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CASE REPORT

Ecchordosis Physaliphora Masquerading as Chordoma: A Case Report

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INTRODUCTION

A notochordal rest is a remnant of the embryonic skeletal column and later forms the nucleus pulposus.[1] It is considered ectopic when located outside the nucleus pulposus.[1] Ecchordosis physaliphora and chordoma are examples of an ectopic notochord remnant.[2]

Although ecchordosis physaliphora and chordoma share similar biological behaviour, they are two separate entities that require different management.[3] Differentiation between them is essential. Ecchordosis physaliphora is benign and self-limiting and does not require surgical intervention.[4] In contrast, chordoma is malignant, exhibits local aggressive behaviour and bony destruction, and requires radical resection as well as radiotherapy.[4]

Ecchordosis physaliphora is a classic benign intradural lesion attached to the clivus by an osseous stalk. Patients are often asymptomatic. Chordoma is a malignant counterpart that exhibits aggressive features and is located extradurally. Intradural chordoma is rare and reported in only few cases. Patients with chordoma often have cranial nerve palsy at the time of presentation and have a dismal prognosis despite surgical intervention and radiotherapy.[3]

CASE REPORT

A 43-year-old lady presented with intermittent non-specific bitemporal and occipital headache for the past 10 years. The pain was not severe and was relieved by rest and sleep. Cranial nerves were intact and neurological examination was unremarkable. A diagnosis was made of tension headache. Nonetheless computed tomography (CT) was performed to exclude any sinister pathology and to reassure the patient.

Incidental findings on CT imaging revealed a retroclival bony defect with no visible stalk. The defect was confined to the retroclival region without extension to adjacent structures, i.e., sphenoid sinus anteriorly, sellar structures superiorly or bilateral internal carotid arteries laterally (Figure 1).

Magnetic resonance imaging (MRI) of the brain was performed to further characterise the lesion. This lesion returned homogeneous hypointensity on T1-weighted images and hyperintensity on T2-weighted images

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Ethics Approval: The patient was treated in accordance with the Declaration of Helsinki. Informed consent was obtained from patient for the examinations.

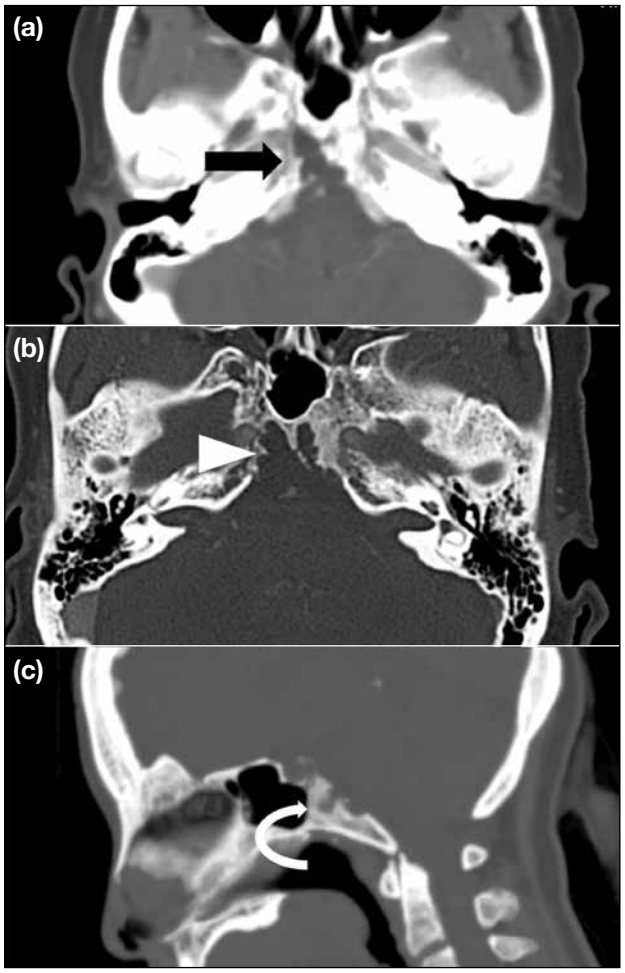

Figure 1. Computed tomography of the brain. (a) There was an isodense lesion with no internal matrix calcification seen extending into the pre-pontine cistern (arrow). It is associated with (b) bony defect (arrowhead) with (c) no visible stalk identified (curve arrow).

