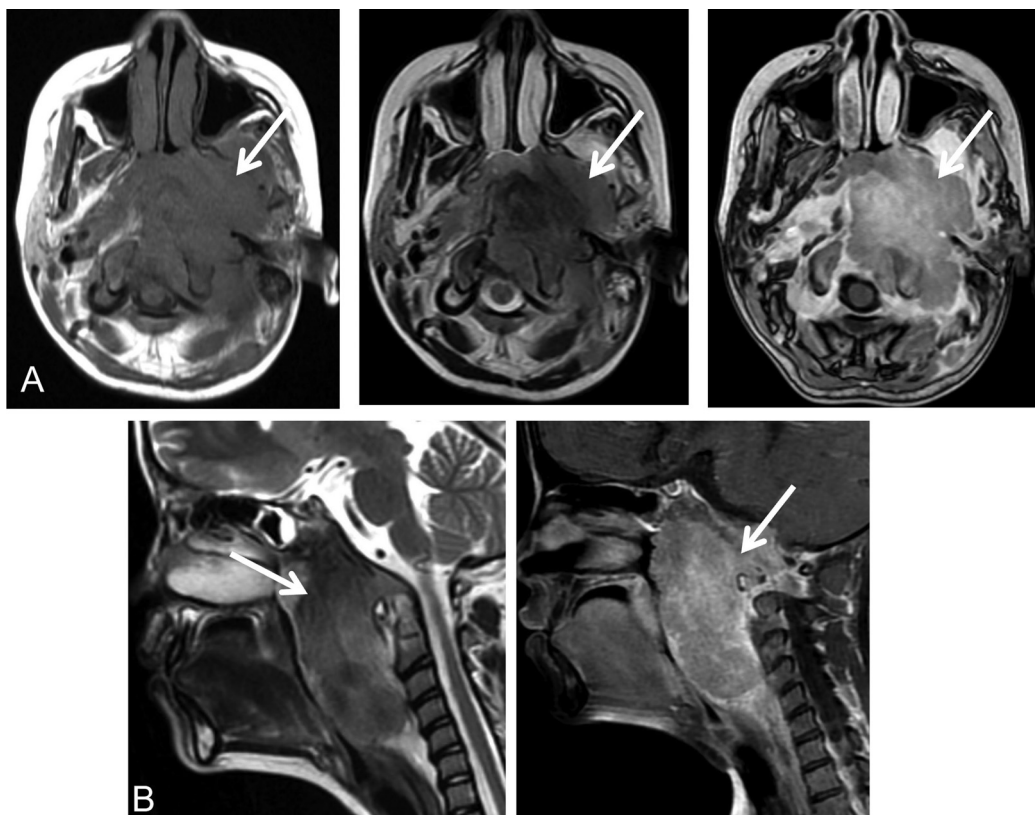


Fig. 21 Nasopharyngeal carcinoma invading clivus: Axial T1-weighted (T1W), T2W, and post-contrast T1W (A); sagittal T2W and post-contrast T1W (B) images showing a large lobulated T1 hypointense, T2 predominantly hypointense heterogeneously enhancing mass (arrows) epicentered in the nasopharynx extending upto left choana. It is causing near complete erosion of the clivus, body of sphenoid, and left half anterior arch of atlas.

