

Multiple Echordosis Physaliphora: A Challenging Diagnosis

Xiao-Ling Zhong^{1,2}, Biao Huang², Chao Liu³, Sheng-Quan Zhan⁴

¹Southern Medical University, Guangzhou, Guangdong 510515, China

²Department of Radiology, Guangdong Academy of Medical Sciences, Guangdong General Hospital, Guangzhou, Guangdong 510080, China

³Department of Pathology, Guangdong Academy of Medical Sciences, Guangdong General Hospital, Guangzhou, Guangdong 510080, China

⁴Department of Neurosurgery, Guangdong Academy of Medical Sciences, Guangdong General Hospital, Guangzhou, Guangdong 510080, China

Key words: Echordosis Physaliphora; Magnetic Resonance Imaging; Notochord; Pathology; Surgery

Echordosis physaliphora (EP) is a benign congenital hamartoma of the ectopic notochordal remnant, accounting for 0.5–2% of all autopsies, and found in 1.5% of all magnetic resonance imaging (MRI) of the brain. It commonly occurs along the midline within the retroclival and the prepontine cistern. EP has a slow growth rate and can vary in size from a few mm to 2 cm.^[1] Giant symptomatic EPs are extremely rare, and only four cases have been reported in the literature.^[2–5] No cases of multiple EPs or EPs with metastasis have been reported.

We report a rare case of giant symptomatic EP with multiple lesions along the midline in the prepontine cistern.

A 34-year-old male presented to Guangdong General Hospital with a 1-year history of diplopia in his left eye. On physical examination, his left eye had limited abduction, but there were no other significant physical abnormalities. Brain MRI disclosed multiple well-defined rounded lesions, measuring 30 mm in diameter, located along the midline within the prepontine cistern, mildly compressing the left cavernous sinus. Lesions were also found within the pontine cistern, suprasellar cistern, cistern ambiens, and superior vermian cistern. The lesions appeared hypointense on both T1-weighted images (T1-WI) and T2-fluid attenuated inversion recovery (FLAIR) images and hyperintense on T2-weighted images (T2-WI) and did not enhance after gadolinium administration [Figure 1]. Computed tomography showed multiple low-attenuation lesions within the pontine cistern, suprasellar cistern, cistern ambiens, and superior vermian cistern and an osseous stalk arising from the retroclivus [Figure 1].

At surgery, through a left frontotemporal craniotomy, multiple cystic gelatinous nodules were revealed ventral to the pons. The multiple cystic tumors were partially resected.

Histological examination revealed scattered physaliphorous cell nests with a lobular growth pattern. The cytoplasm was eosinophilic and vacuolated with a mucin-filled matrix. No mitotic activity or cellular pleomorphism was revealed [Figure 1].

Immunohistochemical staining with proliferation marker MIB-1 showed a relatively low proliferation index [$<2\%$; Figure 1]. A final diagnosis of multiple EPs was made.

The patient's postoperative course was uneventful. Since discharge, he has been followed up in our clinic at regular intervals with MRIs which have demonstrated stable residual tumor with no regrowth of the masses.

Luschka first described his observation of notochord cells along the retroclivus in 1856. Virchow named the lesion "echordosis physaliphora," based on his theory that it was derived from the spheno-occipital synchondrosis. In 1858, Müller theorized the notochordal origin of EP, which was subsequently confirmed in 1894 by Ribbert who coined the definitive term "echordosis physaliphora".^[3]

Phylogenetically, the notochord represents the primitive skeleton of vertebrates that forms during the 3rd week of gestation. It persists in adults as the nucleus pulposus of the intervertebral disc. Occasionally, heterotopic rests of notochordal cells are found outside the nucleus pulposus

Address for correspondence: Dr. Biao Huang,

Department of Radiology, Guangdong Academy of Medical Sciences, Guangdong General Hospital, No. 106 Zhongshan 2nd Road, Guangzhou, Guangdong 510080, China

E-Mail: cjr.huangbiao@vip.163.com

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

© 2015 Chinese Medical Journal | Produced by Wolters Kluwer - Medknow

Access this article online

Quick Response Code:



Website:
www.cmj.org

DOI:
10.4103/0366-6999.167368

Received: 01-07-2015 **Edited by:** Xin Chen

How to cite this article: Zhong XL, Huang B, Liu C, Zhan SQ. Multiple Echordosis Physaliphora: A Challenging Diagnosis. *Chin Med J* 2015;128:2826-8.

