Section: Radiodiagnosis

Case Series

THE POWER OF IMAGING: UNRAVELLING CHORDOMA WITH MRI AND CT

Naveen D[1], Madhu S Kikkeri[2], Achala Shravya G[3], Vinaya Ratna S[4], Sindhu K[5], Poornima G B[6]

[1]Associate Professor, Department of Radiodiagnosis, Sapthagiri Institute of Medical Sciences and Research Centre, Bangalore, Karnataka, India.
[2]Assistant Professor, Department of Radiodiagnosis, Sapthagiri Institute of Medical Sciences and Research Centre, Bangalore, Karnataka, India.
[3-6]Post Graduate, Department of Radiodiagnosis, Sapthagiri Institute of Medical Sciences and Research Centre, Bangalore, Karnataka, India.

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Corresponding Author:
Dr. Madhu S Kikkeri,
Email: drmadhuskikkeri@gmail.com

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Abstract

Chordomas, rare tumours affecting individuals of all ages, exhibit a diverse range of clinical behaviours and demographic variations. These tumours present with unique clinical characteristics and demographic predispositions, contributing to the complexity of diagnosis and management. Despite their rarity, chordomas present significant challenges in both diagnostic accuracy and treatment strategies due to their slow growth and nonspecific symptomatology. Radiological assessment, particularly through computed tomography (CT) and magnetic resonance imaging (MRI), plays a crucial role in achieving precise diagnosis and formulating effective treatment plans. These imaging modalities provide comprehensive insights into chordoma characteristics, aiding in their differentiation from other lesions and guiding therapeutic decisions.

INTRODUCTION

Chordomas are rare malignancies originating from embryonic remnants of the primitive notochord, a foundational cellular structure critical for skull base and vertebral column development. These residual notochord elements typically endure near or within the midline, encased within bony structures.[1]

Given their notochordal origin, chordomas primarily manifest within the axial skeleton. Around 50% arise in the sacrococcygeal region, 35% in the skull base, and the remaining 15% within the vertebrae of the mobile spine.[2]

Although chordoma are generally slow growing lesions, the recurrence rate is high and the location makes it often difficult to treat. Both computed tomography (CT) and magnetic resonance imaging (MRI) are crucial in the initial diagnosis, treatment planning and post-treatment follow-up.[3]

CASE 1

A 4-year female child presented with difficulty in neck movements and weakness of right upper and lower limb. She was referred for MRI brain with whole spine screening which showed a lobulated extra axial T2/T1/FLAIR predominantly isointense lesion arising from lower part of clivus. Anteriorly the lesion was extending into the nasopharynx. Inferiorly it was extending into the cervical spinal canal till C3 level. Posteriorly the lesion was extending into premedullary cisternal space, compressing and posteriorly displacing the underlying brain stem and cervical spinal cord posterosuperiorly [Figure 1 A, B, C]. It showed predominantly peripheral diffusion restriction [Figure 1 E, F] and heterogenous enhancement post contrast [Figure 1 H]. CT sections showed irregular destruction of clivus with few calcific specks within [Figure 1 D]. Patient underwent Lateral craniotomy and subtotal excision of the lesion and the HPE came out as chordoma.

CASE 2

A 46-year-old female presented with headache, paraesthesia on right side of the face and neck. A Plain CT Brain study was ordered which showed an expansile lytic lesion involving the right spine of sphenoid, right lateral aspect of body of sphenoid and clivus with soft tissue component [Figure 2 E]. Further a CE-MRI brain was done which showed a well-defined T2 and FLAIR heterogeneously hyperintense extra axial lesion in the right middle cranial fossa filling the Meckel's cave and extending into the right cerebellopontine angle and posterior cranial fossa on the right side. The lesion was closely abutting and displacing the brainstem towards left and was closely abutting the right lateral aspect of clivus and the basilar artery on the medial aspect with well-maintained basilar artery flow void [Figure 2 A, B, C]. The lesion was hypointense on T1W sequence

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[Figure 2 D]. Post contrast the lesion showed intense heterogenous enhancement with few non enhancing necrotic areas within [Figure 2 F, G]. In CISS sequence the cisternal part of right trigeminal nerve was not separately made out from the lesion [Figure 2 H]. With the above imaging features, DDs of trigeminal schwannoma, chordoma or petroclival meningioma were considered. The patient underwent right Fronto temporo parietal craniotomy and excision of the lesion and the HPE was found to be consistent with a chordoma.

