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Isolated Intradural Prepontine Chordoma Presenting with Imaging Features of Epidermoid Cyst

Cordoma pré-pontino intradural isolado apresentando características de imagem de cisto epidermóide

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Abstract

Chordoma is an erratic aggressive tumor of the brain that typically involves the clivus. The majority of the clivus chordomas reside in the extradural space. Here, we report a unique case of chordoma arising from the prepontine intradural space without bony involvement and presenting with radiological features typical of an epidermoid cyst on magnetic resonance imaging.

Keywords

- cyst
- epidermoid
- chordoma
- clivus
- differential diagnosis
- MRI
- prepontine

Introduction

Chordoma, which is as an erratic, aggressive tumor with slow growth, is thought to be derived from the embryonic notochord remnants, as a cartilage-like structure shaped like a rod serving a scaffold for forming the spinal column.[1] They usually occur in adults with a peak occurrence within the age 50 to 60 years.[2] Approximately 35 to 40% of these tumors have an intracranial localization, where they typically involve the clivus. The majority of the clivus chordomas reside in the extradural space.[3] Here, we report a unique case of chordoma arising from the prepontine intradural space without bony involvement and presenting with radiological features typical of an epidermoid cyst (EC) on magnetic resonance imaging (MRI).

Case Report

History

A 24-year-old patient presented to our clinic with headache complaints. The patient's headache had begun 6 months before admission, increased progressively, aggravated by coughing and laughing and was sometimes accompanied by dizziness. The patient reported no other associated symptoms, and no specific neurological deficits were found in this case.

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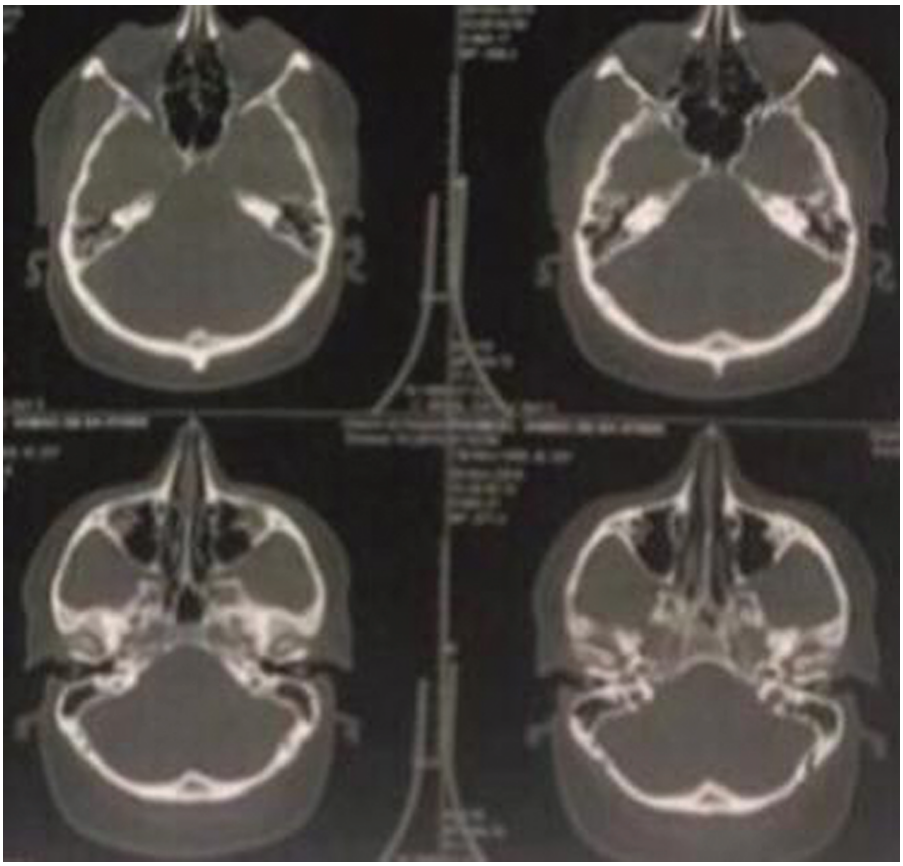

Fig. 1 Brain magnetic resonance imaging.

Radiological Imaging

Brain MRI with and without contrast showed a well-defined hypodense mass about $34 \times 25 \times 36$ mm in prepontine cistern with mass effect on the pons. The tumor had heterogenous high T2 and flair signal intensity. Areas of high T1 intensity were seen. No significant enhancement is seen in areas with fluid restriction is noted in diffusion-weighted imaging (DWI). The finding is suggestive of an epidermoid cyst (►**Fig. 1**).

A brain computed tomography (CT) scan showed a well-defined hypodense mass about $34 \times 25 \times 36$ mm in the prepontine cistern with mass effect on the pons; no calcification and no bone erosion were seen and the clivus was normal (►**Fig. 2**).

