



Imaging Findings of a Nonenhancing Intradural Paramedian Chordoma Mimicking an Epidermoid Cyst

표피양낭종과 유사한 조영증강되지 않는 방정중위치의 경막 내 척삭종

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Intracranial chordoma is a rare tumor, originating from embryonic remnants of the primitive notochord. It typically appears as an enhancing extradural midline tumor with bone involvement. We introduce a rare case of a 27-year-old male who had a nonenhancing intradural chordoma showing paramedian location, involving the left cavernous sinus, Meckel's cave, and prepontine cistern. The pathologic diagnosis was confirmed as an intradural chordoma. The imaging findings of this unusual case of a nonenhancing intradural paramedian chordoma will be presented with the differential diagnosis focused on the epidermoid cyst.

Index terms

Chordoma
Cavernous Sinus
Epidermoid Cyst
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INTRODUCTION

Intracranial chordomas originate from the remnants of the primitive notochord and account for approximately 1% of all intracranial tumors (1, 2). Most intracranial chordomas are extradural tumors that arise from the sphenooctipital synchondrosis in the clivus (2). There are only few reports about intradural chordoma without bone involvement (2). Moreover, only few cases of chordomas reported absence of contrast enhancement (3). We introduce a very rare case of a nonenhancing intradural chordoma, which showed a paramidline location without bone involvement.

CASE REPORT

A 27-year-old male visited the neurosurgery clinic due to recurrent syncope for 6 years. He had a history of several syncope events, especially in case of stress or after urination. His motor

or sensory functions were normal. There was no abnormal finding on electroencephalography, electrocardiogram, and orthostatic blood pressure tests.

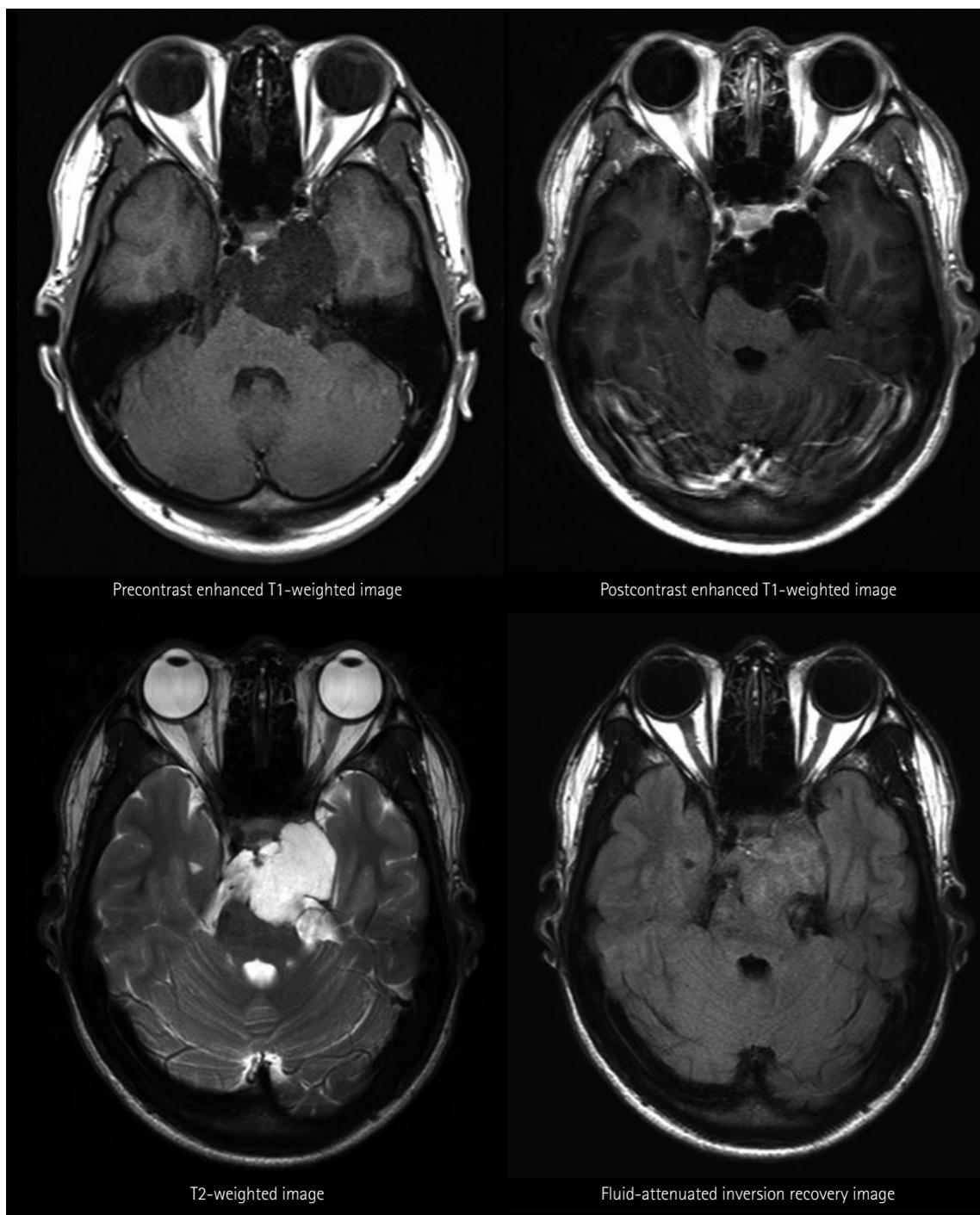
On brain magnetic resonance imaging (MRI), there was an approximately 4.6 × 3.8 × 3.4 cm sized lobulated intradural mass involving the left cavernous sinus and left Meckel's cave, extending to the prepontine cistern, causing deviation of left fifth cranial nerve. The mass showed low signal intensity on T1-weighted images (T1WI), high signal intensity on T2-weighted images (T2WI), and iso- to high signal intensity on fluid-attenuated inversion recovery images (Fig. 1A). On postcontrast enhanced T1WI, the mass did not demonstrate any contrast enhancement (Fig. 1A). On diffusion weighted images (DWI), the mass showed high signal intensity with slightly low apparent diffusion coefficient (ADC) value on ADC map (Fig. 1B), which was approximately 1227.01 × 10⁻⁶ mm²/s in average.