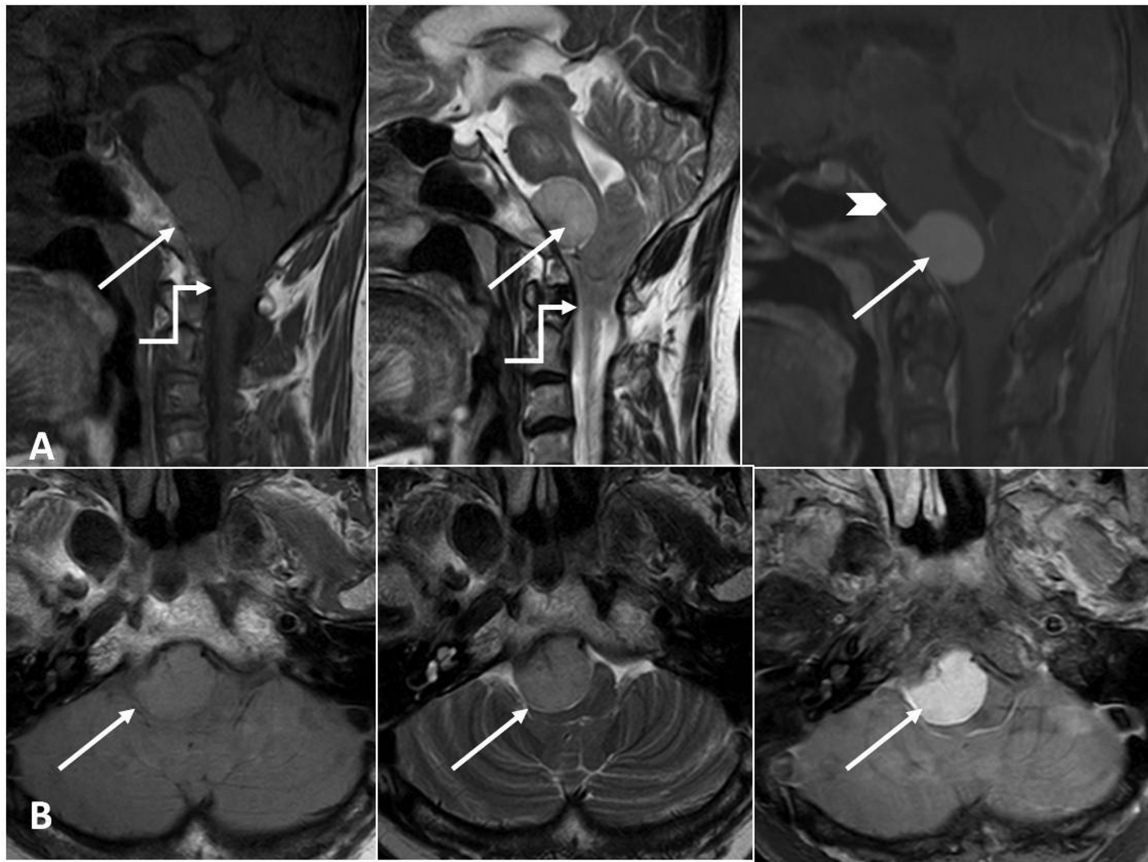


Fig. 22 Clival meningioma. Sagittal (A) and axial (B) T1-weighted (T1W), T2W, and post-contrast T1 fat saturated images showing a well-defined homogeneously enhancing extra-axial lesion (arrow) with broad based at clivus indenting and compressing the medulla. Note dural tail along clivus superior to the lesion (arrowhead). Incidentally peg like tonsils (elbow arrow) due to Chiari 1 malformation.

