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Nonenhancing Prepontine Chordoma with Diffusion Restriction Mimicking an Epidermoid Cyst

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Abstract

Intradural prepontine chordoma is an extremely rare entity and depicts benign features with no recurrence and has a good prognosis as compared with bone endogenous chordoma. Radiologically, it may mimic an epidermoid cyst and present an imaging challenge. Here we present a case of a middle adolescent girl who presented with headache along with limb and facial weakness. A nonenhancing, diffusion-restricting, predominantly intradural cystic-appearing mass lesion was seen in the prepontine cistern. Initially, an epidermoid cyst was suspected, but histopathology revealed it to be a chordoma. Destruction of the clivus is important and chordoma should be included in differential diagnosis.

Keywords
- chordoma
- epidermoid cyst
- diffusion restriction

Introduction

Most chordomas are bone endogenous types, typically manifesting as midline tumors with bone involvement and enhancement.[1] Few chordomas are bone exogenous types between the clivus and the dura mater and may show minor bone destruction and transdural transgression.[2] The spheno-occipital locations of chordomas constitute approximately 25 to 30% of all cases.[3] However, in rare instances (3–4%), they may be purely intradural without bone involvement.[4,5] Intradural chordomas are considered benign with complete surgical resection and have a better prognosis than that of typical chordomas.[5] Here, we present a unique case of a nonenhancing, predominantly intradural cystic chordoma showing diffusion restriction, thereby mimicking an epidermoid cyst in an adolescent girl. We will discuss the imaging findings of this atypical case, focusing on the differentiating features from the epidermoid cyst.

Case Presentation

A girl in her middle adolescence, presented with headache, weakness of the left upper and lower limbs for 5 months, and facial weakness for 4 months. On examination, the patient had a full Glasgow coma scale (GCS) score with normal extraocular movements, left facial palsy, temporal and masseter muscle atrophy, loss of corneal reflex on the left side, and loss of left-hand grip. Brain magnetic resonance imaging (MRI) revealed a well-defined lobulated mass lesion in the prepontine cistern (►**Fig. 1**). The lesion was seen causing

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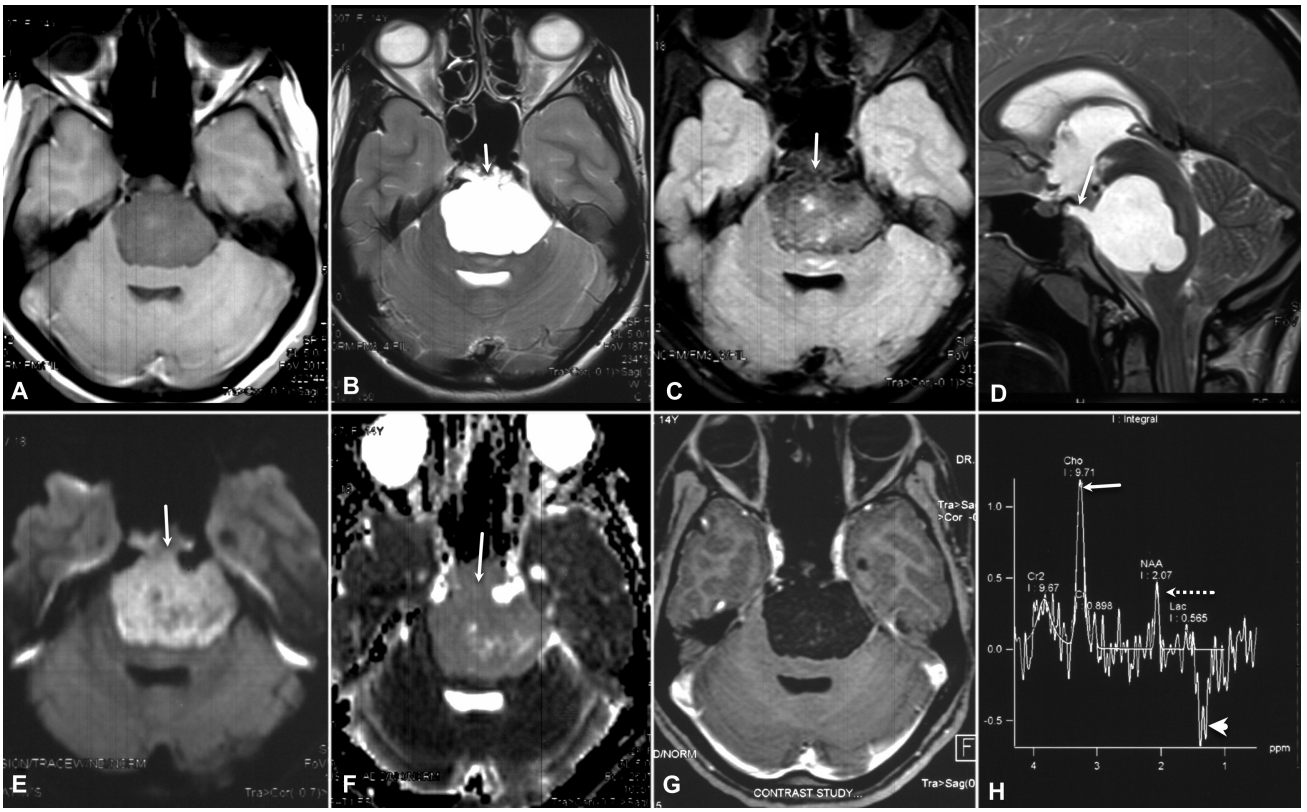