On the subsequent brain computed tomography (CT), the mass showed low attenuation without evidence of bony erosion

or destruction of the adjacent skull (Fig. 1C). Additionally, on transfemoral cerebral angiography, there was no feeding vessel. Based on these findings, the preoperative radiological impres-

sion was an epidermoid cyst.

The patient underwent subtotal tumor excision with left parietotemporal craniotomy. The tumor was consisted of vacuolat-



A

Fig. 1. A 27-year-old man with a nonenhancing intradural paramedian chordoma.

A. The mass shows low signal intensity on precontrast enhanced T1-weighted images, high signal intensity on T2-weighted images, and iso- to high signal intensity on fluid-attenuated inversion recovery images. The mass does not show any enhancement on postcontrast enhanced T1-weighted images.

ed cells with abundant myxoid stroma and physaliferous appearance on hematoxylin and eosin stain (Fig.1D). The pathological diagnosis was confirmed as chordoma with positive results for pancytokeratin, S-100, epithelial membrane antigen, and Ki-67 (markers of cell proliferation; MIB-1) (Fig.1E). There was no evidence of necrosis in the tumor.

After the surgery, the patient complained of impairment of lateral gaze in left eye with diplopia. The patient was discharged

with subsequent follow-up at the outpatient clinic for the residual tumor at the left cavernous sinus, Meckel's cave, and prepontine cistern.

DISCUSSION

Intracranial chordomas, arising from the remnant of primitive notochord, can occur at any site along the course of the

