

# A Case of Ecchordosis Physaliphora Presenting with an Intratumoral Hemorrhage

## Tümör İçi Kanamayla Prezente Olan Ecchordosis Physaliphora Olgusu

### ABSTRACT

Ecchordosis physaliphora is a rare congenital, benign, hamartomatous, retroclival mass derived from notochordal tissue that is typically located intradurally in the prepontine cistern. Ecchordosis physaliphora is usually asymptomatic. In rare cases, ecchordosis physaliphora can be symptomatic due to tumor expansion and compression of the surrounding structures and extratumoral hemorrhage. To our knowledge, ecchordosis physaliphora associated with intratumoral hemorrhage and vasogenic edema has not been previously described. We present a case of 22-year-old man who presented with headache and confusion. MR imaging and CT revealed intracranial ecchordosis physaliphora associated with intratumoral hemorrhage and vasogenic edema. The neurological findings resolved completely after medical therapy.

**KEY WORDS:** Ecchordosis physaliphora, MR, Osseous stalk, CT

### ÖZ

Ecchordosis physaliphora notokord dokusundan kaynaklanan nadir, konjenital, benign, hamartamatöz retroklival kitlelerdir. Tipik olarak prepontin sisternada intradural yerleşim gösterirler. Ecchordosis physaliphora genellikle asemptomatiktir. Nadiren tümörün ekspansiyonuyla çevre dokulara bası ya da kanamalara bağlı semptomatik olan olgular tanımlanmıştır. Bildiğimiz kadarıyla şimdiye kadar, tümör içi kanama ve vazojenik ödemin eşlik ettiği ecchordosis physaliphora olgusu bildirilmemiştir. Biz, baş ağrısı ve konfüzyonla başvuran ve tümör içi kanama ve vazojenik ödemin eşlik ettiği intrakraniyal Ecchordosis physaliphoralı 22 yaşında erkek olgunun BT ve MR görüntüleme bulgularını sunduk. Medikal tedaviyi takiben nörolojik bulgular tamamen düzeldi.

**ANAHTAR SOZCÜKLER:** Ecchordosis physaliphora, MR, Kemik stalk, CT

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## INTRODUCTION

Ectopic notochordosis physaliphora (EP) is a rare intracranial extra-axial mass derived from notochordal tissue that is typically located intradurally in the prepontine cistern and attached to the dorsal wall of the clivus (12). EP is usually asymptomatic, and only a few studies have reported associated symptoms due to tumor expansion and compression of the surrounding structures and extratumoral hemorrhage (1,2,4,5,6,7,9,10). To our knowledge, EP associated with intratumoral hemorrhage has not been previously described. We report a case of headache and confusion in a 22-year-old man in which the diagnosis was based on CT and MR imaging.

## CASE REPORT

A 22-year-old man was admitted to our hospital for a confused state after taking 1000 mg aspirin for a headache. On admission, neurological examination showed right supranuclear facial nerve palsy, dysarthria, and right hemiparesis with a positive Babinski sign. He had truncal ataxia. The patient underwent CT, MRI, MR angiography, and digital subtraction angiography.

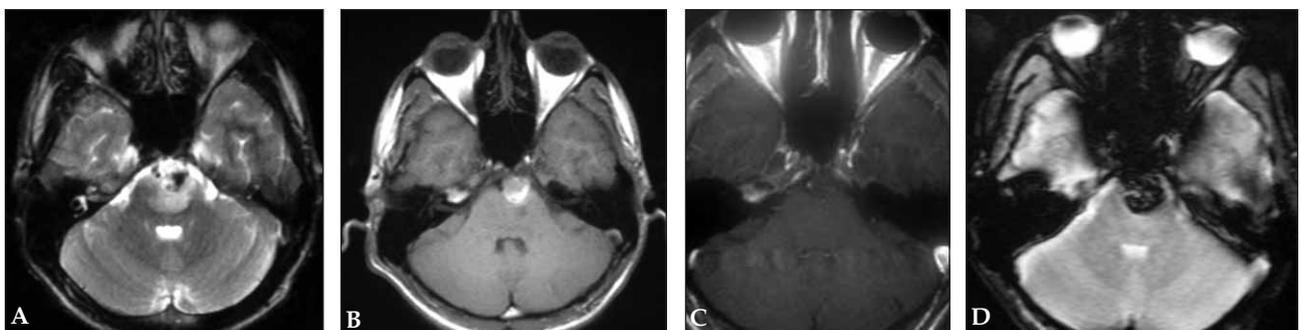
Initial MR imaging of the brain showed an intradural, well circumscribed, extra-axial mass located in the prepontine cistern that was causing a mass effect on the basilar trunk and pons. The mass appeared hyperintense on T2-weighted images and hypointense on T1-weighted images without contrast enhancement and was 2 cm in diameter. The mass had focal areas of hyperintensity on T1-weighted images and hypointensity on T2-weighted images, which likely represented hemorrhage, and restricted diffusion on diffusion-weighted images. T2 gradient echo confirmed an intratumoral