CASE 3

A 26-year-old male presented with severe headache and diplopia. A CT brain was ordered which revealed a large hypodense lesion of soft tissue density measuring 2.8 x 3.1 x 2.6 cm (AP x TR x CC) with coarse internal and peripheral calcifications, epicentered within the body of sphenoid bone causing lytic destruction of the anterior body of clivus and floor of sella [Figure 3 E]. It was followed up with a CE-MRI Brain study which showed a well-defined extra axial T2W, FLAIR heterogeneously hyperintense, T1 hypo to isointense lesion epicentered in the basi-sphenoid and clivus bone (more on the right side) extending laterally into cavernous sinus and anteriorly into the sphenoid sinus bilaterally. Flow void of bilateral ICA was well maintained (Figure 3- A, B, C). Posteriorly the lesion was extending into pre pontine cistern, compressing and displacing the pons posteriorly, closely abutting the basilar artery with well-maintained flow void [Figure 3 A, B, C, D]. On SWI, few areas of blooming were seen in the peripheral aspect [Figure 3 F]. Patient underwent sublabial transsphenoidal excision of the lesion and HPE revealed it to be chordoma.

CASE 4

A 76-year-old female presented with headache and a CT Brain was done which showed a large irregular expansile lytic lesion with soft tissue component epicentred within the clivus causing cortical destruction of the entire clival bone and 1st cervical vertebra [Figure 4 A,B,C,D]. Patient underwent sublabial transsphenoidal excision of the lesion and HPE was consistent with a chordoma.

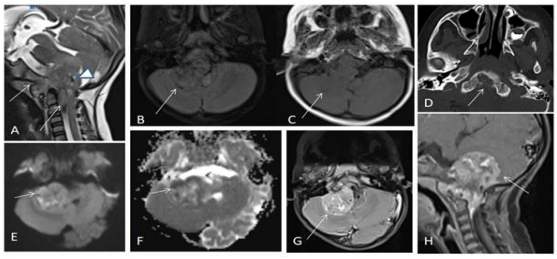

Figure 1: (A) T2WI in sagittal section showing a lobulated extra axial isointense lesion arising from lower part of clivus. Anteriorly the lesion is seen extending into the nasopharynx. Inferiorly it is seen extending into the cervical spinal canal till C3 level (white arrows). Posteriorly the lesion is extending into premedullary cisternal space, compressing and posteriorly displacing the underlying brain stem and cervical spinal cord posterosuperiorly (white arrowhead). (B) FLAIR axial section showing isointense lesion which is posteriorly abutting the right cerebellar hemisphere (white arrow). (C) The lesion is isointense on T1W sequence (white arrow). (D) CT axial section in bone window showing irregular destruction of clivus with few calcific specks within (white arrow). (E & F) DWI and ADC map showing predominantly peripheral diffusion restriction in the lesion (white arrows). (G & H) Post contrast the lesion shows heterogeneous enhancement (white arrows). HPE: Chordoma