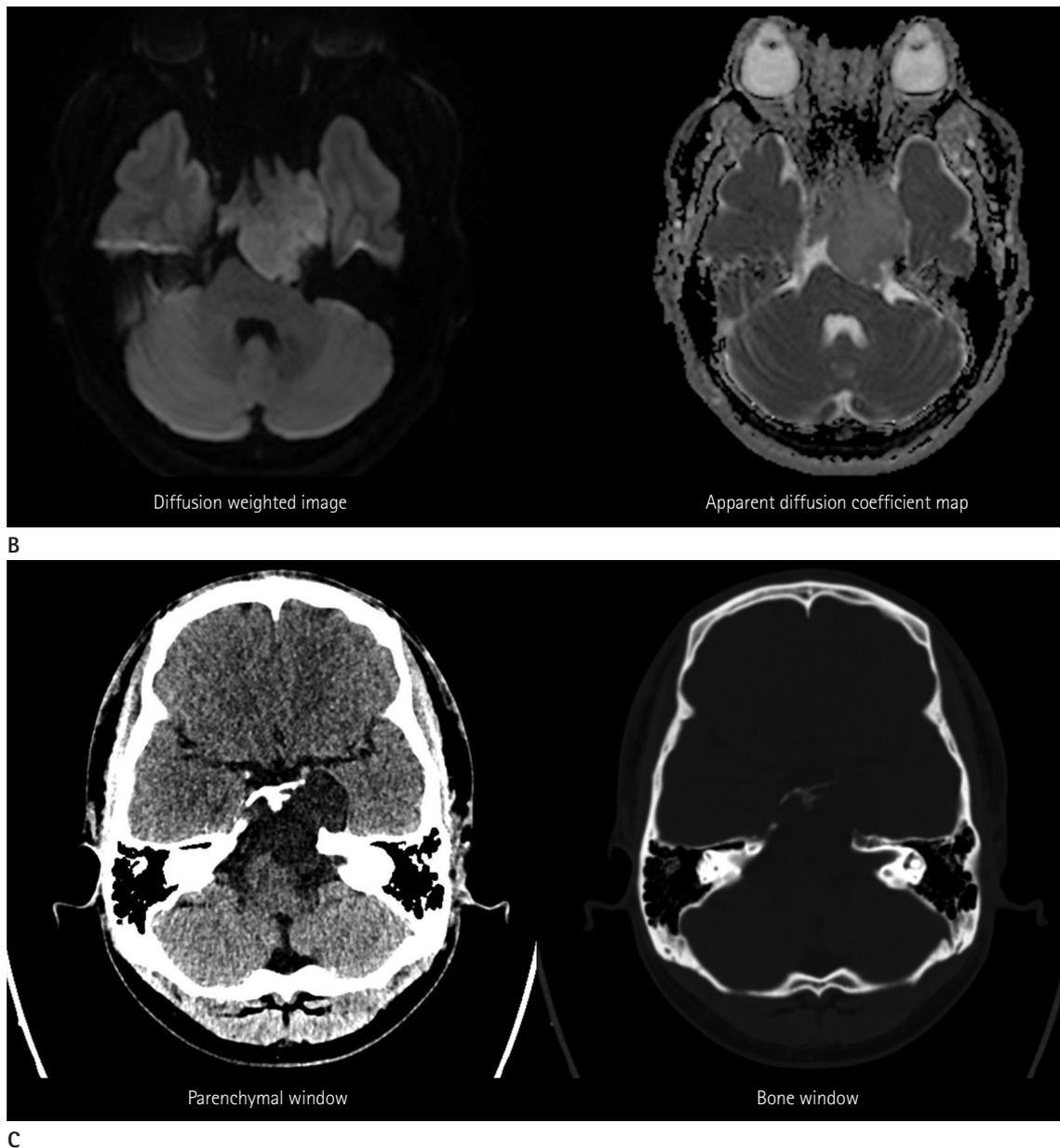
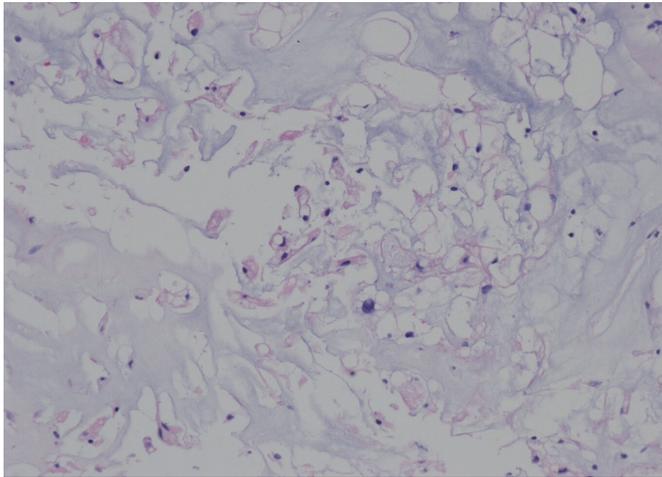


