A Unique Case Of Ecchordosis Physaliphora With Intratumoral Haemorrhage

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Citation

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Abstract

Ecchordosis physaliphora is a small, gelatinous tissue that is considered an ectopic notochordal remnant. This tissue is located in the midline of the craniospinal axis, reaching from the dorsum sellae to the sacrococcygeal region (1-4). Intracranial EP is typically found intradurally in the pre-pontine cistern, where it is attached to the dorsal wall of clivus via a small pedicle (1, 2, 5). Ecchordosis in this region are usually asymptomatic, and only a few studies have reported associated symptoms due to tumour expansion and compression of surrounding structures and extratumoral haemorrhage. To our knowledge retroclival EP with intratumoral haemorrhage has been described only once by Alkan et al. We report a case of retroclival EP with intratumoral haemorrhage in a 31 yr old female in which the diagnosis was based on CT and MR imaging.

INTRODUCTION

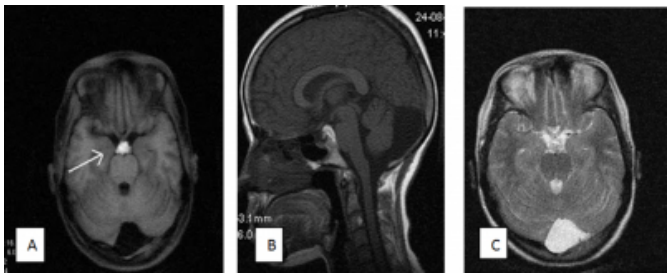
EP is a rare intracranial extra-axial mass derived from notochordal tissue that is typically located intradurally in the pre-pontine cistern and attached to the dorsal wall of clivus (1). Fetal notochordal tissues play a major role in the genesis of the axial skeleton and persist in the adult at the nucleus pulposus of the intervertebral disc (6). Due to a common origin from the fetal notochordal rests, EP and chordomas share common histological and ultrastructural characteristics (3, 7-11). Ecchordosis physaliphora is an extremely rare lesion. It is found incidentally in as few as 0.5-2% of autopsies (3, 8, 10, 11). The natural pathogenesis of the disease is characterized by slow subclinical progression with only rare ostensible manifestations. Symptoms, if occur, are the direct result of involvement of surrounding neurovascular structures and depend on the location of primary mass. EP should be differentiated from retroclival chordoma as accurate identification of nature of these masses has relevance in the determination of patient prognosis and in the planning of therapy.

CASE REPORT

A 31 yr old otherwise healthy woman presented to us with frequent headaches but no associated fever or vomiting. There was no history of blurring of vision, rhinorrhoea or ear discharge. Physical examination findings were normal and there were no neurologic deficits. Magnetic resonance (MR) images of the brain and computed tomographic (CT) scans of the head were obtained.

Figure 1

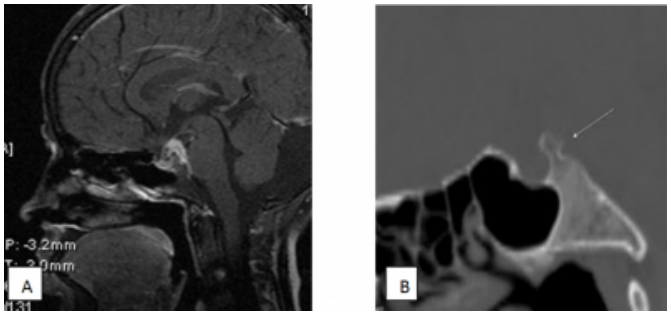
Figure 1: Axial T1-weighted (A) and Sagittal T1-weighted (B) and Axial T2-weighted (C) images showing a retroclival extra-axial mass with haemorrhage in pre-pontine cistern



Initial MR imaging of the brain revealed an intradural, well circumscribed, extra-axial retroclival mass located in the prepontine cistern. The mass appeared hyperintense on T2 and T1-weighted images, which likely represented haemorrhage, and restricted diffusion on diffusion-weighted images (Fig 1A, B, C). Contrast enhanced T1 weighted MR images were obtained and showed no enhancement (Fig 2A). On the T2-weighted image, the homogenously hyperintense signal intensity may have been less due to pulsation artifact in the lesion than in the pre-pontine CSF (Fig 2B). A possible diagnosis of Ecchordosis physaliphora was considered and sagittal CT scan with bone window settings was obtained. An osseous stalk arising from the clivus was well appreciated on sagittal CT scan with bone window settings (Fig 5). No bone destruction was noted.