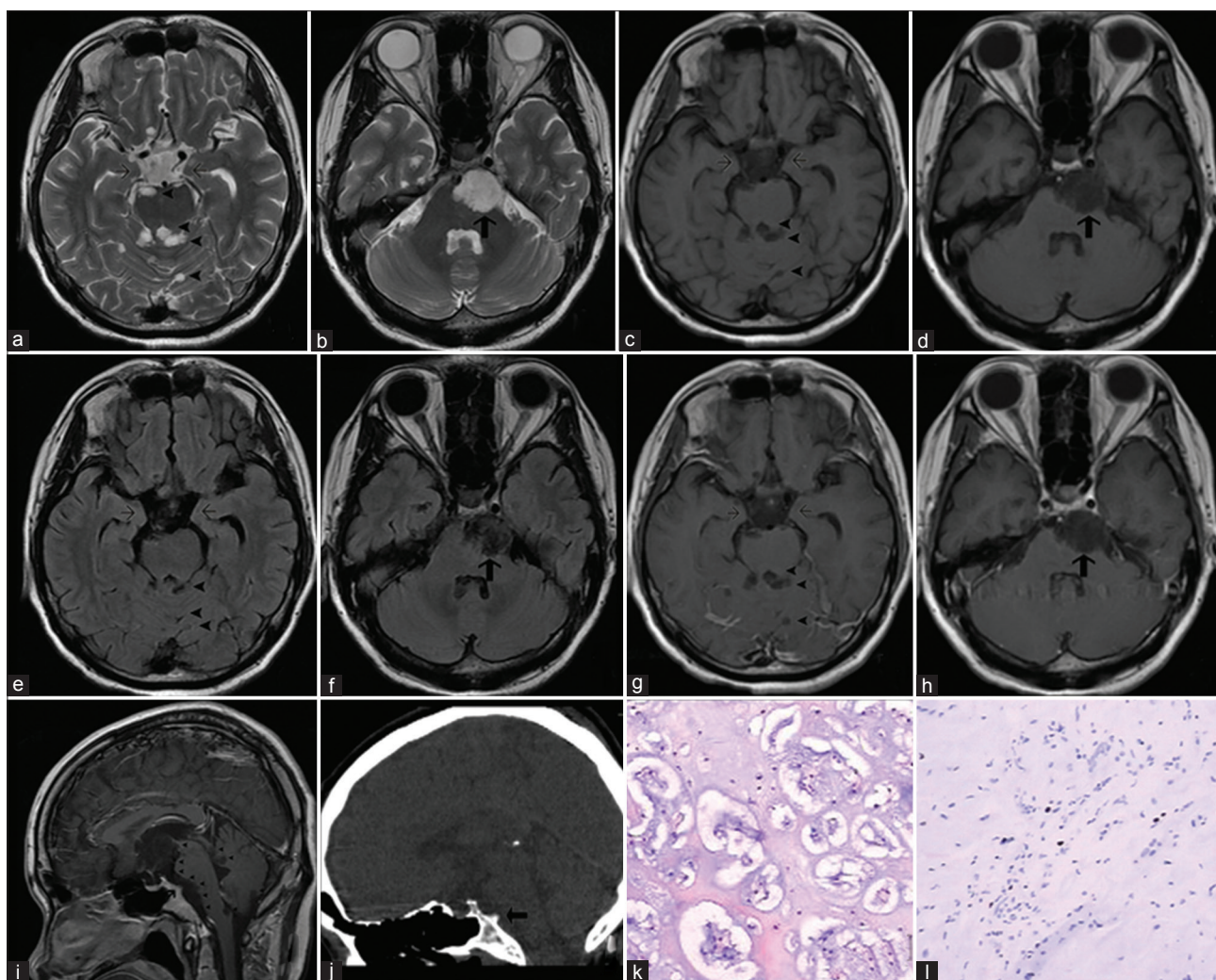


Figure 1: T2-weighted image (a and b), T1-weighted image (c and d), and T2-fluid attenuated inversion recovery (e and f) revealed multiple, well-defined, rounded masses in the pontine cistern, suprasellar cistern, cistern ambiens, superior vermillion cistern, mildly compressing the cavernous sinus. The lesion appears hyperintense on the T2-weighted image and hypointense on the T1-weighted image and T2-fluid attenuated inversion recovery without contrast enhancement (g and h). (i) Sagittal enhanced the T1-weighted image. (j) CT (bone window) showed a subtle stalk-like structure projecting from the clivus (arrow). (k) Hypocellularity of the physaliphorous cells with a lobular growth pattern, eosinophilic and vacuolated cytoplasm with a mixomatous matrix, the absence of mitoses and of cellular pleomorphism (HE, original magnification, $\times 200$). (l) MIB-1 showed a relatively low proliferation index, lower than 2% (Immunohistochemical staining, original magnification, $\times 200$).

anywhere along the craniospinal axis, primarily within the clival or sacrococcygeal regions. EP may be completely intradural or may break through a weak point in the dura mater and become attached to the clival bone by a narrow base.^[3] This could explain the possible pathogenesis of multiple EPs within the midline areas, as documented in our case. As an alternative hypothesis, the notochordal cells at the beginning of embryonic life may disseminate to other spaces, with the largest lesion located along the retroclivus (i.e., the primary lesion, as in our case, with dissemination of the other nodules).