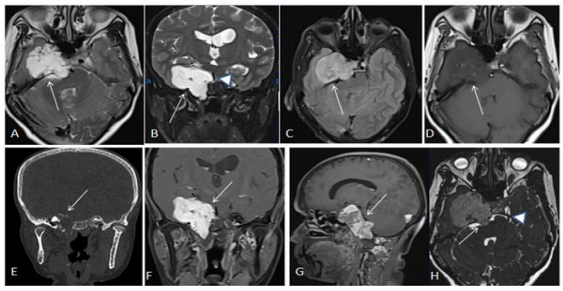

Figure 2: (A & C) Axial sections in T2 and FLAIR showing a well-defined heterogeneously hyperintense extra axial lesion in the base of middle cranial fossa on right side filling the Meckel's cave and extending into the right cerebellopontine angle and posterior cranial fossa on the right side (white arrows). It is seen closely abutting and displacing the brainstem towards left. (B) Coronal T2WI showing the lesion is medially abutting the right lateral aspect of clivus and the basilar artery (white arrow head). (D) The lesion shows hypointensity on T1W sequence (white arrow). (E) Coronal CT section in bone window showing expansile lytic lesion involving the right spine of sphenoid, right lateral aspect of body of sphenoid and clivus (white arrow). (F & G) Post contrast T1FS sequence in coronal and sagittal sections shows intense enhancement of the lesion (white arrows). (H) CISS sequence depicting that the cisternal segment of right trigeminal nerve is not separately made out from the lesion (white arrow). Cisternal segment of left trigeminal nerve is well made out (white arrow head). With above imaging features, DDs of trigeminal schwannoma, chordoma or petroclival meningioma were considered. HPE: Chordoma

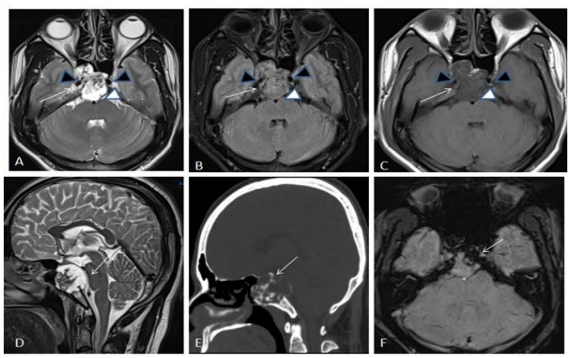

Figure 3: (A, B, C) Axial T2W, FLAIR, T1W sequence showing well-defined extra axial heterogeneously hyperintense lesion epicentered in the basi-sphenoid and clivus bone (more on the right side) extending

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laterally into cavernous sinus and anteriorly into the sphenoid sinus bilaterally (white arrows). Flow void of bilateral ICA is well maintained 9 (black arrow head). Posteriorly the lesion is extending into pre pontine cistern, compressing and displacing the pons posteriorly, closely abutting the basilar artery with well-maintained flow void (white arrow head). The lesion appears heterogeneously hyperintense on T2 (A) with partial suppression on FLAIR (B), iso-hypointnese on T1WI (C). (D) Sagittal T2W sequence showing posterior displacement of pons by the lesion (white arrow). (E) Sagittal CT section in bone window showing lytic destruction of the anterior body of clivus, floor of sella (white arrow). (F) On SWI, there are few areas of blooming in the peripheral aspect (white arrow) . HPE: Chordoma