Fig. 1 Magnetic resonance imaging (MRI) of the brain reveals a well-defined lesion in the prepontine cistern with mass effect over the pons resulting in hydrocephalus. The mass is (**A**) iso-hypointense in T1-weighted imaging (T1WI), and (**B**,**D**) hyperintense in T2WI with partial signal suppression in the (**C**) fluid-attenuated inversion recovery (FLAIR) image. A focal, tongue-shaped extension (*arrows* in **B–F**) of the lesion into the sphenoid, which is best appreciated in sagittal T2WI (**D**). The mass is (**E**) hyperintense on diffusion-weighted imaging with corresponding (**F**) hypointensity to cerebrospinal fluid (CSF) on apparent diffusion coefficient (ADC) maps. (**G**) Postcontrast, fat-suppressed axial T1WI shows no enhancement within the lesion. (**H**) Furthermore, magnetic resonance spectroscopy (MRS) acquired at an intermediate echo time (TE) shows elevated choline (*arrow*) and lipid–lactate (*arrowhead*) peaks with a decreased N-acetyl aspartate (NAA) peak (*dotted arrow*).

mass effect over the pons, midbrain, and thalamus, and consequent mild hydrocephalus. The lesion was iso to hypointense on the T1-weighted image (T1WI; ►**Fig. 1a**), and hyperintense on T2-weighted image (T2WI; ►**Fig. 1b**) with incomplete signal suppression on fluid-attenuated inversion recovery (FLAIR; ►**Fig. 1c**). The lesion was hyperintense in diffusion-weighted imaging (►**Fig. 1e**) with a hypointense signal in corresponding apparent diffusion coefficient (ADC) maps (►**Fig. 1f**) suggesting diffusion restriction. On post-contrast-enhanced T1WI (►**Fig. 1g**), the mass did not demonstrate any contrast enhancement. A tongue-shaped extradural extension of the lesion was seen at the level of the upper clivus, into the sphenoid bone (*arrows* in ►**Fig. 1b–f**). MR spectroscopy at intermediate echo time (TE) revealed an increase in choline (Cho), a decrease in N-acetyl aspartate (NAA), and the presence of lipid–lactate within the mass (►**Fig. 1h**). The subsequent brain computed tomography (CT) scan revealed a well-defined intradural hypodense mass without any calcification in the prepontine cistern (►**Fig. 2a**). Based on imaging, a diagnosis of an epidermoid cyst was considered. However, the patient experienced a rapid deterioration in clinical symptoms, along with hydrocephalus, which contradicted the diagnosis of epidermoid cysts, as patients with epidermoids typically do not present with such symptoms.