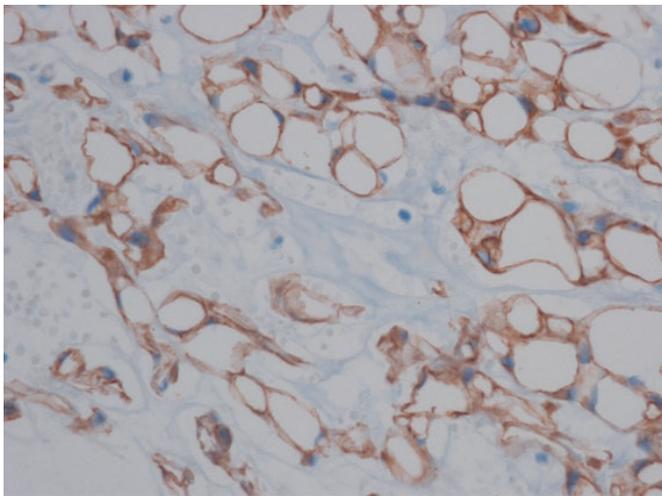
Fig. 1. A 27-year-old man with a nonenhancing intradural paramedian chordoma.

B. On diffusion weighted images, the mass demonstrates high signal intensity with slightly low apparent diffusion coefficient value on apparent diffusion coefficient map.

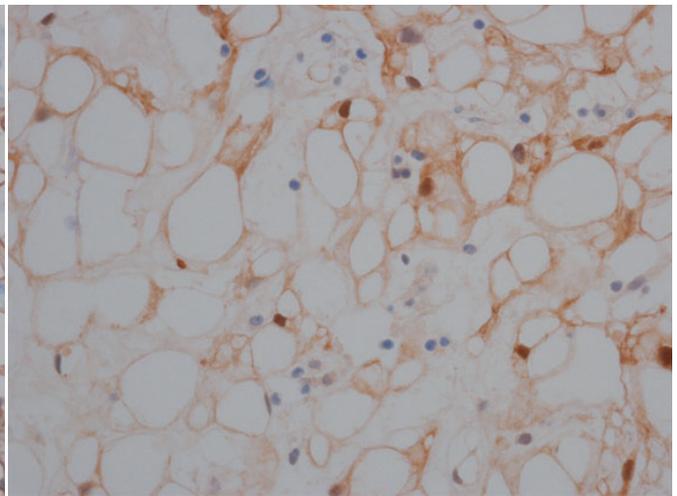
C. On brain computed tomography, a multilobular low attenuating mass is noted at the left cavernous sinus and left Meckel's cave. There is no evidence of bone erosion, demonstrating on bone window images.



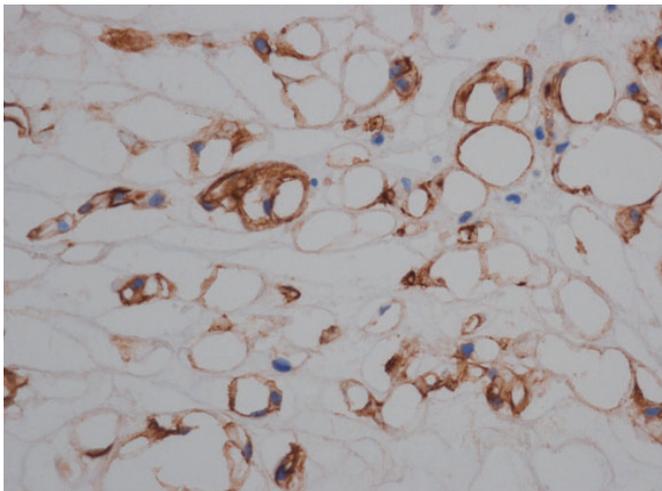
D



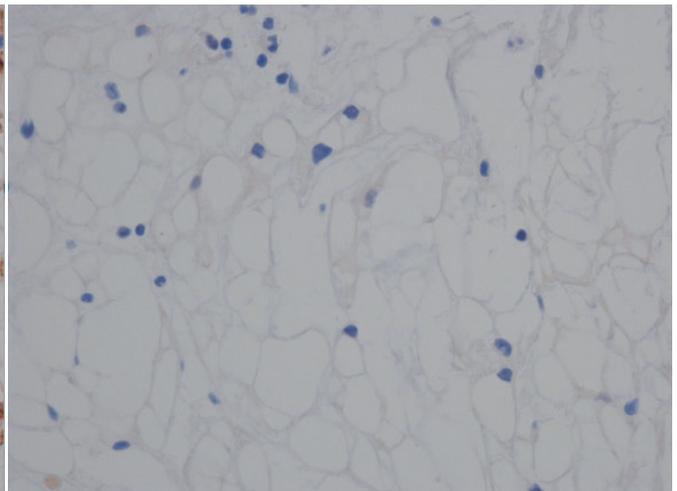
Pancytokeratin



S-100



EMA



Ki-67 (MIB-1)

E

Fig. 1. A 27-year-old man with a nonenhancing intradural paramedian chordoma.

