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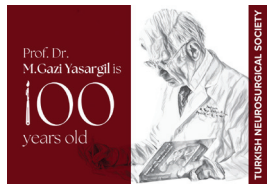

Retroclival Ecchordosis Physaliphora: Diagnosis, Management

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ABSTRACT

AIM: To discuss the diagnostic tests and management options of retroclival Ecchordosis physaliphora (EP).

MATERIAL and METHODS: Four patients with a retroclival EP were assessed. Computerized tomography (CT) and magnetic resonance imaging (MRI) findings were examined thoroughly. Diffusion MRI characteristics were also evaluated. Furthermore, our management protocol has been discussed.

RESULTS: A total of 4 patients with a retroclival EP exhibited similar imaging findings. CT revealed bone changes and a stalk-like connection between the clivus and EP. MRI revealed a lesion that was hyperintense on T2-weighted images and hypointense on T1-weighted images. Neither of the lesions showed contrast enhancement. All lesions were surgically resected. Histopathological examination of the lesions confirmed the diagnosis of benign notochordal remnant.

CONCLUSION: The approach and timing of surgery should be determined according to the lesion parameters in each patient. Large heterogenous lesions that have caused significant bone changes require timely surgery. Small homogenous lesions with a stalk-like connection to clivus should be closely monitored.

KEYWORDS: Notochordal remnants, Chordoma, Retroclival, Benign

ABBREVIATIONS: EP: Ecchordosis physaliphora, **CN:** Cranial nerve, **CT:** Computerized tomography, **MRI:** Magnetic resonance imaging

INTRODUCTION

Ecchordosis physaliphora (EP) is small and benign lesion that originates from notochordal remnants. It is usually located in the midline along the craniospinal axis from the clivus to the sacrococcygeal region, and it has been detected in 0.4%–2% of autopsies (3,14). Typically it is located intradurally in the prepontine cisterna and is attached to the dorsal clival wall via a small pedicle (9,12,13,15).

An EP grows slowly and has been defined as a gelatinous mass. If present in the prepontine region, the EP is usually asymptomatic. However, the lesion may manifest as cranial nerve (CN) palsies if it compresses the brainstem (mass effect). Owing to a common origin, chordomas and EPs have similar histological and ultrastructural characteristics. This makes is challenging to distinguish EPs from chordomas, especially in the prepontine region (4,9,13,16).

The radiological diagnosis of EP and its differentiation from its malignant counterpart chordoma are crucial. Furthermore, studies on EP from a neurosurgical aspect are lacking. Moreover, the previous studies have mostly been radiological

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evaluations and case reports. Thus, case series and studies on the surgical management of EP would be indispensable. In this study, we have discussed the management and diagnostic criteria of EP.

MATERIAL and METHODS

A total of 4875 transsphenoidal endoscopic surgeries were performed for sellar and parasellar pathologies at the Kocaeli University Medical Faculty Neurosurgery Department & Pituitary Research Center, between August 1997 and December 2022. With approval from the institutional review board (17.10.2024 – GOKAEK-2024/15.08), these cases were retrospectively reviewed to identify patients with clival lesions that were consistent with EP. These patients were enrolled in the study, and the following data were collected: sex, age, clinical presentation, imaging findings, operative notes, histopathologic findings, and follow-up data. The demographic features, symptoms, complete medical history, and physical examination findings of the patients were also evaluated. The patients' complaints as well as site and severity of headache were recorded as per the patients' description.

Magnetic resonance imaging (MRI) was performed with a 3T device preoperatively as well as postoperatively at 24 h and 3 months. The MRI protocol consisted of T2-weighted turbo spin echo images (3 mm, gap of 10%, and TR/TE of 4000–4320/90–109) and T1-weighted images obtained before (spin echo, 3–5 mm, gap of 10%–20%, and TR/TE of 600/12) and after (flow compensated spin echo, 3–5 mm, gap of 10%–20%, and TR/TE of 615–647/17) intravenous contrast administration.