Operation, Histological Examination, and Postoperative Course

The patient underwent prepontine mass resection with a retrosigmoid mass resection surgery in a sitting position. After mass removal, tissue samples were sent to the laboratory for pathological examination, which revealed that the tumoral tissue was composed of lobules of cell beating round nuclei and abundant clear or lightly eosinophilic cytoplasm. Areas of chondroid differentiation cells showed cytokeratin, epithelial membrane antigen (EMA), s100 positive, and glial fibrillary acidic protein (GFAP) negative, which are compatible with chordoma (►**Fig. 3**).

Discussion

Chordomas are erratic tumors of the axial skeleton, and they happen mostly in the sacrum and at the base of the skull. It is thought that chordomas arise from fetal notochord remnants remaining in the axial skeleton during life and probably undergoing malignant transformation into chordoma at any point in life.[4,5] Macroscopically, chordomas seem to be a white-gray lobulated, gelatinous, soft tumor with dens fibrous trabeculae.[6] Mucoid material, necrotic areas, current, old hemorrhages, and sequestration and calcification of bone fragments are observed in the tumor.[7] An unfinished pseudocapsule with pressured nearby tissue imitating a true capsule surround the soft tissue frequently.[8] A heterogeneous cytology is displayed by all chordoma subtypes. The large cells containing prominent solitary or multiple vacuoles are the main cell types, also known as physaliphorous cells. These vacuoles are rich in mucopolysaccharides.[9–11] On non-contrast CT, chordomas are characteristically found as well-constrained, heterogeneous, hypoattenuating lesions with widespread lytic bone destruction.[6] The tumor bulk is typically hyperattenuating based on the nearby neuronal axis.[12] Distinguishing between intratumoral calcifications is difficult that are representative of the chordoma's chondroid variant,[13] and appropriated fragments of the damaged clival bone.[6] In the late 1980s and early 1990s, the MRI features of chordoma with the normal MRI sequences were described well.[6] Chordoma is able to impose inconstant signal intensity on T1, typically mostly low-to-

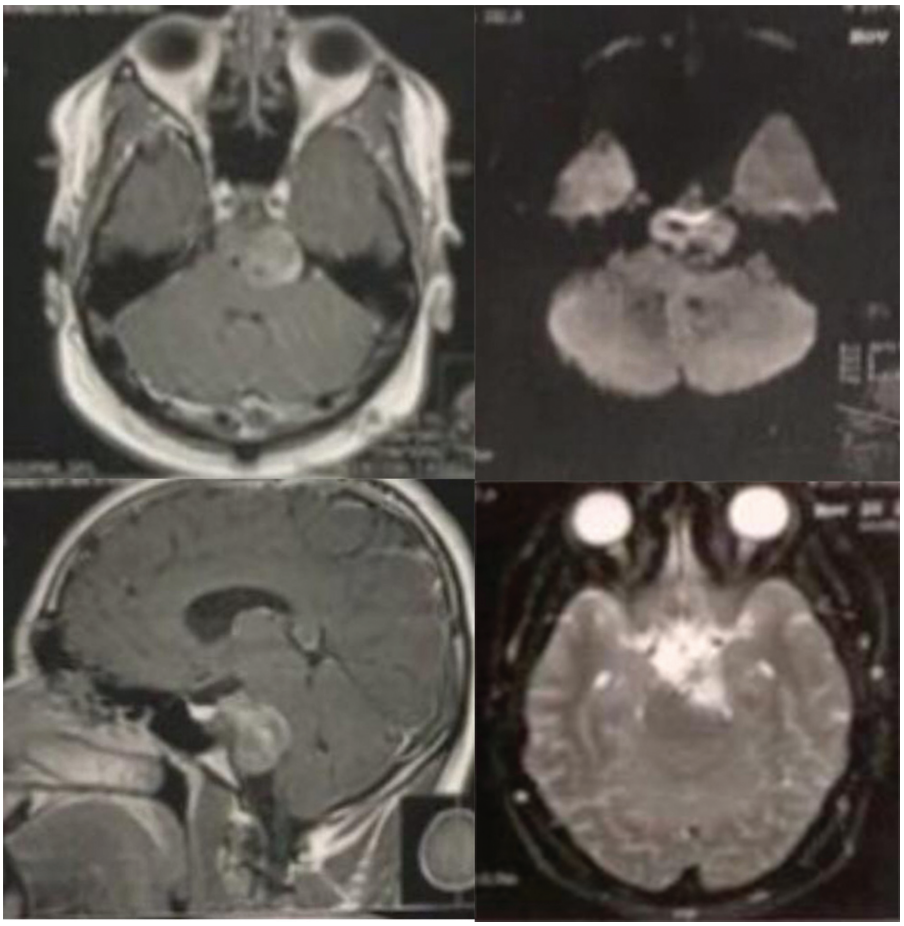

Fig. 2 Brain computed tomography scan.

