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**Radiology**

# Clival Chordoma Presenting with Ophthalmoplegia and Frontal Headache: A Radioclinical Case Report

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**Abstract** | **Case Report**

Chordomas are rare, slow-growing, locally aggressive malignant tumors arising from remnants of the notochord, commonly involving the clivus at the skull base. We present a case of a 38-year-old male with a 2-year history of progressive diplopia and ophthalmoplegia, recently worsened by frontal headaches. Imaging revealed a destructive, expansile skull base mass centered on the spheno-occipital synchondrosis, extending to adjacent critical neurovascular structures. Histopathology confirmed the diagnosis of clival chordoma. This case highlights the characteristic radiological features of chordoma and its potential for extensive local invasion.

**Keywords:** Chordoma, Skull base tumor, Clivus, Diplopia, Ophthalmoplegia.



## INTRODUCTION

Chordomas account for approximately 1–4% of all primary malignant bone tumors, predominantly affecting the axial skeleton, with 35–40% occurring at the skull base [1, 2]. The clival region is the most common intracranial site due to persistence of notochordal remnants. Diagnosis is often delayed due to insidious growth and nonspecific symptoms related to cranial nerve involvement. Imaging plays a critical role in diagnosis, surgical planning, and follow-up.

## CASE PRESENTATION

A 38-year-old male presented with a 2-year history of diplopia and ophthalmoplegia, recently accompanied by frontal headaches for the past 2 months. A prior CT scan had identified a lytic skull base lesion, prompting a surgical biopsy suggestive of a skull base tumor. Further imaging was undertaken in our department as part of presurgical planning.

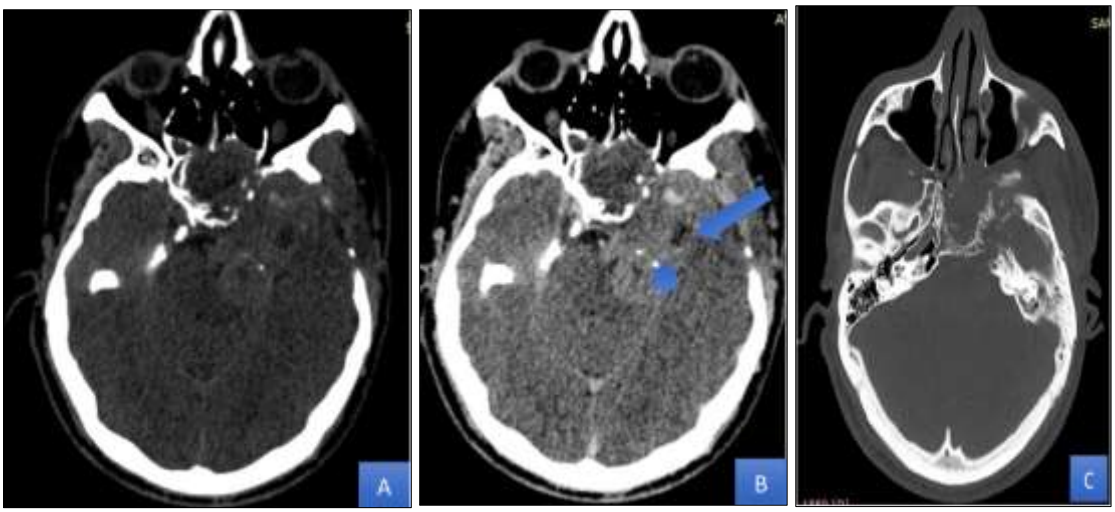
### Imaging Findings

CT and MRI of the brain and skull base were performed. The lesion was a well-circumscribed, lobulated, expansile, destructive soft tissue mass centered on the spheno-occipital synchondrosis extending to the sellar, suprasellar, parasellar, and infratemporal regions. On MRI, the lesion showed heterogeneous intermediate-to-low signal on T1-weighted images and heterogeneous high signal on T2-weighted and FLAIR sequences. Diffusion-weighted imaging (DWI) demonstrated hyperintensity with corresponding low apparent diffusion coefficient (ADC) values.