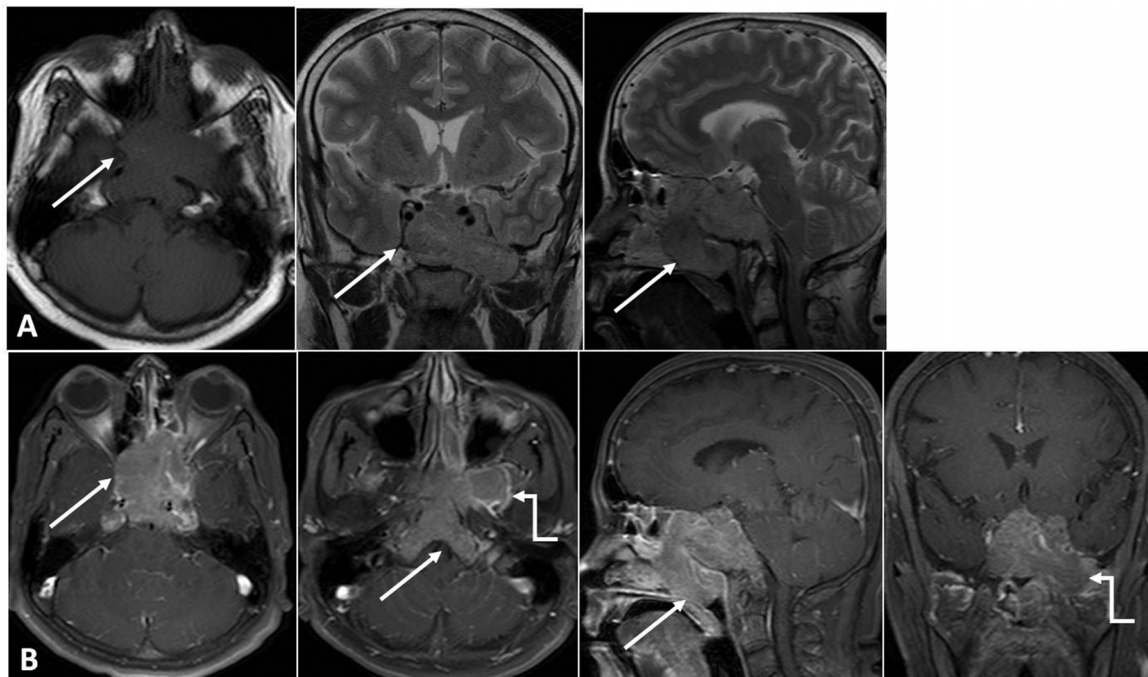


Fig. 23 Neuroendocrine tumor infiltrating clivus: Axial T1-weighted (T1W), coronal, and sagittal T2W (A) and post-contrast fat-saturated (B) images in a 55-year-old female showing a heterogenous mass (arrow) involving nasopharynx, clivus, sella as well as encasing both the petrous and cavernous internal carotid arteries. The tumor also extends into left pterygopalatine fossa (elbow arrow).

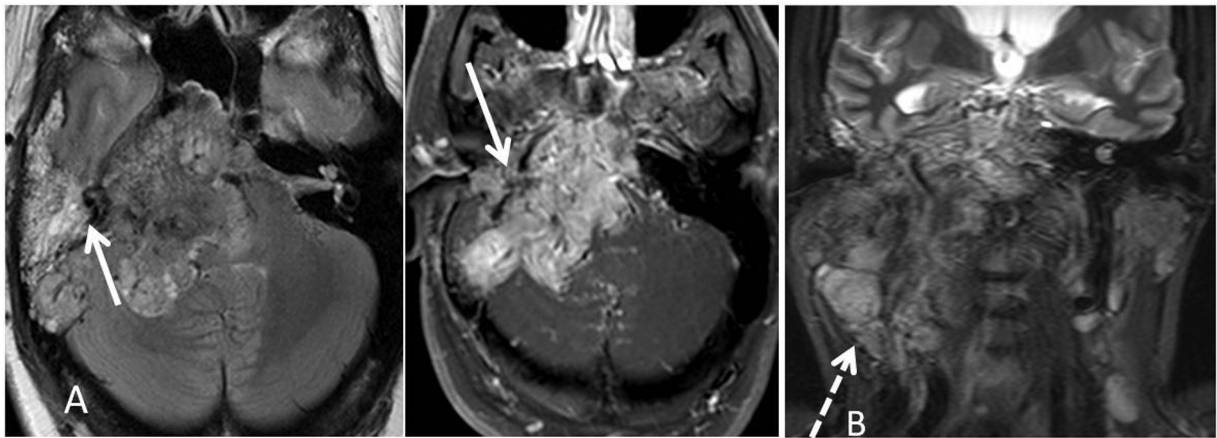


Fig. 24 Paraganglioma invading clivus: Axial T2, axial post-contrast (A) and coronal T2-weighted fat sat magnetic resonance images (B) show a large heterogeneously enhancing mass (arrow) involving right side of skull base and petrous apex with extension into neck (dotted arrow). Note the classic “salt and pepper” appearance (best appreciated in A): T2 bright foci (salt) and T2 hypointense flow voids (pepper).

Table 4 Summary of key imaging features of clival–paraclival lesions

Lesion	Key imaging features
1. Chordoma	<ul style="list-style-type: none"> • T2 very hyperintense, T1 iso to hypointense • Heterogeneous “Honeycomb” enhancement • Midline • Thumb sign • Chondroid variant: calcifications common
2. Chondrosarcoma	<ul style="list-style-type: none"> • T2 hyperintense, T1 hypointense • Intense heterogenous post-contrast enhancement • Off midline • Rings and arcs calcifications
3. Ecthorodosis physaliphora	<ul style="list-style-type: none"> • T2 hyperintense, T1 hypointense • No post-contrast enhancement • Stalk like clival connection on high-resolution heavily T2-weighted sequence like CISS/FIESTA
4. Fibrous dysplasia	<ul style="list-style-type: none"> • Ground-glass density, ballooning and expansion of the affected bone, thinning of the cortex
5. Plasmacytoma	<ul style="list-style-type: none"> • T1 iso to hyperintense; T2 iso to hypointense, with post-contrast enhancement. • Osseo destructive on CT • Solitary lesion
6. Lymphoma	<ul style="list-style-type: none"> • Isointense or hypointense mass on both T1- and T2-weighted images with homogeneous enhancement and restricted diffusion
7. Giant cell tumor	<ul style="list-style-type: none"> • T2 hypointense soft tissue lesion showing homogenous post-contrast enhancement
8. Fungal infection	<ul style="list-style-type: none"> • T2 hypointense
9. Aneurysmal bone cyst	<ul style="list-style-type: none"> • Multiple fluid levels and multiple internal septations on MRI
10. Cholesteatoma	<ul style="list-style-type: none"> • High signal on DWI
11. Meningioma	<ul style="list-style-type: none"> • Hyperdense on noncontrast-enhanced CT • Homogeneous enhancement • Hypo to isointense on T1- and T2-weighted sequences • Dural tail
12. Paraganglioma	<ul style="list-style-type: none"> • On CT, a characteristic “moth-eaten” pattern of destruction of bone • Intense enhancement • “Salt and pepper” pattern on T1- and T2-weighted sequences

Abbreviations: CISS, constructive interference in steady state; CT, computed tomography; DWI, diffusion-weighted imaging; MRI, magnetic resonance imaging.