Treatment and Follow-Up

The lesion was operated on via left frontotemporal craniotomy and the subtemporal approach. The tumor was moderately vascular and could be removed using Cavitron Ultrasonic Surgical Aspirator (CUSA) suction. It was observed to compress the pons and midbrain posteriorly. The fourth, fifth, and sixth cranial nerves and basilar artery were successfully identified and preserved during the procedure. A complete excision of the tumor was achieved. The histopathological analysis diagnosed the tumor as chordoma (►**Fig. 3**). Immunohistochemistry results showed positive cytokeratin, *S-100*, and brachyury expression, while p53 was negative. Integrase interactor 1 (INI-1) expression was retained. The MIB-1 index was determined to be 1%. After careful reassessment of the noncontrast CT (NCCT) bone window, subtle focal erosion of the upper clivus was seen (*arrow* in ►**Fig. 2b**).

The patient was discharged 5 days after the surgery, with no observed complications. One month later, the previously reported headache and lower limb weakness had completely resolved. Mild improvement was noted in the facial muscles. A follow-up MRI scan, conducted 1.5 months later (►**Fig. 2c, d**), revealed the presence of a small residual extradural lesion at the upper clivus, exhibiting diffusion restriction. This finding further supports the likelihood of a chordoma in

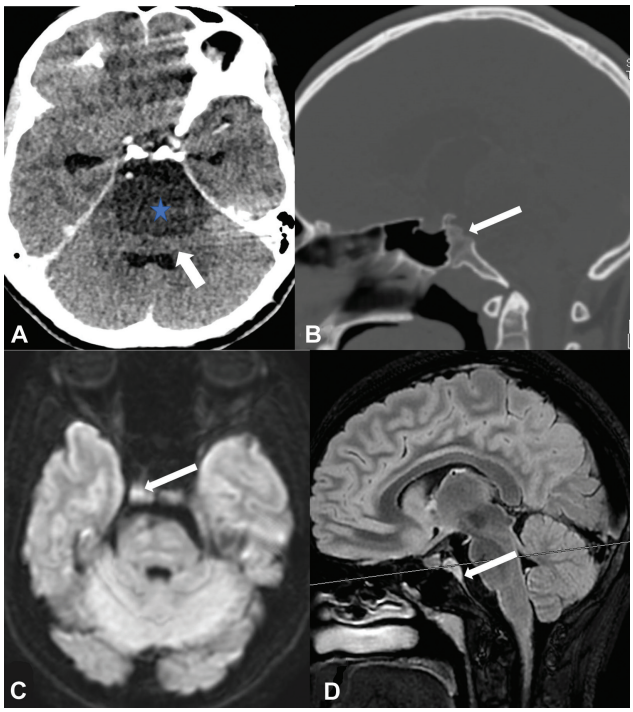

Fig. 2 (**A**) Noncontrast computed tomography (CT) scan of the head shows a well-defined mass lesion in the prepontine space (*), resulting in compression of the pons (*white arrow*). (**B**) The sagittal reconstructed image in bone window setting displays erosion (*arrow*) along the posterior surface of the sphenoclival region. Follow-up magnetic resonance imaging (MRI) after 1.5 months revealed residual extradural lesion between the sphenoclival junction and the dura with (**C**) restricted diffusion (*arrow*) of diffusion-weighted image and (**D**) hyperintense signal (*arrow*) on sagittal fluid-attenuated inversion recovery (FLAIR) image.

that location. As of the current date, the patient remains under continued follow-up.