intermediate signal strength, occasionally with minor hyperintensity foci, related to hemorrhage or mucus.[14] High T2 signal strength with heterogeneous hypointensity is observed in classic chordoma, probably related to hemorrhage, mucous, and calcification as well. The existence of calcification or hemorrhagic foci can be observed in susceptibility-weighted imaging (SWI), which are gradient echo images indicating vulnerability artifacts.[14] It is also possible to find low signal intensity septations probably correlated with areas of cartilage or necrosis observed in histology. Various imaging features may be shown by weakly distinguished chordoma.[15] Yeom et al. indicated hypo intensity on T2-weighted images in 3 weakly distinguished chordoma. Nevertheless, there are no studies in the literature that investigating greater weakly distinguished chordoma cohorts. Chordoma characteristically indicate fair-to-high gadolinium contrast improvement with honeycomb appearance, with linear non-improvement areas.[16] This is probably clarified with the areas of necrosis, connective tissue, or cartilage in the tumor at histology. In delineating the clival chordoma, fat suppression imaging with suppression of the clivus fatty bone marrow may be effective.[12] Epidermoid cysts are among the most prevalent benign intracranial cystic lesions commonly found. Through current imaging technology, their recognition and the radiologic diagnosis are nearly always easily possible.[17] As a result of protein, calcium, lipid, and hemosiderin content[18] together with occasional calcification (more in dermoids), the tumor seems to be isodense or hypodense or occasionally spontaneous hyperdense on CT scan. Magnetic resonance imaging scan is the modality of choice for diagnosing. The lesion is hyperintense on T2-weighted, hypointense on T1-weighted and FLAIR, with hyperintense constraint on diffusion-weighted imaging (DWI) [19]and without any contrast enhancement.[18] When contrast improvement occurs, it is typically at the tumor's margins.[18] In intrinsic lesions, the absence of the tumor edema distinguishes from the gliomas. In extrinsic locations, DWI is useful to differentiate from the arachnoid cyst and abscess.[20,21] Diffusion-weighted imaging is useful to know the remnant by its restriction, differentiation from abscess, and arachnoid cyst.[22] There are very few reports in the English literature on cases that were clinically followed as EC, yet found to be as chordoma in the pathological examination. Such a case was described in 2004, but, in this case, the lesion demonstrated partial bony erosion.[23] The lesion showed hyperintensity on T2-weighted MRI, hypointensity on T1-weighted MRI and hyperintensity on DWI with no contrast enhancement. The tumor was surgically removed and despite a suspicion of EC, the pathological examination revealed a chordoma. In 2018, a similar case of a prepontine mass was reported. A 35-year-old Caucasian woman was admitted to an outpatient clinic with complaints of progressive dizziness, ataxia, and diplopia. The patient's lesion was previously diagnosed as a prepontine EC. The lesion

