Neuroradiological characteristics of ecchordosis physaliphora

Case report and review of the literature

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An extremely rare case of ecchordosis physaliphora is presented in which the authors focus especially on its radiological characteristics. The patient complained of a headache with no other neurological abnormalities. A thorough radiological examination revealed a small intradural prepontine mass with no bone destruction of the clivus. Magnetic resonance imaging was very useful in visualizing this mass as a low signal intensity lesion on T1-weighted images and as a high signal intensity lesion on T2-weighted images without any contrast enhancing effects. At surgery, a cystic gelatinous nodule was found ventral to the pons; the nodule was connected to the dorsal wall of the clivus via a delicate stalk. Histological studies proved that this was an ecchordosis physaliphora. Review of the literature demonstrates that the reported cases of ecchordoses have many common radiological features that would suggest the diagnosis of this rare disease.

Key Words • ecchordosis physaliphora • chordoma • magnetic resonance imaging • computerized tomography

Case Report

This 56-year-old woman presented to our institute with a 3-month history of a persistent headache.

Physical Examination. The patient’s neurological examination was not significant except for the headache and no other signs suggesting increasing intracranial pressure were observed. A cerebrospinal fluid study yielded negative findings.

Radiological Studies. A plain cranial radiograph and a routine plain CT scan showed no destruction of bone structures or abnormal calcification. A magnified CT scan of the bone window revealed a subtle pedicle-like structure protruding from the dorsal wall of the clivus (Fig. 1). A contrast-enhanced CT scan did not disclose any enhancing lesions. Magnetic resonance imaging demonstrated a round extraaxial mass, 15 mm in diameter, located anterior to the pons, which was situated along the midline cranial axis in the intradural space (Fig. 2). The lesion appeared hypointense on a T1-weighted image and homogeneously hyperintense on a T2-weighted image. A sagittal section of the T2-weighted image revealed that the
lesion was slightly compressing the pons, but the clivus was unaffected (Fig. 3). Addition of gadolinium–diethyl-ene triamine pentaacetic acid (Gd-DTPA) contrast medium provided no enhancing effects. Computerized tomography cisternography demonstrated an isolated small filling defect at the prepontine cistern (Fig. 4). Cerebral angiography revealed no pathological findings.

Operation. Although the clear correlation between the patient’s headache and the mass was not explained, exploratory surgery was undertaken. Surgery performed via a left lateral suboccipital craniectomy revealed a cystic gelatinous nodule located ventral to the pons. The mass was easily dissected en bloc from surrounding structures. It was apparently attached to the dura mater at the dorsal aspect of the clivus via a small stalklike structure. After removal of the cystic portion of the mass, this small stalk-like osseous process was found to have penetrated the dura of the clivus and was easily removed with a curette.

Postoperative Course. The patient’s postoperative course was uneventful and her headache resolved. A post-operative MR image demonstrated no evidence of a residual mass and no regrowth has occurred after 2 years of follow-up evaluation.

Pathological Examination. Microsection of the specimen revealed hypocellularity of the physaliphorous cells with a lobular growth pattern. The cytoplasm was eosinophilic and vacuolated with a myxomatous matrix. No mitosis or cellular pleomorphism was revealed by hematoxylin and eosin staining (Fig. 5). Immunohistochemical staining for epithelial membrane antigen, S-100 protein, and vimentin
Discussion

Ectopic notochordal remnants were first described by Luschka in 1856 and were subsequently termed “echordosis physaliphora” by other investigators.\(^1,3,4\) The notochord forms during the 3rd week of gestation and becomes the nucleus pulposus of the intervertebral disc at maturity.\(^4,10\) Heterotopic rests of notochordal cells are occasionally found outside the nucleus pulposus anywhere along the axial skeleton such as within the clivus. Although these rests usually have an intraosseous location, they occasionally perforate through the dorsal wall of the clivus into the subdural or subarachnoid space.\(^4,10\) This could explain the possible pathogenesis of an intradural echordosis physaliphora at the prepontine area. Thus, echordoses physaliphora are thought to be congenital malformations.\(^1,3,4,10,11\)

The main differential diagnoses of echordosis are dermoid and epidermoid cysts and chordoma, all of which occur frequently at the same location as the echordosis.\(^2,8,10\) Chordomas have a notochordal origin and are morphologically similar to echordoses; however, these two are quite different from each other in their biological behavior.\(^2,5\) In contrast to the benign nature of echordoses, chordomas are usually considered to be malignant.\(^2,5\)

Although dermoid and epidermoid cysts are also thought to be congenital malformations rather than true neoplasms, they tend to grow slowly and manifest late in life.\(^8\) To our knowledge, there is little information on the specific radiological features of echordoses in the literature. We therefore conclude that knowledge of the radiographic characteristics of echordosis physaliphora would be of value in diagnosing and differentiating this uncommon disease and in deciding therapeutic strategies.

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**Fig. 4.** Computerized tomography cisternogram demonstrating a filling defect (*arrow*) at the site of the lesion that was revealed by MR imaging.

