

## Case Report

# A case of echordosis physaliphora presenting with an abducens nerve palsy: A rare symptomatic case managed with endoscopic endonasal transsphenoidal surgery

Takahiro Yamamoto<sup>1,2</sup>, Shigetoshi Yano<sup>1</sup>, Takuichiro Hide<sup>1</sup>, Jun-ichi Kuratsu<sup>1</sup>

<sup>1</sup>Department of Neurosurgery, Kumamoto University School of Medicine, Kumamoto, Japan, <sup>2</sup>Center of Neurosurgery, Nobeoka Hospital, Nobeoka, Japan

E-mail: Takahiro Yamamoto - tyamamoto0915@yahoo.co.jp; \*Shigetoshi Yano - yanos@kumamoto-u.ac.jp; Takuichiro Hide - thide@fc.kuh.kumamoto-u.ac.jp; Jun-ichi Kuratsu - jkuratsu@kumamoto-u.ac.jp

\*Corresponding author

Received: 22 September 12

Accepted: 03 December 12

Published: 28 January 13

### This article may be cited as:

Yamamoto T, Yano S, Hide T, Kuratsu J. A case of echordosis physaliphora presenting with an abducens nerve palsy: A rare symptomatic case managed with endoscopic endonasal transsphenoidal surgery. *Surg Neurol Int* 2013;4:13.

Available FREE in open access from: <http://www.surgicalneurologyint.com/text.asp?2013/4/1/13/106562>

Copyright: © 2013 Yamamoto T. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

## Abstract

**Background:** Echordosis physaliphora (EP) is a benign notochordal remnant that is usually asymptomatic; symptomatic cases are extremely rare. Most of the reported symptomatic cases were managed by resection via craniotomy.

**Case Description:** We report a case of a 20-year-old male presenting with abducens nerve palsy. Magnetic resonance imaging performed on admission demonstrated a mass in the retroclival prepontine location. The patient was treated successfully by endoscopic endonasal trans-sphenoidal surgery (ETSS), his postoperative course was uneventful, and the abducens nerve palsy disappeared.

**Conclusion:** ETSS has advantages not only for treatment but also for differentiation between EP and intradural chordoma. This is the first case of symptomatic EP successfully treated solely by ETSS.

**Key Words:** Abducens nerve palsy, chordoma, echordosis physaliphora, endoscopic endonasal transsphenoidal surgery

### Access this article online

#### Website:

[www.surgicalneurologyint.com](http://www.surgicalneurologyint.com)

#### DOI:

10.4103/2152-7806.106562

#### Quick Response Code:



## INTRODUCTION

Echordosis physaliphora (EP) is a benign notochordal remnant that is usually asymptomatic and requires no intervention.<sup>[3-5,15]</sup> However, symptomatic EP cases are extremely rare, as only 11 cases have been reported to date, most of which were managed by resection via craniotomy.<sup>[1,2,4,6-8,10-12,14,15]</sup> Here we report, a rare case of symptomatic EP managed successfully by endoscopic endonasal trans-sphenoidal surgery (ETSS) without resection via craniotomy. We also present a review of the literature and discuss the advantages of ETSS and the differences between EP and intradural chordoma.

## CASE REPORT

### History and examination

A 20-year-old healthy male was admitted to our hospital for sudden onset of diplopia. Physical examination at admission revealed right abducens nerve palsy but no other neurological disorder. The diplopia arose when he gazed straight ahead or to the right. Computed tomography (CT) and magnetic resonance imaging (MRI) demonstrated a lesion, measuring 22 mm in diameter, which was isodense and had no enhancement on CT. On MRI, the lesion appeared heterogeneously hyperintense on a T1-weighted image,

more hyperintense on a T2-weighted image, and had slightly restricted diffusion on diffusion-weighted images. No contrast enhancement was observed after the addition of gadolinium diethylenetriamine pentaacetic acid. The lesion compressed the patient's pons and right abducens nerve [Figure 1].