Conclusion

The clivus can thus be involved by a wide spectrum of conditions ranging from congenital to neoplastic. Owing to the vague clinical presentation and limited clinical accessibility to this region, imaging is of paramount importance in approximating to the final diagnosis and mapping out the disease extent. Hence, a clear understanding of clival anatomy and the radiological appearances of various clival and paraclival pathologies plays a pivotal role in early diagnosis (► **Table 4**) and defining the most effective therapeutic or surgical approach for their management.

Conflict of Interest

None declared.

References

- Rai R, Iwanaga J, Shokouhi G, et al. A comprehensive review of the clivus: anatomy, embryology, variants, pathology, and surgical approaches. *Childs Nerv Syst* 2018;34(08):1451–1458
- Kunimatsu A, Kunimatsu N. Skull base tumors and tumor-like lesions: a pictorial review. *Pol J Radiol* 2017;82:398–409
- Chaljub G, Van Fleet R, Guinto FC Jr, Crow WN, Martinez L, Kumar R. MR imaging of clival and paraclival lesions. *Am J Roentgenol* 1992;159(05):1069–1074
- Kimura F, Kim KS, Friedman H, Russell EJ, Breit R. MR imaging of the normal and abnormal clivus. *Am J Roentgenol* 1990;155(06):1285–1291
- Bahrami S, Yim CM. Quality initiatives: blind spots at brain imaging. *Radiographics* 2009;29(07):1877–1896
- Rathore YS, Sinha S, Mahapatra AK. Transsellar transsphenoidal encephalocele: a series of four cases. *Neurol India* 2011;59(02):289–292
- Caprioli J, Lesser RL. Basal encephalocele and morning glory syndrome. *Br J Ophthalmol* 1983;67(06):349–351
- Chua R, Shapiro S. A mucopyocele of the clivus: case report. *Neurosurgery* 1996;39(03):589–590, discussion 590–591
- Kösling S, Hintner M, Brandt S, Schulz T, Bloching M. Mucocoeles of the sphenoid sinus. *Eur J Radiol* 2004;51(01):1–5
- Sadashiva N, Nandeesh BN, Shukla D, Bhat D, Somanna S, Devi BI. Isolated sphenoid sinus lesions: experience with a few rare pathologies. *J Neurosci Rural Pract* 2017;8(01):107–113
- Fujimoto Y, Harada Y, Toyota S, Wakayama A, So H, Yoshimine T. Primary clival mucocoele: case report. *Neurol Med Chir (Tokyo)* 2011;51(03):250–252
- Clark MP, Pretorius PM, Byren I, Milford CA. Central or atypical skull base osteomyelitis: diagnosis and treatment. *Skull Base* 2009;19(04):247–254
- Khan MA, Quadri SAQ, Kazmi AS, et al. A comprehensive review of skull base osteomyelitis: diagnostic and therapeutic challenges among various presentations. *Asian J Neurosurg* 2018;13(04):959–970
- Bhoi SK, Naik S, Jha M, Pradhan N. Skull base and calvarial tuberculosis presenting as chronic meningitis. *Indian J Tuberc* 2020;67(03):411–413
- Indira Devi B, Tyagi AK, Bhat DI, Santosh V. Tuberculous osteitis of clivus. *Neurol India* 2003;51(01):69–70
- Razek AA, Huang BY. Lesions of the petrous apex: classification and findings at CT and MR imaging. *Radiographics* 2012;32(01):151–173
- Chole RA, Donald PJ. Petrous apicitis. Clinical considerations. *Ann Otol Rhinol Laryngol* 1983;92(6 Pt 1):544–551
- Sabeti S, Yousefi F, Pourabdollah Toutkaboni M. Enchondroma of the skull base in a case of Ollier's syndrome. *Iran J Pathol* 2015;10(03):237–242
- Agrawal A, Shanthi V, Ramakrishna BA. Giant enchondroma of the clivus. *J DattaMeghe Institute Med Sci Uni* 2017;12(03):226
- Gomez CK, Schiffman SR, Bhatt AA. Radiological review of skull lesions. *Insights Imaging* 2018;9(05):857–882