hemorrhage (Figure 1A,B,C,D). CT showed a hyperdense lesion and osseous stalk at the dorsal wall of the clivus, but did not indicate any intratumoral calcification or bone destruction (Figure 2A,B). Cerebral MR angiography and digital subtraction angiography showed displacement of the basilar artery by the mass. No feeding vessels or draining veins were observed that would have suggested a vascular malformation, or any tumor stain. The patient was given mannitol for the brain stem edema. Three days after his admission, his neurological signs resolved except for mild truncal ataxia. A 7-day follow-up MR showed regression of the intratumoral hemorrhage. A 4-week follow-up MR demonstrated resolution of the intratumoral hemorrhage and vasogenic edema. The size of the tumor and resultant compression regressed (Figure 3A,B,C). A chronic lacunar infarct was seen in the left pons. The neurological findings were completely resolved.

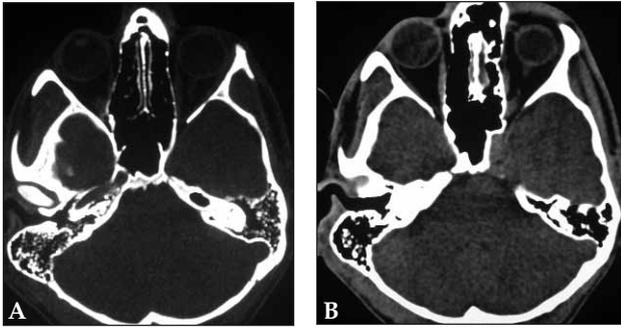
## DISCUSSION

EP is a rare, congenital, benign, hamartomatous lesion arising from an ectopic notochordal remnant. The notochord develops in humans during the third week of embryonic life and persists in adults as the nucleus pulposus of the intervertebral disks. Ectopic notochordal rests can be seen along the midline of the craniospinal axis, from the dorsum sellae to the sacrococcygeal region (12). Intracranial EP is most often located in the prepontine cistern and can occur in any combination of extradural, intradural, and subarachnoid spaces along the clivus and dorsum sellae. Intradurally, EP is usually connected by a thin pedicle with similar nests of notochordal cells within the clivus (7,11).

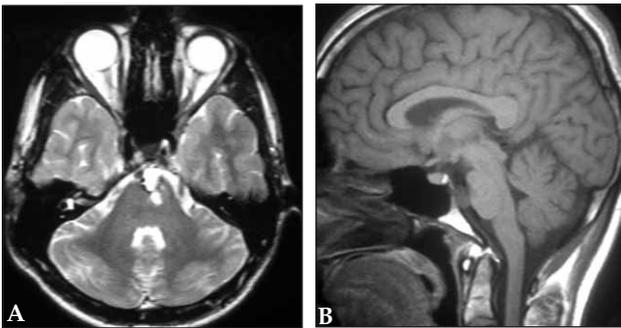
Recognition of the imaging features of EP is helpful in suggesting the diagnosis and



**Figure 1:** Axial T2-weighted (A) and pre-contrast T1-weighted (B) images showing a mass with focal intratumoral hemorrhage in the prepontine cistern. Post-contrast T1-weighted (C) image shows no contrast enhancement. A gradient echo image (D) confirmed the intratumoral hemorrhage.



**Figure 2:** Axial CT image bone (A) and soft-tissue (B) windows showing a hyperdense mass in the left prepontine cistern and osseous stalk at the clivus.



**Figure 3:** Four-week follow-up axial T2-weighted (A), sagittal pre-contrast T1-weighted (B) and gradient echo (C) MR images demonstrated resolution of the intratumoral hemorrhage, decreased size of the tumor, vasogenic edema, and chronic lacunar infarct in the left pons.

differentiating from other retroclival lesions. CT is limited in the detection of EP due to the lesion's small size and beam-hardening artifacts in the posterior fossa. However, the osseous stalk at the dorsal wall of the clivus on thin-section CT images is defined as a morphologic hallmark of EP and does not occur in other retroclival lesions (11). MR imaging is the best modality for the radiological detection of EP, and the characteristic appearance of EP on MR is that of a well circumscribed, round, extra-axial mass in the prepontine cistern. EP demonstrates high signal intensity on T2-weighted images, low signal intensity on T1-weighted images, and no contrast enhancement (8).