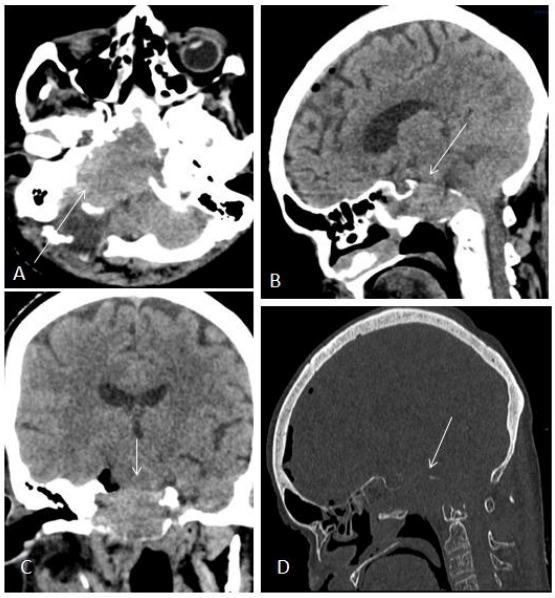

Figure 4: (A, B, C) Representative CT sections in brain window in axial, sagittal and coronal cuts showing a large irregular expansile lytic lesion with soft tissue component epicentred within the clivus (white arrows). (D) Sagittal section in bone window showing expansile lytic lesion showing cortical destruction of the entire clival bone (white arrow). HPE: Chordoma.

DISCUSSION

Chordomas typically impact individuals in their 60s, yet they can occur across all age groups. Recent descriptive epidemiological data from the United States indicates an age-adjusted incidence rate of 0.88 per million persons per year for all chordoma types between 2001 and 2014. In Taiwan, the annual age-standardized incidence rate from 2003 to 2010 was 0.4 per million, with a higher prevalence among males. Asian/Pacific individuals exhibit a higher incidence of cranial chordomas, whereas spinal and sacral chordomas are most common in the Caucasian population.[4]

Chordomas also have implications for children and young adults, with females showing a notable predominance in the paediatric cohort. In this age group, these tumours tend to emerge primarily at the craniocervical junction, with a decreasing frequency as one moves down the spine. This distribution pattern differs from that observed in adults, where the craniocervical and sacrococcygeal regions are more commonly affected.[4]

Pathologically, chordomas are classified into three distinct categories according to the fifth edition of the World Health Organization (WHO) classification. The majority of cases are attributed to conventional and/or chondroid chordomas, with less common subtypes including poorly differentiated chordoma and dedifferentiated chordoma. Conventional chordomas represent approximately 95% of all instances. They predominantly afflict adults, exhibit a higher prevalence in males than females, and are more frequently observed in the sacral region.[2,5,6]

Poorly differentiated chordoma, a rare variant, primarily affects children. The average age at diagnosis is around 10 years, with cases reported from as young as 3 months to as old as 42 years. There's a slight female predominance, and these tumours are more often found in the clivus, skull base, and cervical spine.[2,7]

Dedifferentiated chordoma is an even rarer occurrence, representing only 2% to 8% of cases. Limited epidemiological data indicate an average age of 57, ranging from 15 to 81 years. This subtype is slightly more prevalent in males and tends to occur more frequently in the sacral region.[2,6]

Symptoms experienced by patients with chordomas vary depending on the location of the tumour along the axial skeleton. For instance, those with sacral chordomas may present with symptoms such as back pain, urinary or bowel problems, neuropathy, or difficulties with walking. On the other hand, individuals with clival chordomas may report headaches, double vision, or dysfunction of cranial nerves. Diagnosis of chordomas can often be delayed for months to years due to the slow growth of the tumour, the presence of nonspecific symptoms, and clinicians' limited familiarity with this rare condition.[2]

Intracranial chordomas often originate from the spheno-occipital synchondrosis found in the clivus. The point of onset can occur either at the upper clivus (basisphenoid) or at the lower part of the clivus (basiocciput).[1]

Spinal chordomas typically develop within the vertebral body, mainly due to the association of the notochord with the developing axial skeleton. Vertebral sclerosis can be a notable feature of chordomas. The lytic bone destruction, along with the extension of soft tissue masses, leads to erosion of the outer cortex under pressure and a spiculated periosteal reaction. These lesions frequently result in the widening of both the transverse and intervertebral foramina.[8]

Generally, chordomas appear as midline, aggressively invasive osteolytic lesions with accompanying soft-tissue masses in radiographs. Occasionally, intratumoral calcifications may be discernible.[4]

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The radiological evaluation of intracranial chordomas has been transformed by CT and MRI, enhancing the accuracy of diagnosis and tumour delineation at the skull base. These advances are crucial for surgeons planning extensive resections and radiation oncologists defining tumor boundaries and critical anatomical structures for targeted therapy. Typically, both imaging modalities are required for a thorough pre-treatment assessment.[1]