Figure 2

Figure 2: Sagittal post contrast T1-weighted (A) MR image shows no contrast enhancement. A sagittal CT image bone window (B) showing an osseous stalk at the clivus.



On the basis of above findings, a diagnosis of Ecchordosis physaliphora with intratumoral haemorrhage was made.

DISCUSSION

EP is a rare, congenital, benign hamartomatous lesion arising from an ectopic notochordal remnant. This tissue is located in the midline of the craniospinal axis, reaching from the dorsum sellae to the sacrococcygeal region (1-4). The notochord is the primitive skeleton of vertebrates that forms the nucleus pulposus of the intervertebral disk at maturity. Ectopic notochordal rests can be seen outside the nucleus pulposus anywhere along the axial skeleton (1, 12). Intracranial EP is typically found intradurally in the pre-pontine cistern, where it is attached to the dorsal wall of clivus via a small pedicle (1, 2, 5). Recognition of the imaging features of EP is helpful in suggesting the diagnosis and differentiating from other retroclival lesions. CT is limited in detecting the EP due to its small size and beam hardening artefacts in the posterior fossa (20). However, the osseous stalk at the dorsal wall of the clivus on thin-section CT images is considered as the morphological hallmark of EP and does not occur in other retroclival lesions (5). MR is the best imaging modality for the radiological detection of EP, and the characteristic appearance of EP in MR is that of a well circumscribed, round, extra-axial mass in the pre-pontine cistern. EP demonstrated high signal intensity on T2-weighted images, low signal intensity on T1-weighted images, and no contrast enhancement.

The differential diagnosis of retroclival intradural lesions in the prepontine cistern includes chordoma, dermoid and epidermoid cysts, arachnoid cyst and Ecchordosis physaliphora (1, 13). A diagnosis of chordoma is less likely in our patient as these tumours usually produce symptoms that suggest brainstem involvement or cranial nerve palsies, that show bone destruction, that enhance with contrast material administration (1,13, 14). On MR images, dermoid and epidermoid cysts may have similar signal intensity to that of EP without contrast enhancement (15). However our lesion didn't suppress on fat suppressed sequences and restricted diffusion on diffusion-weighted images. The key imaging feature in this patient was the osseous stalk arising from the dorsal clivus. The absence of symptoms apart from headaches, midline location along the craniospinal axis, lack of contrast enhancement, hyperintense on both T1 and T2-weighted images with restricted diffusion on diffusion-weighted imaging, with no obvious bone destruction also favoured EP with intratumoral haemorrhage as the most likely diagnosis in this patient (5).

EP is a midline intradural lesion that shows no contrast enhancement, likely because of the paucity of vascular structures (1, 5, 17). In contrast, chordomas usually are seen in the extraosseous intradural portion of the clivus, causing local bone destruction (13, 16 radio). A purely intradural chordoma is rare and requires differentiation from EP (1, 13, 16-radio). Both the typical extradural chordoma and rare intradural chordoma show contrast enhancement, which is key feature in differentiating them from EP (1, 5, 13, 14, 16, 18, 19).

In conclusion, EP is a congenital malformation that needs to be considered in the differential diagnosis of retroclival lesions. In rare cases, it may be haemorrhagic or may be symptomatic due to tumor compression of the brain stem. When a retroclival mass is established on MRI, it should be differentiated from other retroclival lesions by using imaging features, especially an osseous stalk, as this is relevant in the patient prognosis and planning of therapy.