### Treatment

ETSS was performed. A gelatinous mass was partially exposed after removing the mucosa of the sphenoid sinus. The clival bone was drilled to entirely expose the mass, which was mainly located in the extradural space and partially located intradurally, accompanied by a small dural defect. The intradural mass had compressed the arachnoid mater, which had a yellowish discoloration [Figure 2]. To avoid damage to the patient's right abducens nerve, the mass in Dorello's canal was partially removed. A waterproof repair of the clivus was performed to avoid cerebrospinal fluid (CSF) leakage using Surgicel® (Ethicon 360, Menlo Park, CA, USA) with fibrinogen and Spongel® (Ethicon 360) with thrombin followed by formation of a pediculate septomucosal flap.

### Postoperative course

The patient's postoperative course was uneventful. Postoperatively, the right abducens nerve palsy improved, but his diplopia persisted when he gazed to the right. Postoperative MRI demonstrated partial removal of the mass, adequate for decompression [Figure 3]. Four months postoperatively, the patient's right abducens nerve palsy and diplopia completely resolved. No residual

mass was revealed on MRI [Figure 3]. At 1-year follow-up, the patient remained symptom- and disease-free.

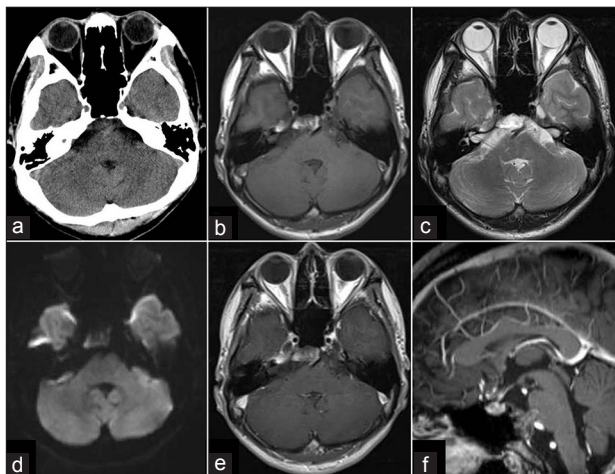
### Pathological findings

Pathological examination of the mass showed hypocellular physaliphorous cells with a lobular growth pattern and eosinophilic cytoplasm with vacuolated mucous droplets. Neither mitosis nor dyskaryosis were visible [Figure 4]. Immunohistochemical study showed positive with antibodies against Cytokeratins. The Ki-67 (MIB-1) index, evaluated using the MIB-1 antigen against Ki-67, was not increased. The MRI findings, pathological aspects, clinical course, and surgical findings suggested EP.

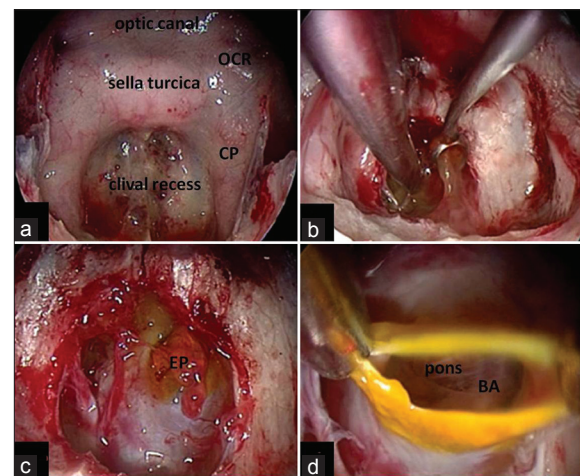
### DISCUSSION

EP is an ectopic notochordal remnant,<sup>[13]</sup> typically located in the intradural prepontine area, but it can be found all along the vertebral column.<sup>[6]</sup> EP is a benign gelatinous nodule, varies in size from a few millimeters to 35 mm, and typically forms a retroclival mass accompanied by a stalk connected to the clival notochordal remnant. It is usually asymptomatic and found incidentally in approximately 2% of all autopsies.<sup>[3-5,13]</sup>

Symptomatic EP cases are extremely rare and only a few cases have been reported to date. To the best of our knowledge, only 11 cases of symptomatic EP, located in the retroclival prepontine region have been reported, but features of EP remain to be elucidated. Of the 11 previously reported cases and our present patient with