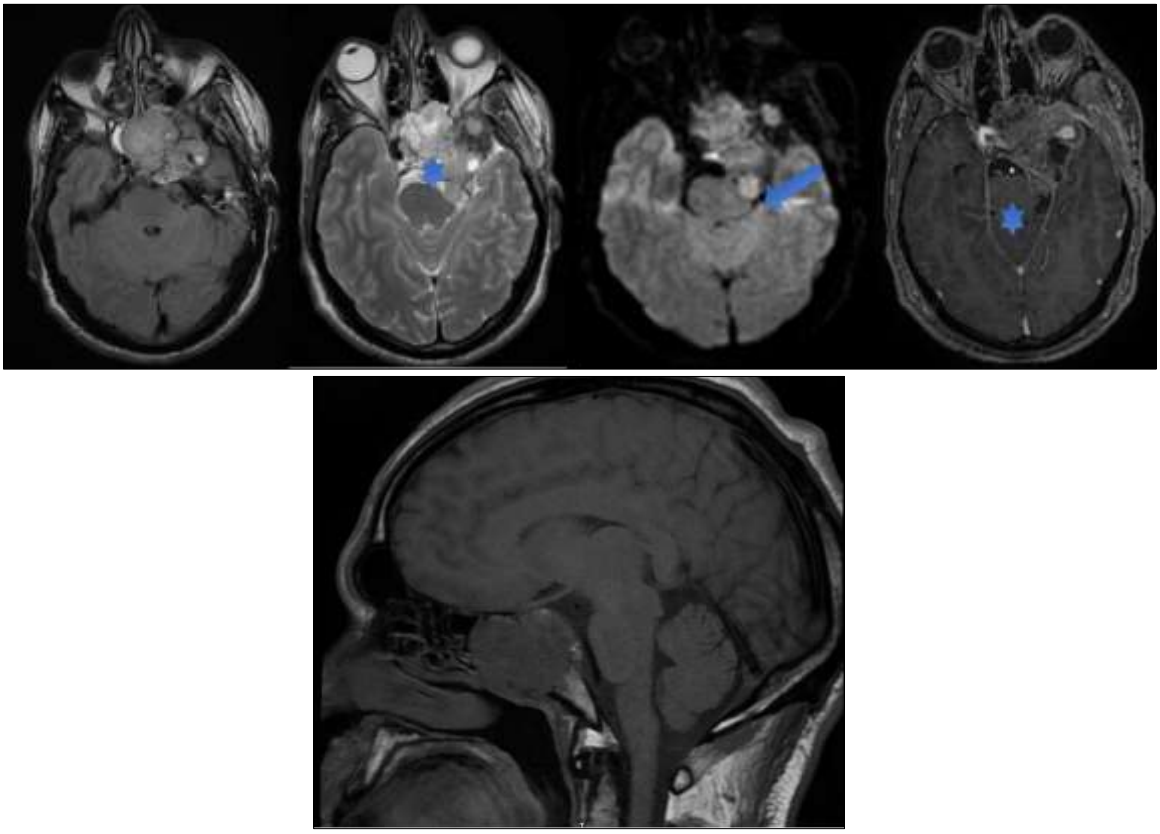
Post-contrast images displayed heterogeneous enhancement with non-enhancing necrotic areas, a honeycomb appearance, and evidence of intratumoral hemorrhage and calcifications, appearing hyperdense on CT, hyperintense on T1, and low signal on T2* gradient echo sequences.

The lesion invaded the left cavernous sinus, abutted the right cavernous sinus without direct invasion, displaced the mesencephalic peduncle and midbrain, and invaded Dorello's canal, Meckel's cave, and the cisternal segments of the trigeminal and abducens nerves. Atrophy of the lateral rectus muscle was noted. It also encased the left internal carotid artery (ICA), reducing its caliber with poor post-contrast enhancement and exhibited a pseudoaneurysmal outpouching.

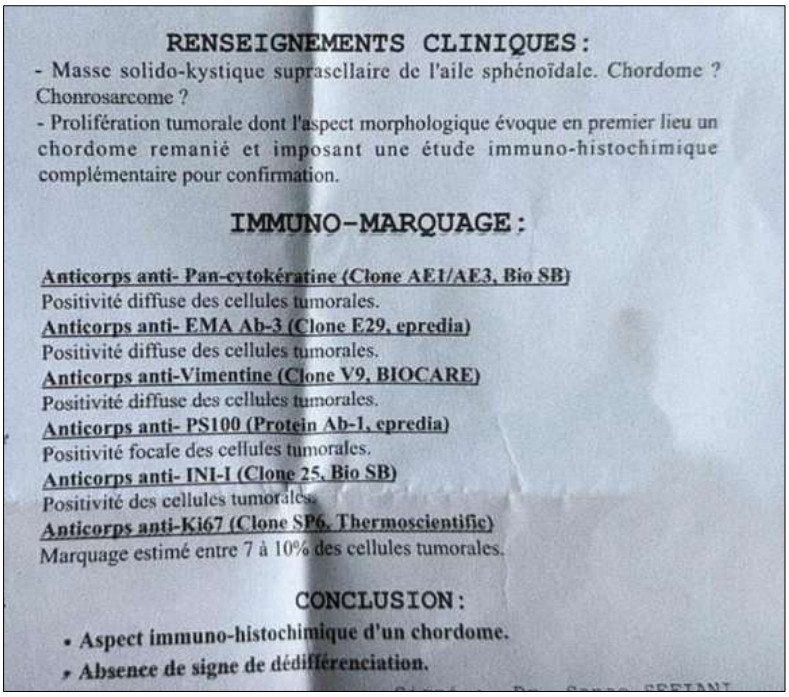
Superiorly, it approached the optic chiasm; laterally, it involved the left temporal lobe and temporal fossa. The lesion extended anteriorly into the sphenoid sinus, ethmoid labyrinth, orbital apex, and posteriorly into the infratemporal fossa, pterygoid region, petrous apex, nd middle ear cavity, resulting in mastoid opacification.




