Ecchordosis physaliphora

History and etymology

Hubert von Luschka (1820-1875), first described the finding of pathologic ectopic notochordal tissue at the posterior clivus in 1856.

Ecchordosis physaliphora is a congenital benign hamartomatous lesion derived from notochord remnants, usually located in the retroclival prepontine region, but can be found anywhere from the skull base to the sacrum.

There has been some controversy as to whether intradural chordoma and large ecchordosis physaliphora are different entities. Some authors (such as Wolfe et al.) proposed the name 'intradural chordoma' for all intradural notochordal remnant lesions [1]. Others (such as Rodriguez et al.) proposed that all intradural notochordal remnant lesions should be called ecchordosis physaliphora, until chordoma are pathologically proven to arise from the intradural compartment [2]. However, they are currently considered distinct pathologies with common origin.

Unlike chordomas which are often symptomatic due to brainstem or cranial nerve compression, patients with ecchordosis physaliphora are usually asymptomatic. They are found in ~2% of autopsies [3].

Pathology

Ecchordosis physaliphora arise from remaining notochord cells along the axis of the spine after embryogenesis. Unfortunately, ecchordosis physaliphora and chordoma are histologically indistinguishable, other than by examining the margins, the latter demonstrating infiltrative growth.

Radiographic features

CT

CT is generally not sensitive for such lesions, mainly because of posterior fossa artifacts and the near CSF density of the mass. The bony clival defect is however visible as a well demarcated smoothly corticated region, without aggressive features.

Occasionally an osseous stalk is seen at the base of the lesion which is said to be pathognomonic in this context.

MRI

Mehnert et al., reviewed 300 consecutive 1.5-T MR imaging studies that included thin-section transverse T2-weighted images of the skull base for the presence of a retroclival EP. In cases in which an EP was identified, two neuroradiologists observed MR signal intensity characteristics, contrast enhancement, size, form, stalk of EP, and signal intensity changes of the adjacent clivus.

Five cases with retroclival EP were found (incidence, 1.7%). In all cases, the ecchordoses was hyperintense on T2-weighted images and hypointense on T1-weighted images. Contrary to the reported findings in chordomas, none of the lesions showed contrast enhancement. In four cases, there were signal intensity changes in the adjacent clivus. A stalklike connection between clivus and EP was seen in three patients.

Because of the benign character of EP and the difficulties in its histopathologic differentiation from chordomas, precise knowledge of the radiologic characteristics of EP is important. On the basis of these five cases and a review of literature, contrast enhancement and the presence of clinical symptoms seem to be highly reliable parameters in the differential diagnosis of intradural chordoma and EP [4)].

A stalk-like connection to the clivus is usually seen if high resolution images are obtained.

Apart from the characteristic location (retroclival, prepontine, and intradural), MRI findings are not specific, with signal similar to CSF:

T1: hypointense

T2: hyperintense

T1 C+ (Gd): variable, however most cases have not shown substantial enhancement [5)].

Case reports

Raffa et al. presented the rare case of a seven-year-old patient with atypical presentation and neuroradiological features of giant ecchordosis physaliphora. The patient underwent cross-sectional imaging due to persistent headaches without neurological or visual symptoms. CT scan imaging of the head revealed a hypodense tumor in the prepontine cistern. This lesion caused smooth scalloping of