An extended endoscopic approach was utilized in all the patients as previously described (1,2,6). In all the patients, a transnasal approach (both nostrils) was utilized. Anterior sphenoidotomy was performed with a microdrill from the inferior border of the ostium. The bone was removed up to the posterior aspect of the nasal septum and the sphenoid floor to access the lower clivus. Neuronavigation was used to verify the anatomic landmarks and lesions.

The operative findings were collected from the operative notes and videos. The lesions of all the study patients had been subjected to histopathological examination. Therefore, the findings were included in the retrospective analysis.

RESULTS

Four patients (male; n=3, female; n=1) with EP were included in the study. The age of the patients ranged from 12 to 42 years (median; 32 years). The most common symptom among the patients was a headache. In addition, two patients complained of restricted lateral eye movements due to 6th cranial nerve palsy. The demographic data and symptomatic findings of the patients are shown in Table I.

Four patients with a retroclival EP exhibited similar imaging findings. Computerized tomography (CT) of the brain revealed bone changes and a stalk-like connection between the clivus and EP in all the patients (Figure 1). MRI of the brain revealed an intradural, well-circumscribed, extra-axial retroclival mass in the prepontine cistern. The lesion appeared hyperintense on T2-weighted images and hypointense on T1-weighted images (Figure 2). None of the lesions exhibited contrast enhancement.

The maximum tumor size did not exceed 15 mm in the asymptomatic patient as well as in the patient who presented with only a headache. However, in the two patients with cranial nerve palsy, the maximum tumor dimensions were 23 and 44 mm (Table II). Brainstem compression was also noted on the MRI.

Histopathologic examination of two resected specimens revealed a tumor that consisted of lobules of mature hyaline cartilage (Figure 3A, Figure 4). Hypercellularity and binucleation were observed in some tumor areas. Mild cellular pleomorphism was also observed (Figure 3B). The tumor had not

Table I: Demographic Data of the Included Patients

Patient no	Sex	Age (years)	Symptoms	Symptom duration (months)
1	Male	12	Headache, cranial nerve palsy	6
2	Male	42	Headache, cranial nerve palsy	3
3	Male	32	Headache	15
4	Female	32	Asymptomatic	-

Table II: Imaging Characteristics of the Included Patients

Patient no	Size (mm)	Contours	Stalk-like connection	T1-signal	T2-signal
1	23	Well circumscribed	Yes	Hypointense	Hyperintense
2	44	Well circumscribed	Yes	Hypointense	Hyperintense
3	27	Well circumscribed	Yes	Hypointense	Hyperintense
4	21	Well circumscribed	Yes	Hypointense	Hyperintense