D. On histologic specimen, the tumor contains physaliferous tumor cells with abundant myxoid stroma. There is no evidence of necrosis (Hematoxylin and eosin stain, $\times 200$).

E. The cells were positive in immunohistochemistry staining for pancytokeratin, S-100, EMA, and Ki-67 (MIB-1) ($\times 400$).

EMA = epithelial membrane antigen, MIB-1 = markers of cell proliferation

embryonic notochord (4). However, most studies reported that chordomas are generally located in the midline, and only few lesions showed an off-midline location (2, 5). Paramedian chordomas are commonly located in the petrous apex and Meckel's cave. A previous study reported that 4 of 28 chordomas had a paramedian location, especially at the petrooccipital junction which is a common location for chondrosarcomas (5).

On CT images, intracranial chordoma is commonly demonstrated as a high attenuating mass with extensive osteolytic bone change (1). However, in our case, the tumor was a low attenuating mass without bone destruction.

Because chordomas are composed of translucent cells containing mucoid fluid and a large intracytoplasmic vacuole, they generally show intermediate or low signal intensity on T1WI and high signal intensity on T2WI. On postcontrast enhanced T1WI, chordomas typically show moderate to marked enhancement and only few cases of chordomas have been reported to demonstrate absent or mild enhancement (1, 6). In our case, there was no contrast enhancement in the tumor. Mild or absent enhancement likely represents necrosis or a large amount of mucinous material in the tumor (1). There was no evidence of necrosis in the pathology slide in our case, thus the large lobules without septa formation within the tumor may have been the cause of nonenhancement.

Considering the overall imaging findings of our case, epidermoid cyst should be included in the differential diagnosis. Intracranial epidermoid cyst is a rare congenital neoplasm that constitute about 1% of all intracranial tumors and located mainly in the cerebellopontine angle (7). The most common site of epidermoid cysts is the cerebellopontine angle, followed by the parasellar area, although rare cases with involvement of Meckel's cave and cavernous sinus have been reported (8, 9). It commonly shows low T1 signal intensity and high T2 signal intensity, without contrast enhancement. The high signal intensity on DWI is attributed to the T2 shine-through effect, showing no diffusion restriction on the ADC map (7). The mean ADC value of epidermoid cysts was reported as $1197 \times 10^{-6} \text{ mm}^2/\text{s}$ in a previous report (7). On the other hand, classic chordomas showed a mean ADC value of $1474 \pm 117 \times 10^{-6} \text{ mm}^2/\text{s}$ (10). In our case with a mean ADC value of $1227.01 \times 10^{-6} \text{ mm}^2/\text{s}$, the ADC value does not provide useful information for differential diagnosis.

In conclusion, we report a rare case of a nonenhancing intradural paramedian chordoma. Epidermoid cyst should be included as a differential diagnosis, and on routine MRI it is diagnostically challenging to make a final diagnosis.

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표피양낭종과 유사한 조영증강되지 않는 방정중위치의 경막 내 척삭종

김민정 · 박예원* · 임수미

두개 내 척삭종은 원시 척삭에서 기원하는 드문 종양이다. 이는 주로 조영증강하는 경막 외 정중선 종양으로 발현하며 인접한 뼈 구조를 침범하는 형태로 발현한다. 본 증례에서는 정중선이 아닌 좌측 해면정맥동, 메켈 공간, 그리고 전교뇌수조에 위치하는 드문 형태로 비조영증강 경막 내 척삭종이 발현하였다. 수술 후 병리학적 최종 진단에서는 경막 내 척삭종으로 확인되었다. 따라서, 본 증례 보고를 통해 비전형적인 양상의 비조영증강 방정중위치의 경막 내 척삭종의 영상 소견을 소개하고 표피양낭종과의 감별진단에 초점을 두고자 한다.

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