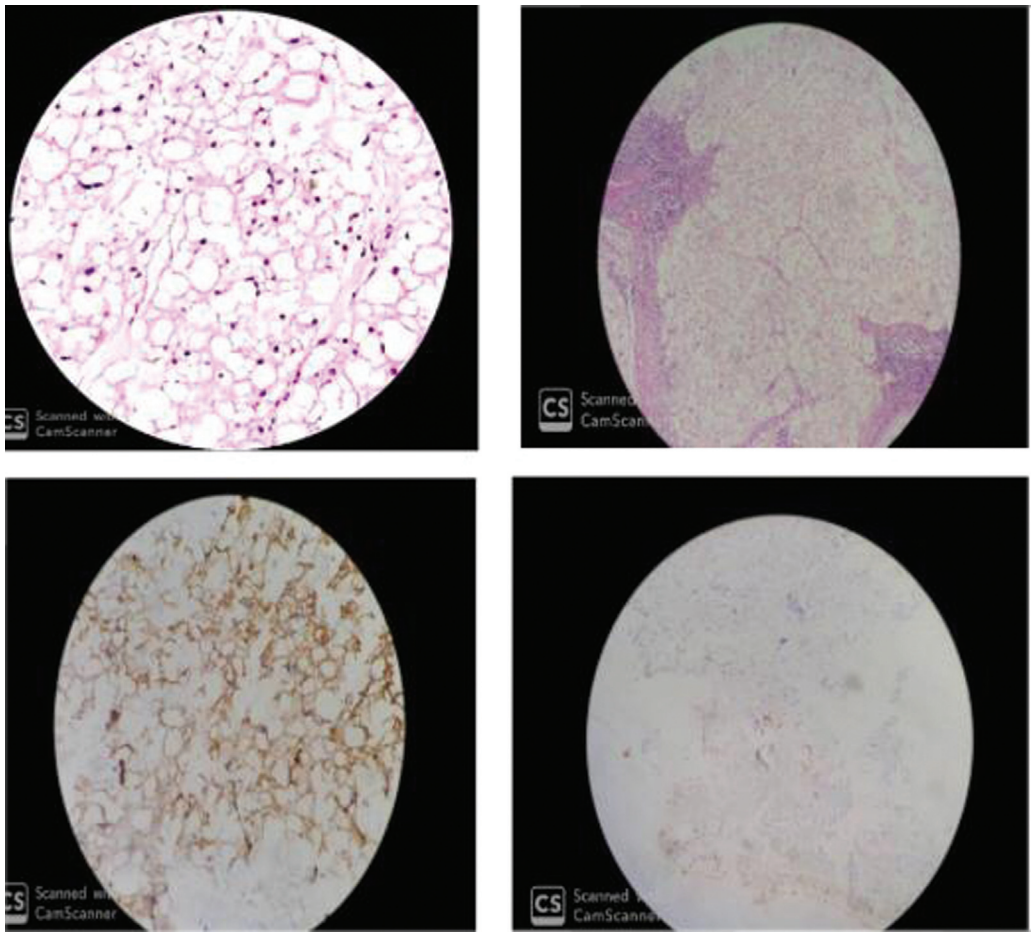

Fig. 3 Microscopical pathology of the tumor.

seemed as hypointense in T1 and hyperintense on T2-weighted images. The lesion also indicated hyperintensity on DWI and hypointensity on apparent diffusion coefficient (ADC) images. The lesion did not demonstrate any contrast enhancement. No obvious destruction of the clivus or bony erosion with conserved normal fatty marrow on MRI. The patient was operated, and a gray-purple soft tumor tissue was noted perioperatively, having neither macroscopical features of a typical EC nor a vascular pattern typical of chordomas. Unexpectedly, the frozen section analysis reported a chordoma. Similar to the frozen section, the pathological analysis also revealed a chordoma. The patient was discharged on the 5th postoperative day, following radiotherapy referral for the residual tumor. The pathological analyses revealed the following features.[3] In 2017, A 44-year-old female visited a clinic with progressively worsening right facial dysesthesia, which had persisted for 10 years. The MRI indicated a $32 \times 30 \times 16$ mm tumor in the prepontine-to-interpeduncular cistern, which was hypointense on T1-weighted images (T1WIs), isointense on fluid-attenuated inversion recovery (FLAIR) sequences, hyperintense on T2WIs, iso-to-hyperintense on DWI, and hyperintense on ADC map ($1.2–1.6 \times 10–3$ mm^2/s). No deceptive contrast improvement exists on postcontrast T1WIs. In consideration of these imaging properties, we supposed that the tumor was an EC according the DW results. The tumor was light-yellowish, transparent, jelled, and cystic after removing the mass, unlike an EC. The pathology was compatible with ecchordosis physaliphora. Follwing the surgery, the symptoms of the patient were enhanced, and she was discharged on the postoperative day.[24] In 2014, a similar case of prepontine mass was reported. The MRI examination of a 15-year old girl with complaints of headache revealed a well-defined, lobulated prepontine mass that was profoundly hypointense on T1- and FLAIR sequences and hyperintense on T2W, suggesting a lesion with fluid content. Small scattered areas of hyperintensity were evident on T1 and FLAIR sequences, presumed as likely signs of a hemorrhage. The clivus was intact with no bone erosion. Peroperatively, the tumor was greyish and semi-solid in texture, and the cranial nerves were stretched and encased by the tumor. Pathological analysis revealed a chordoma. The patient required a second excision 13 months later as the residual tumor expanded in size; the patient also received 3 months of radiotherapy. The most recent MRI performed two years postsurgery showed no tumor progression.[25] In 2012, another prepontine chordoma was reported, which was initially suspected as a benign EC. The CT examination of a 32-year-old male with complaints of dysarthria showed a hypodense pontine mass with no contrast improvement or clival bony erosion. The MRI revealed a multi lobulated contour bulging lesion in the prepontine cistern and invagination into

the pontine parenchyma, which somewhat covered the mid basilar artery. The mass was hyperintense on T2-weighted MRI and hypointense on T1. Most of the mass indicated no post-contrast improvement, excluding some minor dominant multiple reticulonodular heterogenous improvements. On DWI, a heterogenous and mildly incremented signal intensity was indicated by this mass. The postoperative pathological analysis revealed a chordoma, and centrally enhanced foci were defined as profound vascularity.[26]

Conclusion

The chordoma case reported here had radiologic features similar to those of ECs. In cases in which there is a tumor near the clivus, chordoma should always be considered as a differential diagnosis, even without the presence of bone erosion.

Conflict of Interests
The authors have no conflict of interests to declare.

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