References

1. Wolfe JT 3rd, Scheithauer BW. "Intradural chordomas" or "giant ecchordosis physaliphora"? report of two cases. Clin Neuropathol 1987;6:98-103.
2. Lantos PL, Louis DN, Rosenblum MK, Kleihues P. Tumours of the nervous system. In; Graham DI, Lantos PL, eds. Greenfield's neuropathology. 7th ed. Vol2. London, England: Arnold, 2002; 767-1052.
3. Congdon CC. Benign and malignant chordomas: management challenge. J Neurosurg 1997; 86:182-189.
4. Rodriguez L, Colina J, Lopez J, Molini O, Cardozo J. Intradural prepontine growth:giant ecchordosis physaliphora? Neuropathol 1999; 19:336-340.
5. Toda H, Kondo A, Iwasaki K. Neuroradiological characteristics of ecchordosis physaliphora:case report and review of the literature. J Neurosurg 1998; 89:830-834.
6. Ulich TR, Mirra JM. Ecchordosis physaliphora vertebralis. Clin Orthop 1982; 163:282-289.
7. Sassin JF. Intracranial chordoma. In: Vinken, P.J., Bruyn G. W.(eds.), Handbook of Clinical Neurology. Vol.18. Amsterdam: North Holland, 9175:151-164.

8. Ho KL. Ecchordosis physaliphora and chordoma: a comparative ultrastructural study. Clin Neuropathol 1985; 4:77-86.
9. Willis RA. Chordoma. Pathology of tumours. London: Butterworths & Co, 1967:937-943.
10. Wyatt RB, Schochet Jr SS, Mc Cormick WF. Ecchordosis physaliphora. An electron microscopic study. J Neurosurg 1971; 34:672-677.
11. Rich TA, Schiller A, Suit HD, Mankin HJ. Clinical and pathologic review of 48 cases of chordoma. Cancer 1984; 56:182-187.
12. Macdonald RL, Cusimano MD, Deck JH, Gullane PJ, Dolan EJ. Cerebrospinal fluid fistula secondary to ecchordosis physaliphora. Neurosurgery 1990; 26:515-519.
13. Katayama Y, Tsubokawa T, Hirasawa T, Takahata T, Nemoto N. Intradural extraosseous chordoma in the foramen magnum region: case report. J Neurosurg 1991; 75:976-979.
14. Mehnert F, Beschorner R, Kuker W, Hahn U, Nagele T. Retroclival ecchordosis physaliphora: MR imaging and review of the literature. AJNR Am J Neuroradiol 2004; 25:1851-1855.
15. Vinchon M, Pertuzon B, Lejeune JP, Assaker R, Pruvo JP, Christiaens JL. Intradural epidermoid cysts of the cerebellopontine angle: diagnosis and surgery. Neurosurgery 1995; 36:52-57.
16. Mapstone TB, Kaufman B, Ratcheson RA. Intradural chordoma without bone involvement: nuclear magnetic resonance (NMR) appearance- case report. J Neurosurg 1983; 59:535-537.
17. Akimoto J, Takeda H, Hashimoto T, Haraoka J, Ito H. A surgical case of ecchordosis physaliphora [in Japanese]. No Shinkei Geka 1996; 24:1021-1025.
18. Yuhi F, Asakura T, Tomosugi T, Kusumoto K, Awa H, Uetsuhara K. A case of clival chordoma without bone destruction [in Japanese]. No Shinkei Geka 1986; 14:547-552.
19. Erdem E, Angtuaco EC, Van Hemert R, Park JS, Al-Mefty O. Comprehensive review of intracranial chordoma. Wis Med J 1992; 91:627-628.
20. Alkan O, Yildirim T, Kizilkilic O, Tan M, Cekinmez M. A case of Ecchordosis Physaliphora presenting with an intratumoral haemorrhage. Turkish Neurosurgery 2009. Vol 19, No.3, 293-296.

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