

# Chordoma of the Sellar Region Mimicking Meningioma: A Literature Review and Illustrative Case

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## ABSTRACT

**Background:** Chordoma is a rare malignant tumor that accounts for 1-4% of primary malignant bone tumors arising primarily in the axial skeleton and primarily affects adults. It seems to have difficulty with diagnosis and treatment.

**Case description:** A 63-year-old man with a history of diabetes treated with oral antidiabetic medication and hypertension. Presented with two years of bilateral decrease in visual acuity. Neurological examination revealed left ptosis, divergent strabismus, and higher cerebral function disorders. Brain MRI revealed a sellar and suprasellar lesion evokes a sellar meningioma, macro adenoma. Hormonal laboratory tests revealed that the prolactin level was elevated to 140ng/ml without any other hormonal disorder. The patient underwent partial surgical resection of the lesion through endoscopic endonasal. Histopathological examination revealed a chondroid chordoma.

**Postoperatively** the patient had no endocrinological or neurological complications, but he kept the strabismus and the ptosis in the left eye.

**Conclusion:** In the context of progressive ophthalmological signs by a sellar and suprasellar lesion, the differential diagnosis should include chordoma along with meningioma, macro adenoma and metastasis.

**Complete and early surgical removal is the first line treatment.**

**Keywords:** chordoma, meningioma, sellar region, surgical remove.

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## I. INTRODUCTION

Chordomas represent a rare entity of slow grow malignant tumors, and classified as tumors of low to intermediate malignancy with a propensity for locally aggressive behavior and rare metastasi; with a slight male predominance. Chordomas are histologically reminiscent of embryonic notochordal tissue, they nearly always arise within bone [1], [2]. It seems to have a therapeutic and diagnostic problem, especially in the sellar and suprasellar location. Several differential diagnoses should be considered to manage the treatment strategy.

We report the case of a patient with chordoma mimicking sellar and supra sellar meningioma.

We conducted a literature review to discuss the management of this rare situation in light of the previously reported case.

## II. CLINICAL CASE

This is a 63-year-old man with a history of diabetes treated with oral antidiabetic medication. He presented with two years of headaches and bilateral decrease in visual acuity.

Neurological examination revealed left ptosis, divergent strabismus, and higher cerebral function disorders, but no endocrine signs were observed. Magnetic resonance imaging (MRI) revealed a sellar and suprasellar lesion that appeared hyperintense on T2-weighted sequence, isointense on T1-weighted sequence, and homogeneously contrast-enhancing, extending laterally to the cavernous sinus. The lesion was compressing the optic chiasma upward and the pituitary stalk, and no other lesions were identified on complete MRI imaging. Hormonal laboratory tests revealed that the prolactin level was elevated to 140 ng/ml without any other hormonal disorder. The preoperative differential diagnoses included meningioma, macro adenoma, and petroclival chondroma with sellar extension. Partial resection through endoscopic endonasal surgery was performed, and a yellowish hemorrhagic tumor with a relatively fibrous texture, aspirable in places, was found. The procedure was limited by hemorrhagic bleeding, with a total blood loss of 150 cc. Histopathological examination revealed a tumor proliferation arranged in lobules and layers surrounded by an abundant myxoid matrix, sometimes chondroid. Immunohistochemical study demonstrated the tumor cells expressed cytokeratin AE1/AE3 (clone AE1/AE3) and EMA

(clone E29), consistent with the diagnosis of a chondroid chordoma.

The patient was hospitalized for 5 days postoperatively, without any endocrinological or neurological complications, but the patient kept the ophthalmological disorders, which are ptosis and strabismus of the left eye, one month later the ptosis improved. The postoperative CT scan showed no tumor recurrence.

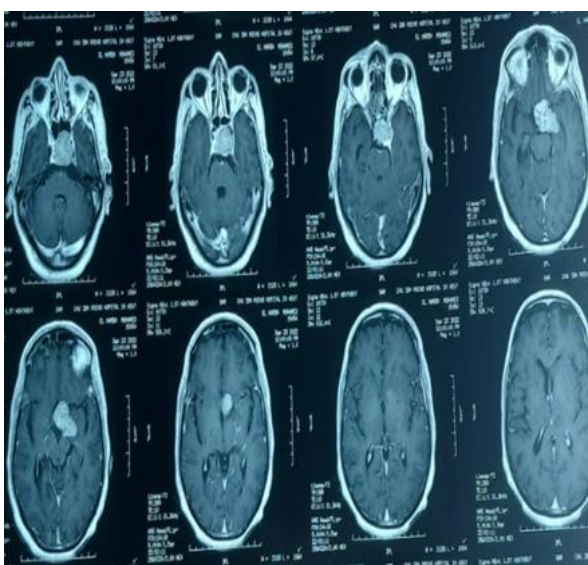
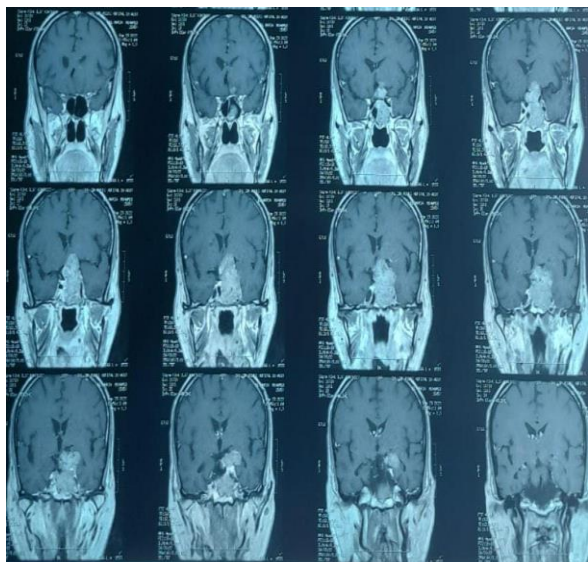