EP is most often confused with intracranial chordoma. The appearance of chordoma on CT is

that of a centrally located, well circumscribed, expansive soft-tissue mass arising from the clivus with associated extensive lytic bone destruction and intratumoral calcification. Chordomas demonstrate intermediate to low signal intensity on T1-weighted MR images and very high signal intensity on T2-weighted MR images. Enhancement is marked and often heterogeneous (8). Chordoma and EP have similar signal intensity on MR images. Unlike intracranial extradural chordomas, the majority of EPs show no contrast enhancement or extensive bone destruction and are usually asymptomatic. Chordomas usually occur in the extradural location, with intradural chordomas being very rare. Intradural chordomas tend not to have any osseous involvement but do tend to be well circumscribed and encapsulated (3). Both EP and intradural chordoma have similar anatomical locations and clinical behaviors and originate from the same embryological remnant. Thus, differentiating between the two lesions based on histopathological and radiological features is likely to be difficult. During a histopathological assessment, a negative MIB-1 index stain has been proposed to be useful in the diagnosis of EP, but intradural chordomas can also demonstrate a negative MIB-1 index stain (6). In radiological examinations, the presences of an osseous stalk at the dorsal wall of the clivus and lack of contrast enhancement help to differentiate EP from an intradural chordoma (8). Some authors have hypothesized that intradural chordoma and EP represent the same spectrum of disease and that intradural chordoma may actually represent a symptomatic form of EP (6). Although our patient was symptomatic, the presence of an osseous stalk and lack of contrast enhancement suggested EP as the most likely diagnosis. Histologic confirmation was not available in this case. Therefore, the diagnosis of an intradural chordoma could not be completely excluded.

The differential diagnosis of EP also includes dermoid, epidermoid, arachnoid cysts and partially thrombosed vertebralbasilar aneurysm. Arachnoid cysts appear as sharply marginated, homogeneous, unilocular masses with signals that are identical to cerebrospinal fluid (CSF) on both CT and MR. Epidermoid cysts are usually less sharply defined, more heterogeneous, and more commonly seen at the cerebellopontine angle cistern. Dermoids are extra-axial masses containing fat or calcifications and

are usually located in the midline (11). Cerebral MR angiography and digital subtraction angiography are useful for differentiating a partially thrombosed vertebrobasilar aneurysm from EP.

EP is usually asymptomatic due to the small size and indolent growth pattern. Only a few case studies have reported EP-associated findings. The associated findings included headache, sudden sensorineural hearing loss, diplopia, fatal pontine hemorrhage, hemiparesis, hemihypoesthesia, CSF fistula, and subarachnoid hemorrhage (1,2,4,5,6,7,9, 10). Fracasso et al. reported hemorrhage areas within the EP on histopathological examination (4). In our case, intratumoral hemorrhage and resultant vasogenic edema due to tumor compression of the brain stem may have been responsible for the sudden onset of symptoms. The neurological findings disappeared after resolution of the intratumoral hemorrhage and vasogenic edema. These findings suggest that the intratumoral hemorrhage rather than the active proliferation of tumor cells may have been more important in the onset of clinical findings in this patient. Although the patient presented in an acute clinical situation, our neurosurgery department recommended observation considering the rapid clinical and radiological improvement. The neurological findings resolved completely after medical therapy. Accurate identification of the nature of a retroclival mass is relevant in the determination of patient prognosis and planning of therapy. Treatment for classic chordoma involves extensive surgery and postoperative radiotherapy (6). Complete surgical resection can be considered as a curative procedure in intradural chordoma (6) while EP is not treated surgically unless it is large and symptomatic (6, 8).

### CONCLUSION

In conclusion, EP is an unusual cause of a retroclival mass. In rare cases, EP can be

symptomatic due to tumor compression of the brain stem. The radiological diagnosis of EP is based on the prepontine cistern location, lack of bony destruction and contrast enhancement, and the presence of an osseous stalk at the dorsal wall of the clivus. When a retroclival mass is established on MRI, any associated osseous stalk should be evaluated on thin-section CT.

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