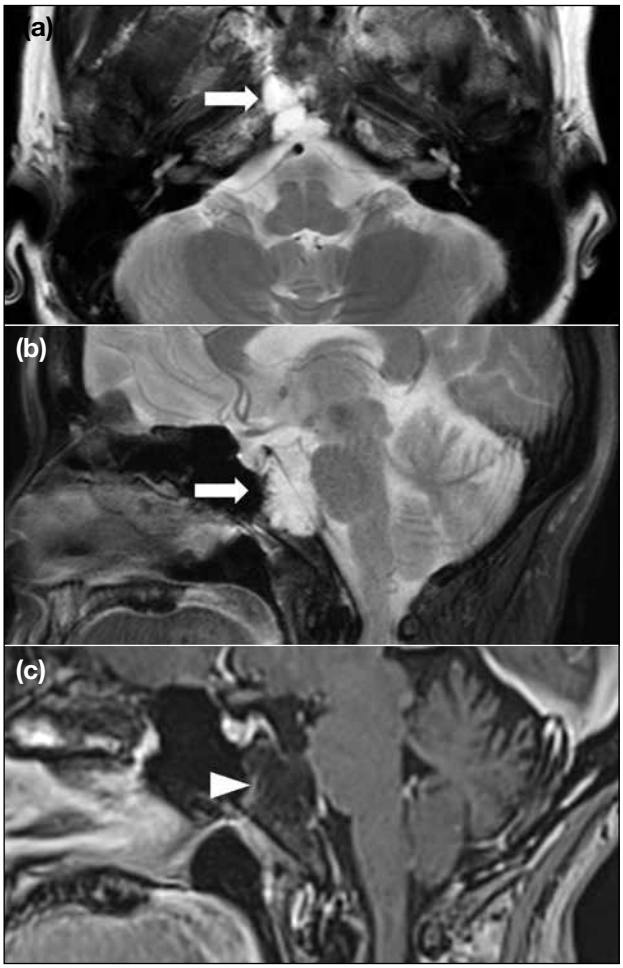

Figure 2. Magnetic resonance image of the brain. A homogenous retroclival lesion demonstrated homogenous hyperintense signal on T2-weighted image (arrow) in (a) an axial view and (b) a sagittal view. (c) This lesion showing hypointense signal on T1-weighted image without enhancement in post-gadolinium image (arrowhead).

without contrast enhancement post-gadolinium administration. It was located in the upper two thirds of the retroclival region with protrusion into the pre-pontine cistern (Figure 2). CT and MRI findings were suggestive of ecchordosis physaliphora although other differential diagnoses such as cystic chordoma, metastasis, or abscess were included in the report. The presence of normal blood parameters in the absence of fever and no history of primary malignancy excluded the possibility of metastasis or abscess.

In view of the differential diagnoses and the possibility of a benign or malignant lesion, the patient was counselled for transsphenoidal clival biopsy to confirm the diagnosis. Initial histopathology was consistent with chordoma. Nonetheless due to discrepancy between the intraoperative and pathological findings, the neurosurgeon asked the pathologist to re-review the tissue sample. In the neurosurgeon's opinion, the bony hard lesion was not consistent with chordoma, instead favouring a congenital lesion such as ecchordosis physaliphora. After re-evaluation, the final histopathological diagnosis was amended accordingly (Figure 3). In addition, the pathologist emphasised the importance of radiological findings in differentiating ecchordosis physaliphora from chordoma due to the challenges of histological interpretation.