**Fig. 5.** Light photomicrograph showing the hypocellular physaliphorous cells with a lobular growth pattern. The eosinophilic cytoplasm was vacuolated with myxomatous matrices. No mitosis or cellular pleomorphism is noted. H & E, original magnification × 200.
Ecchordosis physaliphora

Because of the lesion's small size and possible artifacts in the posterior fossa, CT scanning seems to have had limited utility in the detection of ecchordosis. Our case also showed no definite abnormalities on either a plain skull x-ray film or a CT scan (both with and without contrast enhancement). However, a magnified bone window CT scan revealed the osseous process as a tiny stalk. The osseous stalk is defined as a morphological hallmark of ecchordoses and does not exist in other lesions.2,3 Because of the lesion's small size and possible artifacts in the posterior fossa, CT scanning seems to have limited utility in the detection of ecchordosis. Our case also showed no definite abnormalities on either a plain skull x-ray film or a CT scan (both with and without contrast enhancement). However, a magnified bone window CT scan revealed the osseous process as a tiny stalk. The osseous stalk is defined as a morphological hallmark of ecchordoses and does not exist in other lesions.1,2,10 Furthermore, CT cisternography disclosed a small isolated filling defect that was probably contiguous with this subtle stalk. These characteristic radiological features seem to be sufficiently specific to distinguish this lesion from other pathological entities. If an ecchordosis is suspected, magnified bone window CT scanning combined with CT cisternography would be useful in determining the appropriate diagnosis.

Magnetic resonance imaging was of great use in detecting and precisely localizing the small ecchordosis physaliphora in the present case. It was situated near the midline axis of the clivus at the intradural space and had no bone involvement. The intradural space along the midline cranial axis is a common area for ecchordoses, and this location is one of the key factors in differentiating ecchordoses from other pathological entities.1,2,4,9 Chordomas usually occur in the extradural intraosseous portion, thus causing local bone destruction.3,5 Epidermoid and dermoid cysts frequently grow in the intradural space, but mainly at the cerebellopontine angle (CPA) cistern.9 The ecchordosis in our case appeared hypointense on a T1-weighted image and hyperintense on a T2-weighted image. This signal characteristic is common to all other reviewed cases, but it is nonspecific. A contrast enhancement study, however, provides important diagnostic information: addition of Gd-DTPA had no enhancing effects in any of the reviewed cases, possibly because of the paucity of vascular structures in the ecchordoses.10 In contrast, chordomas are commonly enhanced by the addition of contrast media in CT scanning and MR imaging.2,5,10 There have been a few reports of a purely intradural variant of chordomas.2,5,10 Although the differential diagnosis between intradural chordomas and ecchordoses is quite difficult, the former shows an enhancement effect whereas the latter does not. Therefore, the presence or absence of any enhancing effects is another useful factor in distinguishing ecchordoses from chordomas. Dermoid and epidermoid cysts may have a similar signal intensity to ecchordoses, but they occur chiefly in the CPA, as mentioned earlier.6 The radiological features of these pathological entities have been summarized in Table 2. Taken together, we suggest that the presence of an osseous stalk, an intradural location along the midline cranial axis, and the lack of enhancement by a contrast agent would be supportive for the differential diagnosis.

As a result of recent advances in neuroimaging systems, the odds of incidentally encountering ecchordoses should increase, even if they are small and clinically silent. We believe that this analysis will be of great help in diagnosing and differentiating this rare disease. However, the accumulation of more cases and clinical data will be necessary to establish the precise diagnostic criteria of ecchordosis physaliphora.

### Table 1

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Sex</th>
<th>Location</th>
<th>Plain X-Ray Film</th>
<th>CT Scan</th>
<th>CT Cisternogram</th>
<th>MR Image</th>
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<tr>
<td>Kurokawa, et al., 1988</td>
<td>M</td>
<td>C-2 level</td>
<td>slight hyperostosis</td>
<td>normal</td>
<td>shadow defect</td>
<td>hypo</td>
</tr>
<tr>
<td>Macdonald, et al., 1990</td>
<td>F</td>
<td>prepontine</td>
<td>ND</td>
<td>normal</td>
<td>ND</td>
<td>hypointense</td>
</tr>
<tr>
<td>Watanabe, et al., 1994</td>
<td>M</td>
<td>prepontine</td>
<td>slight hyperostosis</td>
<td>enlarged preprotine area</td>
<td>shadow defect</td>
<td>hyper</td>
</tr>
<tr>
<td>Akimoto, et al., 1996</td>
<td>F</td>
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<td>normal</td>
<td>normal</td>
<td>shadow defect</td>
<td>hyper</td>
</tr>
<tr>
<td>Present report</td>
<td>M</td>
<td>prepontine</td>
<td>normal</td>
<td>normal</td>
<td>shadow defect</td>
<td>hyper</td>
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* Gd = Gd-DTPA-enhanced image; hypo = hypointense; hypointense; ND = not described.

### Table 2

<table>
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<tr>
<th>Pathological Diagnosis</th>
<th>Usual Location (relation to)</th>
<th>Contrast Enhancing</th>
<th>Bone Involvement</th>
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<tr>
<td>ecchordosis chordoma</td>
<td>intradural</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td></td>
<td>midline</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td></td>
<td>midline (w/in clivus)</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td>intradural chordoma</td>
<td>intradural</td>
<td>yes</td>
<td>no</td>
</tr>
<tr>
<td>dermoid &amp; epidermoid</td>
<td>CPA</td>
<td>usually no</td>
<td>usually no</td>
</tr>
</tbody>
</table>

### References
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