High-resolution CT scans characteristically show intracranial chordomas as well-circumscribed, expansile soft-tissue masses emanating from the clivus, with extensive associated bone destruction. These tumours generally have higher attenuation than the surrounding neural tissues, with irregular calcifications that suggest bone debris rather than intrinsic calcifications. After contrast administration, the lesions often enhance markedly, and areas of low attenuation may be visible, corresponding to the myxoid and gelatinous components identified on pathological examination.[1]

MRI is the preferred method for detailed visualization of intracranial chordomas, offering superior tissue contrast and anatomical detail, significantly outperforming CT in mapping the extent of the lesions.[1]

Chordomas typically exhibit low-to-intermediate signal on T1-weighted MRI images and marked hyperintensity on T2-weighted images, primarily due to the presence of vacuolated cellular components containing high fluid content. Occasionally, small areas of hyperintense signal on T1-weighted images may indicate intratumoral haemorrhage or proteinaceous material. Following gadolinium injection, moderate enhancement is usually observed, although mild enhancement may occur if the tumour is necrotic. Complications such as spinal cord compression and invasion of adjacent nerves and blood vessels can be visualized. Given their midline location and aggressive nature, chordomas often involve vital neurovascular structures. In cases where vascular encasement is suspected, CT and MR angiography may be necessary to assess arterial patency.[4,8]

According to available literature, cervical chordomas display several distinctive characteristics. These include lytic and sclerotic bone destruction accompanied by a soft tissue mass. This mass may present with a "collar button" appearance in the sagittal plane and display a multi-lobular configuration surrounding the vertebrae in the axial plane. Other notable features include pressure erosion of the outer cortex and a spiculated periosteal reaction, as well as widening of the transverse and intervertebral foramina. Encasement of the vertebral artery is also observed, along with calcification. On CT scans, the tumour composition appears heterogeneous and hypodense compared to adjacent muscle. In magnetic resonance T2-weighted images, the composition typically appears heterogeneous and hyperintense, resembling cerebrospinal fluid, with hypointense septa.[8]

Another consideration for differential diagnosis is Benign Notochordal Cell Tumour (BNCT). BNCT typically preserves trabecular architecture without causing bone destruction or expansion. Additionally, it lacks soft tissue extension and does not exhibit enhancement following contrast administration. These radiographic features are commonly relied upon to distinguish BNCT from chordoma.

Chondrosarcoma shares similar MRI characteristics with chordoma. However, it more frequently involves the neural arch rather than the vertebral body. Moreover, chondrosarcoma often presents with a chondroid matrix, which manifests as characteristic rings and arcs.[8]

Chordomas are characterized by a wide range of clinical behaviours, spanning from relatively slow-growing to highly aggressive tumours. Despite presenting as indolent in certain cases, loco-regional recurrence is a common occurrence, observed in up to 50% of patients in some case series. Additionally, approximately 30%-40% of patients eventually develop distant metastatic disease, with common sites including the lungs, liver, and bones. Less frequently, metastases may involve lymph nodes, soft tissues, and dermal sites.

When managing loco-regional recurrent chordomas, guidelines typically advocate for a multi-modal approach, combining surgical intervention with radiation therapy. Other local control options are available and may include definitive radiation therapy (without surgical intervention), as well as innovative techniques such as radiofrequency ablation and cryoablation.[2]

CONCLUSION

Chordoma, a rare spinal tumour affecting a wide age range, poses significant challenges in treatment due to its propensity for relapse, local recurrence, and metastasis. Both CT and MRI play crucial roles in diagnosing chordoma, with CT excelling in assessing bone abnormalities and MRI highlighting specific tumour features. Their combined use enables accurate diagnosis and informs treatment decisions.

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