Most patients with EP are asymptomatic due to its small size and slow growth, but they can become symptomatic when the tumor compresses adjacent structures. To the best of our knowledge, only 4 cases of giant symptomatic EP, located in the retroclival prepontine region, have been reported.^[2-5] Of the 4 giant reported cases and our present patient with

symptomatic EP found in the retroclival prepontine location, with a size from 20 mm to 40 mm. Three were male and one was female with a mean age of 28 years (range: 12–63 years). The major symptoms presented were headache and diplopia. On MRI, those lesions were hypointense on T1-WI and hyperintense on T2-WI and demonstrated no contrast enhancement. Moreover, only Krisht *et al.* described a small bone stalk arising from the retroclivus. In our present case of multiple EPs, the imaging characteristics are consistent with those of previously reported EP.

Chordomas can also have a notochordal origin and are morphologically similar to EP. Both EP and chordomas are composed of cells containing multiple clear cytoplasmic vacuoles indenting the nucleus (known as physaliphorous cells) with copious extracellular pools of mucin. Immunohistochemical staining with S-100 protein, keratin, and epithelial membrane antigen markers is not sufficient

to differentiate between chordoma and EP. However, MIB-1 >5% may predict a chordoma.

On MRI, a chordoma usually arises extradurally, shows bone destruction, and enhances with contrast administration. In our case, the key imaging feature was the osseous stalk arising from the basisphenoid portion of the clivus, which is considered a morphologic hallmark of EP and is absent in other retroclival lesions, such as dermoids, epidermoids, arachnoid cysts, and neuroenteric cysts. Dermoid cysts often show shortened T1 relaxation times, and epidermoid cysts are more commonly seen within the cerebellopontine angle cistern. An arachnoid cyst can have attenuation and signal intensity similar to the cerebrospinal fluid. However, they are most commonly seen in the middle cranial fossa and are rarely seen in a retroclival location. In addition, lack of contrast enhancement and the low proliferation index favored multiple EPs as the most likely diagnosis in our case.

EP can be managed expectantly with serial MRI follow-up since it has an indolent growth and rarely causes symptoms throughout a person's life. Our case demonstrates that these lesions can, in certain cases, become large and cause compressive symptoms, typically including headache and diplopia. Surgical treatment is considered the gold standard for symptomatic EP, usually performed by craniotomy. A few studies have reported that endoscopic endonasal transsphenoidal surgery is a feasible, minimally-invasive alternative to craniotomy which allows for complete excision of a clival EP with less postoperative complications.

To our knowledge, this is the first case describing multiple EPs. An MRI study revealing multiple lesions along the midline which are homogeneously hypointense on T1-WI and T2-FLAIR and hyperintense on T2-WI without contrast enhancement and without bony involvement can be helpful in the differential diagnosis. The characteristic imaging feature of an osseous stalk arising from the basisphenoid portion of the clivus may help make the proper diagnosis and guide treatment.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Golden LD, Small JE. Benign notochordal lesions of the posterior clivus: Retrospective review of prevalence and imaging characteristics. *J Neuroimaging* 2014;24:245-9.
2. Yamamoto T, Yano S, Hide T, Kuratsu J. A case of ecchordosis physaliphora presenting with an abducens nerve palsy: A rare symptomatic case managed with endoscopic endonasal transsphenoidal surgery. *Surg Neurol Int* 2013;4:13.
3. Krisht KM, Palmer CA, Osborn AG, Couldwell WT. Giant ecchordosis physaliphora in an adolescent girl: Case report. *J Neurosurg Pediatr* 2013;12:328-33.
4. Choudhri O, Feroze A, Hwang P, Vogel H, Ajlan A, Harsh G 4th. Endoscopic resection of a giant intradural retroclival ecchordosis physaliphora: Surgical technique and literature review. *World Neurosurg* 2014;82:912.e21-6.
5. Takeyama J, Hayashi T, Shirane R. Notochordal remnant-derived mass: Ecchordosis physaliphora or chordoma? *Pathology* 2006;38:599-600.