**Axial brain CT before (A)and after contrast administration (B) including bone window reconstructions (C) objectives Skull base well circumscribed, lobulated expansile destructive soft tissue mass centered on the spheno-occipital synchondrosis sellar, suprasellar and left parasellar region, it displays heterogenous enhancement delimitating areas of necrosis(ARROW) and a honey comb appearance (STAR)**



**MRI demonstrates the same extensive bone-infiltration on the spheno-occipital synchondrosis sellar, suprasellar and left parasellar region T1WI signal is intermediate/low, with possible small foci of T1-hyperintensity due to intralesional haemorrhage. T2WI shows a heterogeneously hyperintense lesion due to abundant fluid component combined with vacuolated-cystic areas, along with less intense areas of proteinaceous and haemorrhagic content. Enhancement is typically moderate to intense, with a heterogeneous "honeycomb" appearance due to areas of necrosis and cystic degeneration**



RENSEIGNEMENTS CLINIQUES :

- Masse solido-kystique suprasellaire de l'aile sphénoïdale. Chordome ? Chonrosarcome ?
- Prolifération tumorale dont l'aspect morphologique évoque en premier lieu un chordome remanié et imposant une étude immuno-histochimique complémentaire pour confirmation.

IMMUNO-MARQUAGE :

Anticorps anti- Pan-cytokératine (Clone AE1/AE3, Bio SB)
Positivité diffuse des cellules tumorales.
Anticorps anti- EMA Ab-3 (Clone E29, epredia)
Positivité diffuse des cellules tumorales.
Anticorps anti-Vimentine (Clone V9, BIOCARE)
Positivité diffuse des cellules tumorales.
Anticorps anti- PS100 (Protein Ab-1, epredia)
Positivité focale des cellules tumorales.
Anticorps anti- INI-I (Clone 25, Bio SB)
Positivité des cellules tumorales.
Anticorps anti-Ki67 (Clone SP6, Thermoscientific)
Marquage estimé entre 7 à 10% des cellules tumorales.

CONCLUSION :

- Aspect immuno-histochimique d'un chordome.
- Absence de signe de dédifférenciation.

**Immunohistochemical profile of a suprasellar sphenoidal mass showing markers consistent with chordoma, including diffuse positivity for cytokeratin, EMA, and vimentin, with focal PS100 and preserved INI-1. Findings support a diagnosis of conventional chordoma without dedifferentiation**

## DISCUSSION

Clival chordomas are locally aggressive tumors derived from notochordal remnants, predominantly affecting the midline skull base [1]. Clinically, they manifest with cranial neuropathies, headaches, or visual disturbances due to their proximity to vital neurovascular structures [2, 3].

Radiologically, chordomas typically appear as well-defined, lobulated, midline lesions causing bone destruction. MRI is the modality of choice, with tumors characteristically exhibiting low to intermediate T1 and high T2 signal intensities, often with internal hemorrhage, necrosis, and calcifications [4]. The 'honeycomb' enhancement pattern and involvement of adjacent structures, including the cavernous sinus, ICA, and cranial nerves, are classical findings [5].

**Differential Diagnoses Include:**

- Meningioma: Typically extra-axial, iso- to hypointense on T1 and T2, with homogeneous enhancement and hyperostosis rather than osteolysis [6].
- Craniopharyngioma: Common in the sellar-suprasellar region, usually cystic with calcifications, predominantly seen in pediatric or older adult populations [7].

Histopathology remains the definitive diagnostic modality, typically showing physaliphorous (vacuolated) cells in a myxoid stroma.

Treatment involves maximal safe surgical resection, often via an endoscopic endonasal or open skull base approach, followed by adjuvant radiotherapy, with proton beam therapy preferred due to its conformal dose distribution [8].

## CONCLUSION

This case illustrates the classic radiological features of a clival chordoma with extensive skull base and intracranial extension. Recognition of its imaging characteristics is crucial for early diagnosis, differential consideration, and preoperative planning due to its aggressive behavior and potential for significant morbidity.

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