Discussion

Chordomas are midline tumors and are found along the course of the embryonic notochord.[1] They mostly appear in adults (fourth and fifth decades of life), with a male predilection; they occur rarely among children.[3] However, in the literature, intradural (prepontine) chordoma has been described in an 11-year-old girl who has similar imaging features to our case.[6] It can show off-midline location.[7] Rarely, the aberrant location of the notochord may occur during ossification, and remnants of the notochord may persist into the extradural or intradural spaces.[8] Another theory suggests that the remnant fragments of the notochord in the extradural space may migrate into the intradural space, especially in the setting of early head trauma.[9] This would allow chordoma to grow in intradural locations. Chordomas exhibit myxoid matrix, *S-100*, cytokeratin with newly diagnosed brachyury (T gene product), and galectin-3 (a member of the galactoside-binding protein family) expression patterns as highly sensitive and specific markers in the diagnosis of chordoma.[10] On CT, intracranial chordomas commonly appear as high attenuating masses with extensive osteolytic bone changes.[3] However, in our case, the lesion was intradural and a low attenuating mass with subtle bone erosion. For differential diagnosis of prepontine lesions, ecchordosis physaliphora (EP) can be easily ruled out from epidermoid cyst and intradural cystic chordoma by imaging and clinical features[11] as EP is mostly asymptomatic, incidentally detected, less than 2 cm in size, shows T1 hypointense/T2 hyperintense signal, without contrast enhancement and no diffusion-weighted restriction.[12,13] Chordomas have specific neural symptoms and signs induced by cranial nerve and brainstem compression as evident in this case. Epidermoid cysts have no bone erosion and fewer cranial symptoms. Intradural chordomas have a better prognosis with no recurrence after surgical excision.[14,15] Ito et al[14] reported zero recurrences in 18 intradural chordomas among 17 patients with 5 to 144 months of follow-up after total/subtotal removal. Seventeen patients had an MIB-1 index of less than 5%. However, they remained in the surrounding bone even after complete excision as in our case (►**Fig. 2**). In a study of 28 biopsy-proven cases of chordoma, the majority of the patients (64%) demonstrated either no/minimal (11/28, 39%) or mild enhancement (7/28, 25%), and the remaining cases showed moderate (4/28, 14%) and marked enhancement (6/28, 21%).[16] They may display honeycomb-like enhancement.[17] The lack of enhancement is due to the mucinous component. A review of the literature of 11 patients by Guadalupi et al[18] demonstrated diffusion restriction in 4 patients.[17,19,20] Poorly differentiated chordomas exhibit lower signal intensity on T2WI and ADC values ($0.875 \pm 0.100 \times 10^{-3}$ mm^2/s), reflecting increased tumor cellularity. The arrangement of physaliphorous cells within myxoid stroma may impede free extracellular water motion and result in diffusion restriction. Therefore, reaching a definitive imaging diagnosis without histopathological correlation can pose a challenge.

Take-Home Messages

- Chordoma should be considered in the differential diagnosis of cystic lesions of the prepontine region like epidermoid cysts as both are nonenhancing and show diffusion restrictions.
- A short clinical history, significant compression on the brainstem, hydrocephalus, multiple cranial nerve involvement, and extradural extension favor the diagnosis of chordoma.
- We should look carefully for erosion of the clivus or spheno-occipital region on NCCT, as intradural chordomas may have minimal bony erosion.

Authors' Contributions

B.D.C., S.A., S.D., E.S., S.J., A.G., L.J.D.S., and M.S. contributed to the acquisition, analysis, conception, design, and drafting of the work. B.D.C. and E.S., along with A.G. and L.J.D.S., contributed to the final draft, revisions, upload, and submission of the final revised work. All the authors have agreed both to be personally accountable for their contributions and to ensure that questions related to the accuracy or integrity of any part of the work, even ones in