the dorsal clivus without aggressive erosion or calcification, and an osseous stalk was also identified between the lesion and the dorsal wall of the clivus. Sagittal T1 weighted image (T1WI) MRI showed a bilobed, solid and cystic, well-defined lesion, measuring 3.5 cm in terms of craniocaudal diameter, found alongside the midline within the prepontine cistern. After the evaluation of radiological images, the patient was then subjected to endoscopic transnasal complete tumor excision. Histological examination revealed sheets and lobules of clear cells with cytoplasmic globules “physaliphorous cells”, and myxoid stroma. There was nuclear pleomorphism associated with focal areas of necrosis. After full recovery and discharge, the patient was followed up for the first year with four-month interval brain MRI scans showing no evidence of residual tumors. On the 12 months follow-up scan, the MRI scan revealed a 1.5 x 0.7 cm recurrent mass in the retroclival right paramidline prepontine cistern, which was most notably seen on the diffusion-weighted images. A Series of proton beam therapy with annual MRI scans demonstrated regression of the tumor, eventually allowing the patient to live free of neurological symptoms up to this day. Results suggest that the utilization of radiological imaging such as CT and MRI scans was successful in identifying the ecchordosis physaliphora and differentiating it from chordomas. It can also be inferred that atypical radiological and histopathological findings of ecchordosis physaliphora lesions might suggest that they are prone to recurrence, which is an atypical feature for such entities. Further studies are recommended to explore and better understand these uncommon observations in patients with ecchordosis physaliphora [6].

Ghimire et al., described the case of a 65-year-old lady who presented with mutism and a right hemiparesis. Imaging showed a severe spontaneous tension pneumocephalus. The cause was diagnosed as Ecchordosis physaliphora (EP). EP is a rare cystic congenital hamartomatous benign notochordal tumor (BNCT) arising from an ectopic notochordal remnant. To the authors' knowledge, this is the first case of EP to be described in the literature which presented with a life-threatening but treatable condition of severe tension pneumocephalus [7].

Stahl-Hoffmann et al., report a case of symptomatic ecchordosis physaliphora (EP) in a 34-year-old woman who presented with progressive diplopia due to palsy of the left sixth cranial nerve. Repeated magnetic resonance imaging (MRI) disclosed typical characteristics of a congenital EP lesion with compression of the left abducens nerve presumably because of a secondary herniation of the arachnoid mater. They performed an augmenting combined recess resect procedure on the left eye. No progression of the lesion was observed over a period of 5 years. For differential diagnostics an EP has to be distinguished from skull base tumors, such as chordoma and chondrosarcoma [8].

A 78-year-old man, who died due to heart infarction. The 6-mm asymptomatic gelatinous lesion was fixed to the basilar artery on its ventral aspect. Small EPs can be easily overlooked in autopsy. Ecchordosis physaliphora and intradural chordoma share some similarities that may be misleading and may even result in the wrong diagnosis and therapy. The recently reported new entity BNCT poses a similar problem [9].

A 60-year-old man presenting with memory loss underwent magnetic resonance imaging, which revealed an intradural retroclival mass without bone involvement.

The patient underwent an endoscopic transsphenoidal-transclival procedure with subtotal removal of the tumor. Histological findings confirmed the diagnosis of a chordoma.

Even if some parameters exist for a differential diagnosis, ecchordosis physaliphora and intradural chordoma may represent different aspects of the spectrum of the same pathology. Intradural clival chordomas have a better prognosis with respect to classic chordomas. Therefore, in subtotal removal such as that performed in our case, postoperative radiation therapy should be performed only if a regrowth of the remnant is seen during neuroradiological follow-up [10].

A case of 22-year-old man who presented with headache and confusion. MR imaging and CT revealed intracranial ecchordosis physaliphora associated with intratumoral hemorrhage and vasogenic edema. The neurological findings resolved completely after medical therapy [11].

Srinivasan A, Goyal M, Kingstone M. Case 133: Ecchordosis physaliphora. Radiology. 2008 May;247(2):585-8. doi: 10.1148/radiol.2472050101. PubMed PMID: 18430884 [12].

Rotondo et al., reported a case of ecchordosis physaliphora, intradurally located in the prepontine cistern, that unusually presented associated with symptoms. MRI detected and precisely located the small mass. At surgery, a cystic gelatinous nodule was found ventral to the pons, contiguous with the dorsal wall of the clivus via a small pedicle. Histological examination diagnosed the lesion as an ecchordosis physaliphora [13].

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