**Figure 1:** (a) Unenhanced cranial CT performed on admission. The mass is isodense with no visible bone destruction, (b) T1-weighted MRI performed on admission. The mass appears heterogeneously hyperintense, (c) T2-weighted MRI performed on admission. The mass appears more hyperintense than by T1-weighted MRI, (d) Diffusion-weighted images demonstrate that the mass has slightly restricted diffusion, (e) Gadolinium-enhanced axial T1-weighted MRI performed on admission showing no contrast enhancement, (f) Gadolinium-enhanced sagittal T1-weighted MRI performed on admission demonstrate a lesion in the retroclival prepontine location with no contrast enhancement

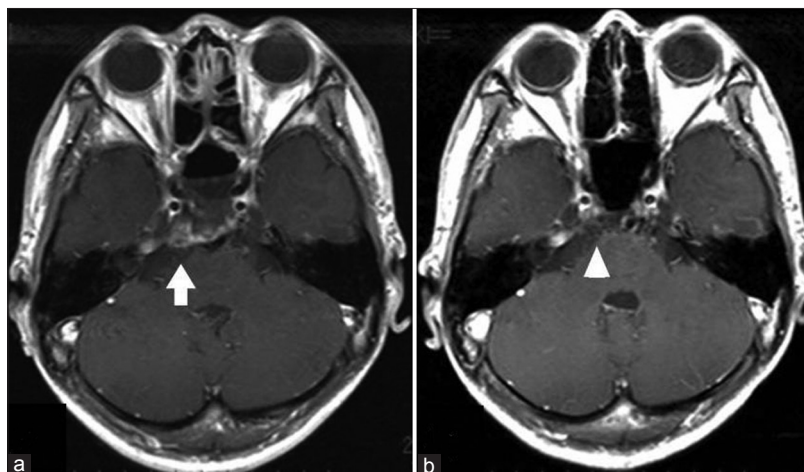


**Figure 2:** Operative findings. (a) An image obtained before drilling the clival recess. The patient underwent ETSS, (b) After removing the mucosa of the sphenoid sinus, the clival bone was drilled to entirely expose the mass. The gelatinous and yellowish-brown mass mainly located in the extradural space, (c) The mass is partially located intradurally accompanied with a small dural defect, (d) An image obtained after removal of the mass. The mass is partially removed and decompression is performed. OCR = opticocarotid recess, CP = carotid protuberance, EP = echordosis physaliphora, BA = basilar artery

symptomatic EP found in the retroclival prepontine location, 5 were male and 7 were female with a mean age of 40.9 years (range, 12–75 years). The major symptoms presented were headache (33.3%), diplopia (25%), and subarachnoid hemorrhage (16.6%) [Table 1].<sup>[1,2,4,6-8,10-12,14,15]</sup>

To date, nine cases of the reported symptomatic EP cases were resected by craniotomy, one case was treated by craniotomy with endoscopic assist, and one case was treated by endoscopic transthoracic approach.<sup>[4,10]</sup> The present EP case is the first case treated solely by ETSS. The advantage of endoscopic surgery

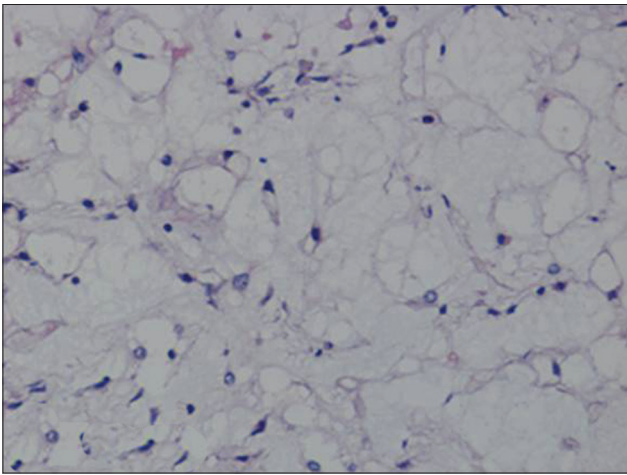
was described by Cha *et al.*, who treated a symptomatic EP case by craniotomy with endoscopic assist.<sup>[4]</sup> An endoscope allows the surgeon to view a wider area to identify detailed anatomical features and relationships between EP and cranial nerves. We believe that ETSS is valuable for approaching a retroclival prepontine lesion. Ciarpaglini *et al.* reported similar approach by ETSS in the case of intradural chordoma.<sup>[5]</sup> By ETSS, the surgeon can approach a retroclival prepontine lesion without retracting the brain. To expose the mass, the clivus must be opened, which is a minimally invasive approach.