Postoperatively the patient developed cerebral spinal fluid leakage and two further endoscopic repairs were

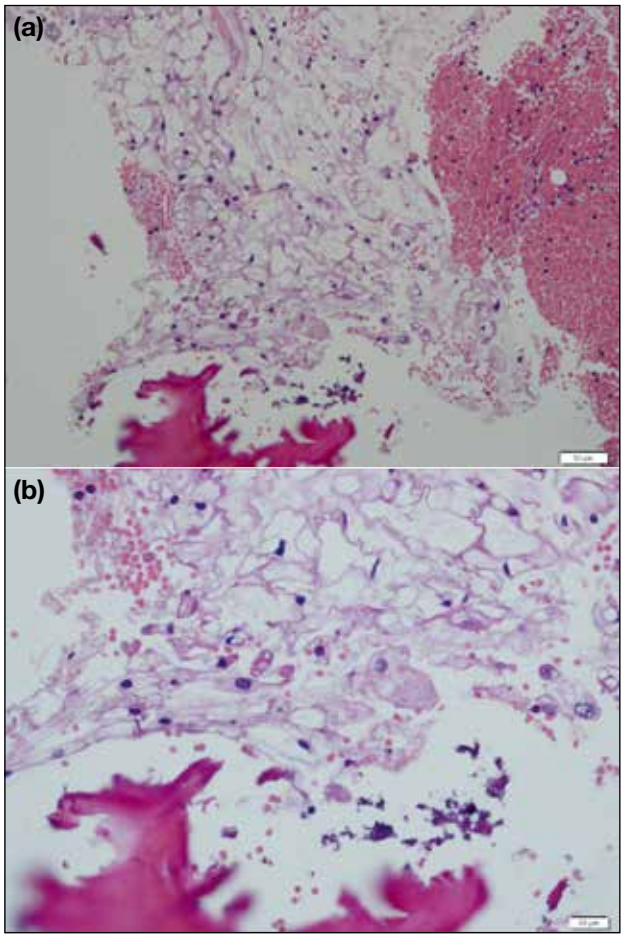

Figure 3. (a: 100x, b: 400x) Most biopsy fragments appeared as normal bone trabecula with normal haematopoietic elements located within the marrow spaces. There were only a few tiny clusters of clear-looking cells that vaguely resembled the physaliferous cells with small uniform round nuclei and no nuclear pleomorphism, increased mitosis, or atypia. These appeared to be lying either free or within the marrow spaces without destruction of adjacent tissues. There was absence of any lobular proliferation of such cells with the attendant cellular pleomorphism, tissue destruction and necrosis, and accompanying inflammatory reaction that would be expected with chordoma. Immunohistochemistry showing cell clusters positive to cytokeratin AE1/AE3, EMA, and to S100 protein. Whilst these markers are positive for chordoma, they are also known to be positive for other lesions of the notochord including ecchordosis physaliphora. The latter was favoured over chordoma because of the absence of cellular proliferation forming tumour lobules and absence of tissue destruction, necrosis, and inflammation.

required during the same admission. She was discharged home well after the second procedure and remained asymptomatic at her last neurosurgical follow-up.

DISCUSSION

Notochords are phylogenetically considered to be the primitive skeleton of vertebrates. As development of the axial skeleton progresses, the notochord becomes the nucleus pulposus.[3] Intriguingly, at the extreme poles of the axial skeleton (dorsum sellae and sacrococcygeal region), the outcome of notochord development is more variable and correlates with the presence of aberrant notochord elements in later life.[3]

Notochordal remnants were reported in 2% of random autopsies by Rippert in 1894, who named these lesions as ecchordosis.[5] Ecchordosis physaliphora is a benign ectopic notochordal remnant along the midline of the craniospinal axis.[6] Ecchordosis physaliphora is a rare small benign hamartomatous lesion located intradurally, and shows a slow-growing pattern.[3] It is attached to the clivus by a small pedicle and associated with bony defect at the retroclivus.[3] The malignant counterpart is chordoma, arising extradurally and associated with extensive bony erosion.[6] There are some rare cases of extraosseous intradural chordoma and this subtype is believed to have a better prognosis than the classic one.[5]

Patients with ecchordosis physaliphora are asymptomatic due to its small size and indolent growth rate.[3] There are exceptional rare cases of symptomatic ecchordosis physaliphora where it has expanded to an unusually large size and manifests as a mass effect by compressing adjacent structures.[6] An atypical extratumoural bleed may also be encountered occasionally.[6] Our patient reported only intermittent non-specific headache that had not worsened over the years. She had no other signs of increased intracranial pressure, cranial nerve palsy, or neurological deficit. These symptoms may be attributed to the disease itself but and are most likely secondary to tension headache rather than symptoms of an ecchordosis physaliphora.