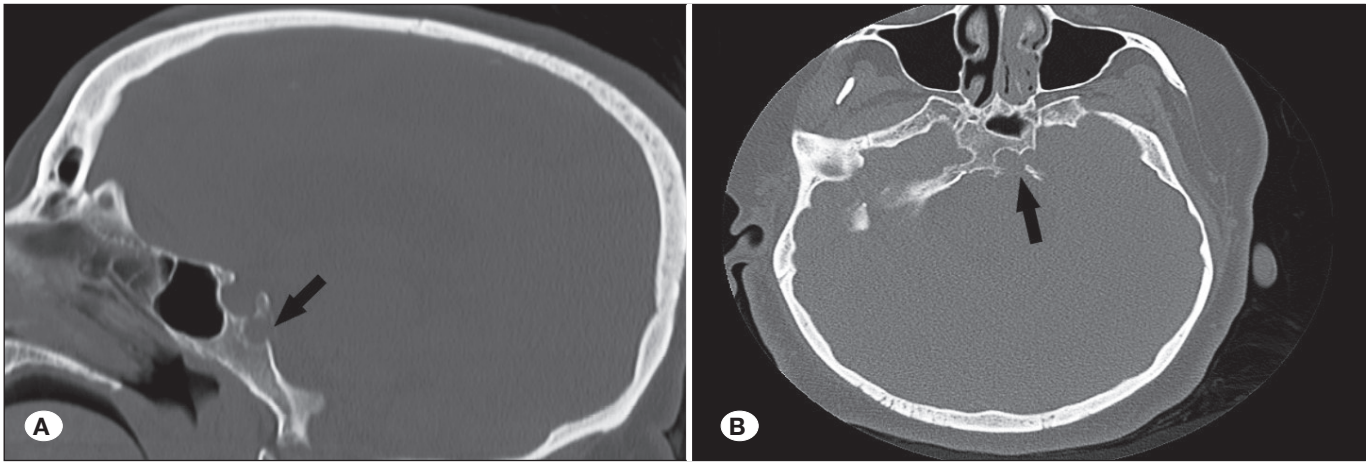

Figure 1: Preoperative sagittal computerized tomography (CT) scan of a representative patient. **A)** Sagittal and **B)** Axial CT images (black arrows shows bone changes).

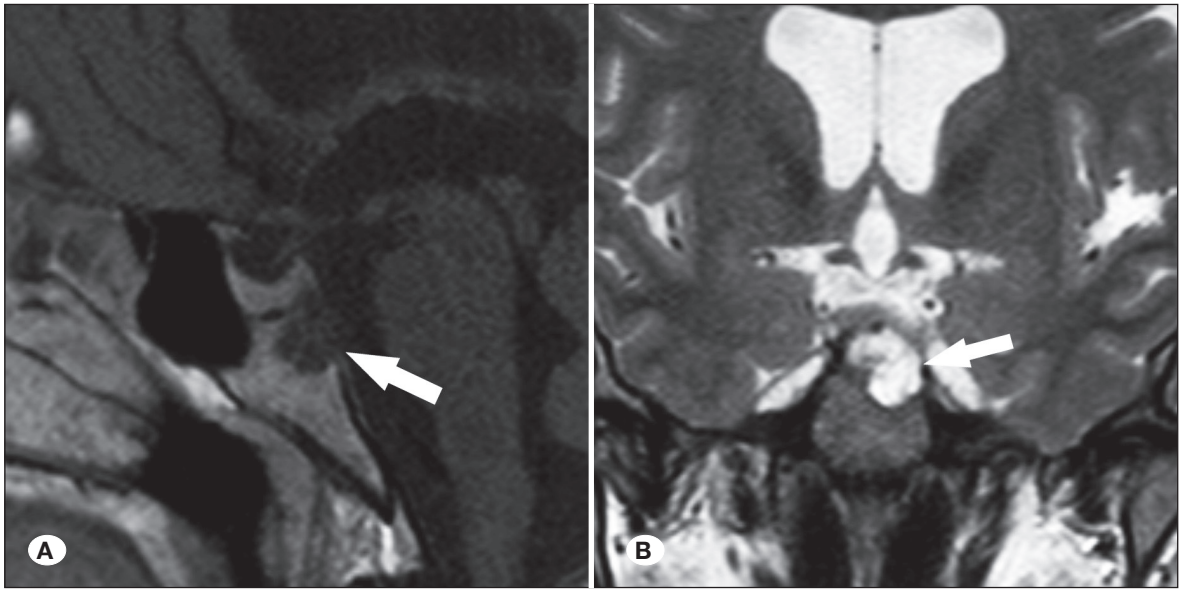

Figure 2: Preoperative magnetic resonance imagings of a representative patient. **A)** Sagittal T1-weighted image showing a hypointense lesion in the retroclival area (white arrow). **B)** Coronal T2-weighted image showing a hyperintense lesion (white arrow).

invaded the surrounding tissue such as brain tissue and bone. Furthermore, mitotic figures were not identified. Based on the radiological, clinical, and histopathologic findings, the patients were diagnosed with skull base enchondroma.

DISCUSSION

EP which was first described by Virchow in 1857 as pathological ectopic notochordal tissue at the posterior clivus, which he named as "*ecchondrosis* physaliphora." The lesion's notochordal origin was described by Muller in 1858. The definitive term *"ecchordosis"* was first used by Ribbert in 1894, confirming Muller's theory (4,5).