- Ustabasioglu FE, Samanci C, Asik M, et al. Aneurysmal bone cyst of sphenoid bone and clivus misdiagnosed as chordoma: a case report. *Brain Tumor Res Treat* 2015;3(02):115–117
- Aghaghazvini L, Sedighi N, Karami P, Yeganeh O. Skull base aneurysmal bone cyst presented with foramen jugular syndrome and multi-osseous involvement. *Iran J Radiol* 2012;9(03):157–160
- Shibao S, Toda M, Yoshida K. Giant cell tumors of the clivus: case report and literature review. *Surg Neurol Int* 2015;6(Suppl 25):S623–S627
- Zhao J, Qian T, Zhi Z, et al. Giant cell tumor of the clivus: a case report and review of the literature. *Oncol Lett* 2014;8(06):2782–2786
- Yuh SJ, Woulfe J, Corsten MJ, Carrau RL, Prevedello DM, Kassam AB. Diagnostic imaging dilemma of a clival lesion and its clinical management implications. *J Neurol Surg B Skull Base* 2014;75(03):177–182
- Mehnert F, Beschoner R, Küker W, Hahn U, Nägele T. Retroclival ecchordosis physaliphora: MR imaging and review of the literature. *Am J Neuroradiol* 2004;25(10):1851–1855
- Theodorou DJ, Theodorou SJ, Kakitsubata Y. Imaging of Paget disease of bone and its musculoskeletal complications: review. *AJR Am J Roentgenol* 2011;196(6, Suppl):S64–S75
- Baráth K, Huber AM, Stämpfli P, Varga Z, Kollias S. Neuroradiology of cholesteatomas. *AJNR Am J Neuroradiol* 2011;32(02):221–229
- Ahmed M, Sureka J, Koshy CG, Chacko BR, Chacko G. Langerhans cell histiocytosis of the clivus: an unusual cause of a destructive central skull base mass in a child. *Neurol India* 2012;60(03):346–348
- D'Ambrosio N, Soohoo S, Warshall C, Johnson A, Karimi S. Craniofacial and intracranial manifestations of Langerhans cell histiocytosis: report of findings in 100 patients. *Am J Roentgenol* 2008;191(02):589–597
- Erdem E, Angtuaco EC, Van Hemert R, Park JS, Al-Mefty O. Comprehensive review of intracranial chordoma. *Radiographics* 2003;23(04):995–1009
- Azzopardi C, Grech R, Mizzi A. Teaching NeuroImages: chordoma. *Neurology* 2014;83(10):e110–e111
- Raut AA, Naphade PS, Chawla A. Imaging of skull base: pictorial essay. *Indian J Radiol Imaging* 2012;22(04):305–316
- Amita R, Sandhyamani S, Nair S, Kapilamoorthy TR. Plasmacytoma of the clivus. *Asian J Neurosurg* 2017;12(03):573–575
- Kalwani N, Remenschneider AK, Faquin W, Ferry J, Holbrook EH. Plasmacytoma of the clivus presenting as bilateral sixth nerve palsy. *J Neurol Surg Rep* 2015;76(01):e156–e159
- Bhartiya M, Pachisia A, Kapoor R, Vardhan V. Clivus and dural involvement in a case of multiple myeloma: a rare complication of multiple myeloma. *Indian J Hematol Blood Transfus* 2016;32(Suppl 1):135–137
- Marinelli JP, Modzeski MC, Lane JL, et al. Primary skull base lymphoma: manifestations and clinical outcomes of a great imitator. *Otolaryngol Head Neck Surg* 2018;159(04):643–649
- Deconde AS, Sanaiha Y, Suh JD, Bhuta S, Bergsneider M, Wang MB. Metastatic disease to the clivus mimicking clival chordomas. *J Neurol Surg B Skull Base* 2013;74(05):292–299
- Chen X, Dai J, Ai L, et al. Clival invasion on multi-detector CT in 390 pituitary macroadenomas: correlation with sex, subtype and rates of operative complication and recurrence. *Am J Neuroradiol* 2011;32(04):785–789
- Rao AB, Koeller KK, Adair CF. Armed Forces Institute of Pathology. From the archives of the AFIP. Paragangliomas of the head and neck: radiologic-pathologic correlation. *Radiographics* 1999;19(06):1605–1632
- Debnam JM. Imaging of the head and neck following radiation treatment. *Pathol Res Int* 2011;2011:607820