Fig. 1. MRI (axial and coronal sections) of chordoma, mimicking a sellar meningioma with suprasellar extension.

### III. DISCUSSION

Ribbert coined the term chordoma in 1894, to describe a malignant tumor that develops slowly from the remnants of the notochord. The notochord plays a crucial role in the development of the first axial skeleton during embryogenesis. It originates from ectodermal cells in the third week of human embryo development and degenerates after the sixth month of fetal life, leaving behind remnants of notochordal tissue [3].

Chordomas are tumors that affect the axial skeleton, with a higher incidence of lesions at both ends. The distribution of these lesions is as follows [4]: 48-60% in the sacrococcygeal region, 39% in the spheno-occipital region, and 13-20% in

the mobile spine, including 8% in the cervical spine, 5% in the lumbar spine, and only 2% in the thoracic segment [5].

The clinical presentation of chordoma is dependent on the size and location, and is often delayed due to the slow progression of these tumors [6]. While chordomas have a distinct appearance under light microscopy, it is important to consider several pathological differential diagnoses [7].

TABLE I: INTRACRANIAL CHORDOMAS. DIFFERENTIAL DIAGNOSIS [8]

	Elements for diagnosis of chordoma	Elements against diagnosis of chordoma
Invasive pituitary adenoma	Sphenoid tumour with involvement of clivus	Contrast enhancement not very heterogenous, well delineated tumour, no pontine cistern extension, exclusive extension to sphenoid bone, asymmetrical bulging of superior surface of pituitary
Lymphoma	Clivus and nasopharynx involvement	Signal change in clivus without morphological change; no pontine cistern extension
Chondroma	Clivus involvement; heterogeneous and lobulated appearance after gadolinium	Lateral location

Immunohistochemical staining is useful for differentiating chordomas from other tumors. Chordomas are typically positive for cytokeratin, epithelial membrane antigen, S-100 protein, vimentin, and sometimes carcinoembryonic antigen. A nuclear transcription factor called brachyury, associated with fetal notochord, has also been identified as a specific marker for chordomas [9]-[11]. In contrast, chordoid meningiomas, which may be mistaken for chordomas based on their intracranial location, do not express cytokeratin or brachyury and have a different histological pattern. In a recent case, cytokeratin staining was strongly positive, but the absence of a meningioma-like pattern and positive staining for brachyury helped confirm the diagnosis of chordoma [12]. The positivity for cytokeratin, led to the original diagnosis of chordoma in our case.

Chordoid meningiomas are a rare form of meningioma that can develop in both adults and children. Unlike childhood meningiomas, those found in adults typically exhibit little to no chronic inflammation and are not linked to blood disorders. Although chordoid meningiomas share some morphological features with chordomas, distinguishing between the two is essential due to the differences in their behavior. A combination of morphology, immunohistochemistry, and/or electron microscopy is the best approach to achieve this distinction [13].

The primary treatment modality for chordomas typically consists of surgical resection followed by adjuvant radiotherapy. The post-operative imaging is the preferred method for documenting the presence or absence of residual tumor. Unfortunately [14]. The most effective treatment approach for patients with clival chordomas involves a combination of gross total resection through endoscopic endonasal surgery (supplemented with an open approach if necessary) and postoperative radiation therapy (RT) [15].

TABLE II: IMMUNOHISTOCHEMICAL PANEL TO AID IN THE DIFFERENTIAL DIAGNOSIS FOR CHORDOMA DEMONSTRATING THE MOST FREQUENT IMMUNOPROFILES [16]

	Cytokeratin	EMA	S 100	Brachyury	Other
Chordoma	Positive	Positive	Positive	Positive	-
Chordoid meningioma	Variable	Variable	Variable	Negative	PR
Chordoid glioma	Variable	Variable	Positive	Negative	GFAP
Metastatic adenocarcinoma	Positive	Positive	Negative	Negative	Sites specific immunohistochemical markers

EMA: epithelial membrane antigen.

PR: progesterone receptor.

GFAP: glial fibrillary acid protein.

#### IV. CONCLUSION

In the context of progressive ophthalmological signs by a sellar and suprasellar lesion, the differential diagnosis should include chordoma along with meningioma, macro adenoma, and metastasis.

A complete and early surgical removal is the first line of treatment.

#### FINANCIAL DISCLOSURE

The authors declared that this study has received no financial support.

#### ETHICAL APPROVAL

Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient. Our institution has exempted ethical approval.

#### CONFLICT OF INTEREST

The authors declare having no conflicts of interest in this article.

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