Embryologically, the notochord, which represents the primitive skeleton of vertebrates, develops during the third week of gestation. Heterotopic rests of notochordal cells are occasionally found outside the nucleus pulposus, anywhere along the axial skeleton. These rests, which are usually intraosseous, occasionally perforate the dorsal clival wall and enter the subdural or subarachnoid space (4,12,13).

EP is a slow-growing small lesion, measuring a few millimeters to centimeters, that appears as a gelatinous nodule (12), with a predisposition to males (9). They are typically found intradurally in the prepontine cistern, attached to the dorsal clival wall.

Based on the imaging studies, the differential diagnoses of EP include chordoma, dermoid and epidermoid cysts, arachnoid cyst that rarely occurs with chondroid tumors, and metastasis (11,12). Chordomas are hypothesized to be the malignant equivalent of EP, because both entities are of notochordal origin and share common histological and ultrastructural charac-

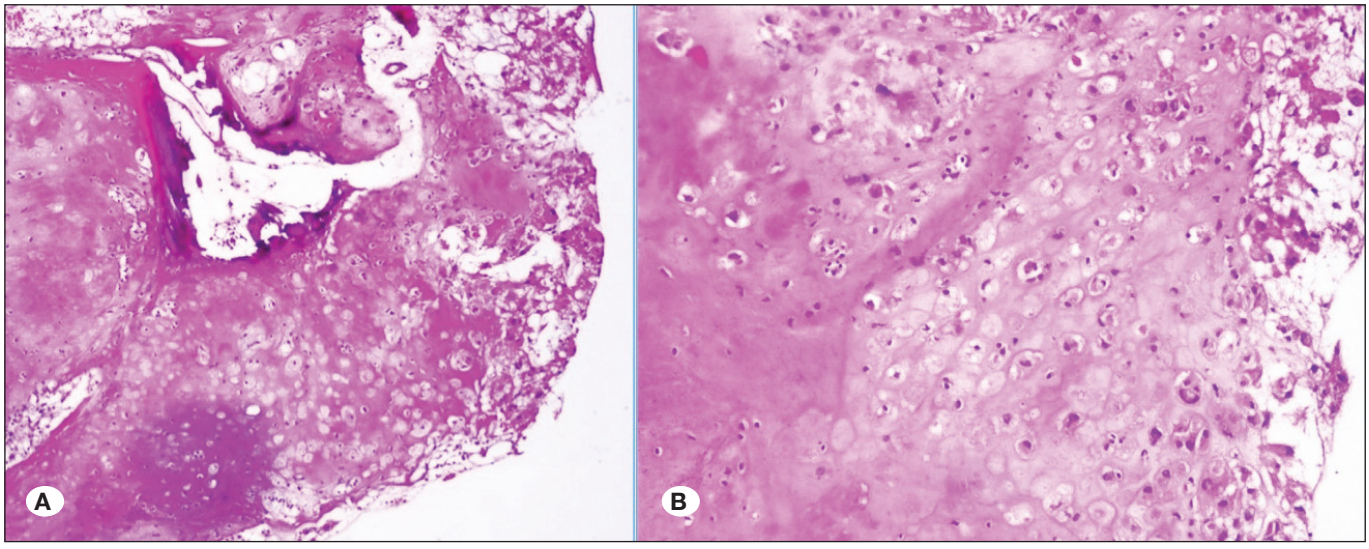

Figure 3: Histopathological examination of the lesion. **A)** The tumor consists of mature hyaline cartilage (Hematoxylin and eosin stain at 10x). **B)** High-power magnification highlights the mild pleomorphism with occasional binucleation (Hematoxylin and eosin stain at 15x.).