**Figure 3:** (a) Postoperative gadolinium-enhanced axial T1-weighted MRI obtained 1 week after surgery revealing a residual mass on the right Dorello's canal (arrow), (b) Gadolinium-enhanced axial T1-weighted MRI obtained 4 months after surgery does not reveal a residual mass (arrow head)

**Table 1: Reported cases of symptomatic echordosis physaliphora**

Case No.	Author	Year	Age (years)/sex	Symptoms	Treatment	Approach	Outcomes	Follow-up
1	Stam <i>et al.</i>	1982	75/M	SAH	Conservative treatment	-	Died	-
2	Macdonald <i>et al.</i>	1990	66/F	CSF leak	Total resection	Trans sphenoidal	Resolved	Recurrence free
3	Akimoto <i>et al.</i>	1996	51/F	Headache, Diplopia	Total resection	Presigmoid	Resolved	Undocumented
4	Toda <i>et al.</i>	1998	56/F	Headache	Total resection	Lateral suboccipital	Resolved	Recurrence free
5	Cha <i>et al.</i>	2002	49/M	Headache, Dizziness	Total resection	Trans maxillary, Endoscopic assist	Resolved	Recurrence free
6	Takeyama <i>et al.</i>	2006	12/M	Hemiparesis, Diplopia	Subtotal resection	Undocumented	Resolved	Recurrence free
7	Ling <i>et al.</i>	2007	45/F	Hearing loss, tinnitus	Total resection	Trans petrosal	Undocumented	-
8	Fracasso <i>et al.</i>	2007	48/F	SAH	Conservative treatment	-	Died	-
9	Rotondo <i>et al.</i>	2007	47/F	Headache, facial pain	Total resection	Presigmoid	Resolved	Recurrence free
10	Miki <i>et al.</i>	2008	59/M	Gait disturbance, dementia	Total resection	Endoscopic transthoracic approach	Resolved	Recurrence free
11	Alkan <i>et al.</i>	2009	22/M	SAH	Conservative treatment	-	Resolved	Spontaneous loss, Recurrence free
12	Present case	2012	20/M	Diplopia	Subtotal resection	Endoscopic transsphenoidal approach	Resolved	Spontaneous loss, Recurrence free



**Figure 4: A pathological examination of the mass showing hypocellular physaliphorous cells with a lobular growth pattern and eosinophilic cytoplasm with vacuolated mucous droplets. Neither mitosis nor dyskaryosis is visible. (H and E, ×200)**

In our case, the residual mass disappeared after 4 months follow-up. From our review, two of three followed-up cases, which had residual mass after treatment, showed spontaneous loss.<sup>[2,14]</sup> This could be a feature of EP. Therefore, it may be advisable to follow up with MRI periodic control for residual mass of EP after treatment. It needs additional studies.

Next, we wish to distinguish EP from intradural chordoma. Interventions of intradural chordomas should be aimed at total removal of the tumor, which may require adjuvant radiotherapy for the residual tissue.<sup>[4,5]</sup> Using MRI, Mehnert *et al.* reported differential diagnoses between EP and a chordoma and found that all EP cases showed no contrast enhancement by gadolinium-enhanced T1-weighted MRI.<sup>[9]</sup> It is difficult to pathologically differentiate EP from an intradural chordoma. Although typical physaliphorous cells were found in the present case, it is well known that they are present in both EPs and intradural chordomas.<sup>[16]</sup> Macdonald *et al.* investigated the immunohistochemical differences between EP and a chordoma and found that routine immunohistochemistry using stains for cytokeratin, vimentin, epithelial membrane antigen, carcinoembryonic antigen, and S-100 was not helpful for differentiation.<sup>[8]</sup> In our case, immunohistochemical study showed positive with antibodies against cytokeratins, but this was not helpful for differentiation. Amer *et al.* explored differential diagnoses between EP, chordomas, and benign notochordal cell tumors and found that an increased Ki-67 (MIB-1) index indicated proliferative activity in chordomas with respect to EPs.<sup>[3]</sup> In our case, the Ki-67 index was not increased.