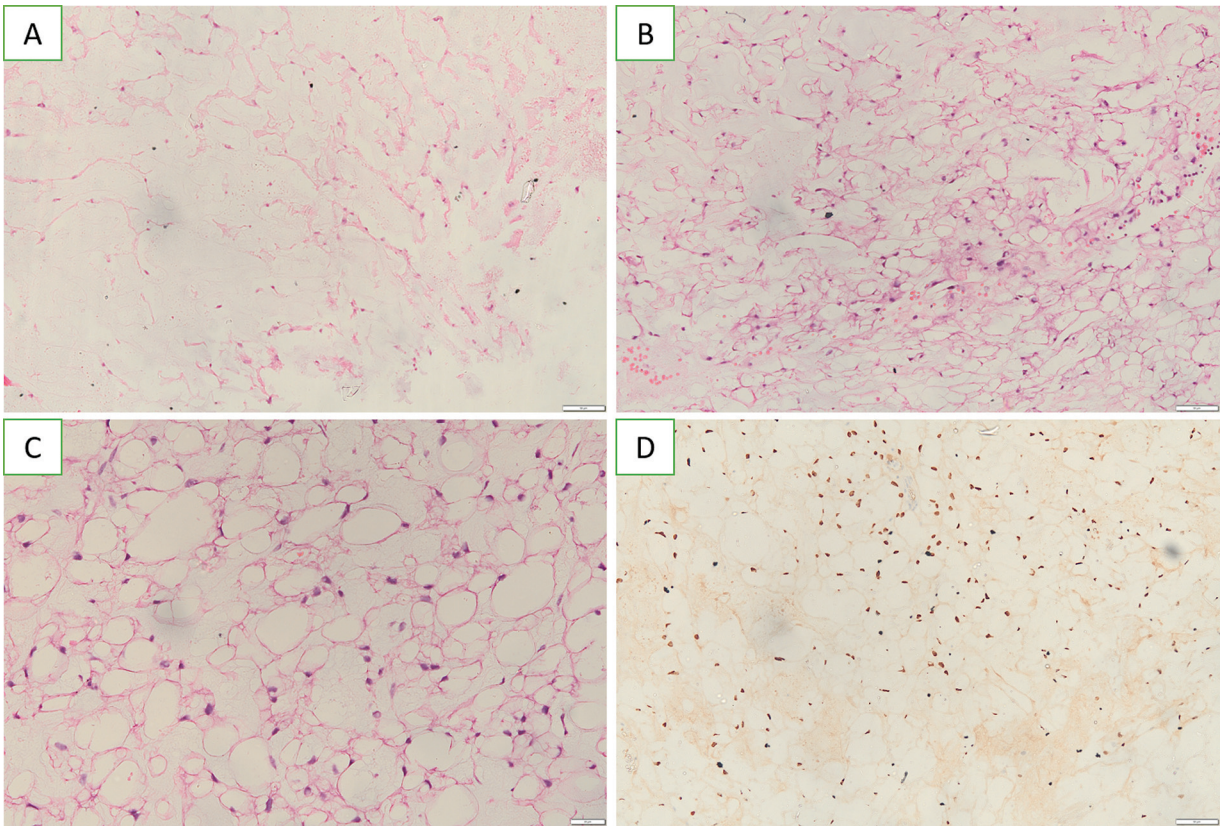

Fig. 3 Histopathological examination. (**A**) The lesion's frozen section (10x, hematoxylin and eosin [H&E]) shows a sparse cellular tumor in a myxoid background. (**B**) Biopsy from the lesion (10x, H&E) shows a sparsely cellular tumor in a myxoid background, characterized by tumor cells with clear cytoplasm. (**C**) At higher magnification (20x, H&E), individual tumor cells with clear and bubbly cytoplasm, known as physaliphorous cells, are observed. (**D**) Immunohistochemistry (20x) for Brachyury reveals strong nuclear positivity.

which one was not personally involved, are appropriately investigated and resolved, and the resolution documented in the literature. All the authors have read and approved the manuscript.

Patients' Consent

Written informed consent to publish this information was obtained from study participants.

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None.

Conflict of Interest

None declared.

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