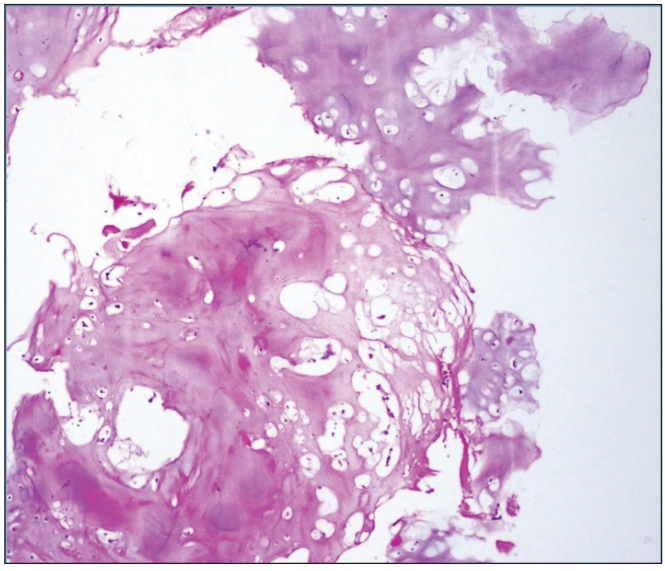

Figure 4: Histological examination of a tumor section. The tumor consists of lobules of mature benign cartilage (Hematoxylin and eosin stain at 10x.).

teristics. However, EPs are usually located intradurally, whereas chordomas are usually located extradurally. Furthermore, chordomas usually cause bone destruction, whereas EPs occasionally cause destruction. On CT, chordomas exhibit intratumoral calcification and local bone invasion. However, the hallmark finding of EPs on thin-sectioned CTs is an osseous stalk connecting the lesion to the dorsal clival wall. This finding is not observed in other retroclival lesions. (14)

Chordomas produce a mass effect due to compression of the brainstem and CNs at the skull base. Thus, patients with chordomas are almost always symptomatic. However, patients with EP are usually asymptomatic (3,7,9,12,14). Furthermore, hemorrhage is a rare complication in EPs.

MRI is the preferred modality for differentiating EPs from chordomas. EPs appear homogeneously hypointense on T1-weighted images and hyperintense on T2-wighted images, Furthermore, a lack of contrast enhancement distinguishes EPs from chordomas or other malignant chondroid tumors or metastases. EPs also appear isointense to hyperintense on diffusion-weighted imaging and hyperintense on an apparent diffusion coefficient map (1.2–1.6 × 10^3 mm^2/second) (10). In our study, no lesion exhibited contrast enhancement.

It remains unclear whether EP is a precursor of chordoma (4,8,11), and randomized studies on this topic are lacking. Previously, it has been debated whether intradural chordomas and large EPs are different entities or whether they can be clubbed as one (9,12,15). Although Wolfe and Scheithauer proposed intradural chordoma be used for all lesions (15), Rodrigues et al. stated that all symptomatic, intradural, extraosseous physaliphorous lesions should be classified as a giant or symptomatic EP as long as an intradural chordoma is absent (12).

A major limitation of this study was the limited number of patients. Larger series and longer follow-ups are needed in order to make it clearer about complications, recurrence and resection rates.

CONCLUSION

To our knowledge there are no significant criteria at present to distinguish simple notochordal remnants from their aggressive types such as chordomas. Such a criteria would aid in identifying cases that require surgical management or follow-up. We propose that the surgical approach and timing for each patient be decided on the basis of an individual's radiological and clinical parameters. Large heterogenous lesions that have caused significant bone changes require timely surgery. However, small homogenous lesions with a stalk-like connection to clivus should be closely monitored. One should also consider

that EPs may be a precursor of chordomas. Hence, if there are any changes in the shape, size, or MRI characteristics of the tumor, the lesion should be surgically resected.

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Declarations

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Availability of data and materials: The datasets generated and/or analyzed during the current study are available from the corresponding author by reasonable request.

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AUTHORSHIP CONTRIBUTION

Study conception and design: MC
Data collection: MC, CV, YA
Analysis and interpretation of results: MC
Draft manuscript preparation: MC, BC
Critical revision of the article: BC, IA
Other (study supervision, fundings, materials, etc...): IA, SC
All authors (MC, BC, CV, YA, IA, SC) reviewed the results and approved the final version of the manuscript.

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