Unlike ecchordosis physaliphora, which is typically retroclival in origin, chordoma is located centrally within the clivus.[7] It causes extensive lytic bony destruction and intratumoural calcification.[7] Most individuals present with symptoms such as headache and cranial nerve palsies.[3] These symptoms are due to local bony destruction and mass effect on the adjacent brainstem and cranial nerves.[3]

Although chordoma has a lower prevalence compared with ecchordosis physaliphora, their differentiation is of great clinical concern as patients require very different management strategies.[7] The former, due to its aggressive nature, often necessitates radical resection and adjuvant radiotherapy, whereas the latter rarely requires surgery.[7]

As our radiological report included either diagnosis of ecchordosis physaliphora or chordoma, the patient underwent clival biopsy to exclude the later.

Although both ecchordosis physaliphora and chordoma share a similar notochordal origin, histopathologic differentiation between the two is challenging.[4] Nishiguchi et al[4] subjected 38 patients initially diagnosed histopathologically with chordoma to be re-evaluated by two separate radiologists and two separate pathologists. In the consensus, five patients were found to have a benign notochord cell tumour. They are difficult to distinguish based on histopathology, immunohistochemistry and ultrastructural studies, instead relying on the infiltrating growth of chordomas.[7] A few authors have proposed differentiation on the basis of hypocellularity, sparse pleomorphism and absence of mitoses, but these are not definitive criteria.[8] In our patient, the final diagnosis of ecchordosis physaliphora was made instead of chordoma due to the absence of cellular proliferation, tissue destruction, or necrosis inflammation. Nonetheless the pathologist also emphasised the importance of radiological findings that were equally important when distinguishing the two due to the dilemma in histological interpretation. What role does imaging contribute to this diagnostic conundrum?

CT and MRI remain important when assessing a clival lesion. CT poses limitations in detecting ecchordosis physaliphora in view of the often small size of the lesion and beam hardening artefacts in the posterior fossa.[6] Nonetheless if an osseous stalk is identified connecting to the clival notochord remnant, it is considered a morphological hallmark of ecchordosis physaliphora.[3] Chordoma, on the contrary, will show extensive bony destruction and tumoural calcifications.[1] Although CT findings in our patient showed no visible osseous stalk or tumoural calcification, the MRI findings were typical and consistent with ecchordosis physaliphora. The lesion was hypointense on T1-weighted images, hyperintense on T2-weighted images, and there was no visible contrast enhancement in post-gadolinium study. Nonetheless due to the rarity of this congenital lesion, most radiologists including ours are not familiar with its appearance and signal characteristics.

Based on all cases reported in the literature, ecchordosis physaliphora (including symptomatic lesion) demonstrates MRI signal characteristics similar to those seen in our patient. The presence or absence of contrast enhancement has been repeatedly shown to be helpful in determining and distinguishing ecchordosis physaliphora from chordoma.[6] Ecchordosis physaliphora does not demonstrate enhancement in post-contrast images, whereas chordoma usually enhances considerably in post-gadolinium study.[9] This reflects the usual histopathological features of some degree of vascular proliferation in the latter, as opposed to scant vascular networks in the former.[4] Hence, lack of enhancement is a very useful criterion for differentiating ecchordosis physaliphora from chordoma or other malignant tumour including rare atypical intradural chordoma.[5]

CONCLUSION

In view of the difficulties in differentiating ecchordosis physaliphora and chordoma based on histology alone, precise knowledge of neuroradiological (specifically magnetic resonance) imaging characteristics is especially important in making the diagnosis. This case report emphasises the significance of observing the absence of contrast enhancement, with T1 and T2 lengthening similar to that of cerebral spinal fluid, and retroclival location as classic features in clinching the diagnosis of ecchordosis physaliphora. In doing so, the patient is spared an unnecessary surgical procedure, with the attendant risks of potential complications.

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