From a review of the literature, gadolinium-enhanced T1-weighted MRI, the Ki-67 index, and surgical findings

of a gelatinous nodule were the most available differential criteria. In our case, the mass was gelatinous, had no enhancement on MRI, and the Ki-67 index was not increased. Moreover, the residual mass disappeared after 4 months. These findings and the clinical course suggested a diagnosis of EP in our case. Operative findings are important for a diagnosis of EP and to differentiate from an intradural chordoma. Therefore, ETSS has advantages not only for resection but also for diagnosis.

## CONCLUSION

Here we present a rare case of symptomatic EP that was managed successfully by ETSS without resection via craniotomy. ETSS has advantages over resection by craniotomy, but additional studies are needed to clarify the features of EP and to conveniently differentiate EP from an intradural chordoma.

## REFERENCES

1. Akimoto J, Takeda H, Hashimoto T, Haraoka J, Ito H. A surgical case of echordosis physaliphora [in Japanese]. *No Shinkei Geka* 1996;24:1021-5.
2. Alkan O, Yildirim T, Kizilkiliç O, Tan M, Cekinmez M. A case of echordosis physaliphora presenting with an intratumoral hemorrhage. *Türk Neurosurg* 2009;19:293-6.
3. Amer HZ, Hameed M. Intraosseous benign notochordal cell tumor. *Arch Pathol Lab Med* 2010;134:283-8.
4. Cha ST, Jarrahy R, Yong WH, Eby T, Shahinian HK. A rare symptomatic presentation of echordosis physaliphora and unique endoscope-assisted surgical management. *Minim Invasive Neurosurg* 2002;45:36-40.
5. Ciarpaglini R, Pasquini E, Mazzatenta D, Ambrosini-Spaltro A, Sciarretta V, Frank G. Intradural clival chordoma and echordosis physaliphora: A challenging differential diagnosis: Case report. *Neurosurgery* 2009;64:E387-8.
6. Fracasso T, Brinkmann B, Paulus W. Sudden death due to subarachnoid bleeding from echordosis physaliphora. *Int J Legal Med* 2008;122:225-7.
7. Ling SS, Sader C, Robbins P, Rajan GP. A case of giant echordosis physaliphora: A case report and literature review. *Otol Neurotol* 2007;28:931-3.
8. Macdonald RL, Cusimano MD, Deck JH, Gullane PJ, Dolan EJ. Cerebrospinal fluid fistula secondary to echordosis physaliphora. *Neurosurgery* 1990;26:515-8.
9. Mehnert F, Beschoner R, Küker W, Hahn U, Nägele T. Retroclival echordosis physaliphora: MR imaging and review of the literature. *AJNR Am J Neuroradiol* 2004;25:1851-5.
10. Miki T, Nakajima N, Akimoto J, Wada J, Haraoka J. Neuroendoscopic trans-third ventricle approach for lesions of the ventral brainstem surface. *Minim Invasive Neurosurg* 2008;51:313-8.
11. Rotondo M, Natale M, Mirone G, Cirillo M, Conforti R, Scuotto A. A rare symptomatic presentation of echordosis physaliphora: Neuroradiological and surgical management. *J Neurol Neurosurg Psychiatry* 2007;78:647-9.
12. Stam FC, Kamphorst W. Echordosis physaliphora as a cause of fatal pontine hemorrhage. *Eur Neurol* 1982;21:90-3.
13. Stewart MJ, Burrow Jle F. Echordosis physaliphora sphenoccipitalis. *J Neurol Psychopathol* 1923;4:218-20.
14. Takeyama J, Hayashi T, Shirane R. Notochordal remnant-derived mass: Echordosis physaliphora or chordoma? *Pathology* 2006;38:599-600.
15. Toda H, Kondo A, Iwasaki K. Neuroradiological characteristics of echordosis physaliphora. Case report and review of the literature. *J Neurosurg* 1998;89:830-4.
16. Wyatt RB, Schochet SS Jr, McCormick WF. Echordosis physaliphora. An electron microscopic study. *J Neurosurg* 1971;34:672-7.