



E-QUID: ANSWER / *Neurology*

Cervical chordoma with moderate bone impairment in a child. Answer to October e-quid[☆]

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Case report

A 4-year-old boy, without personal antecedents, born at term after a normal pregnancy, presented progressive right hemiparesis over one month. He was referred for a brain and medulla MRI (Fig. 1), completed by a CT scan (Fig. 2). The diffusion sequence was negative.

DOI of original article: <http://dx.doi.org/10.1016/j.diii.2012.04.011>.

[☆] Here is the answer to the case previously published in the n° 10/2012. As a reminder we publish again the entire case with the response following.

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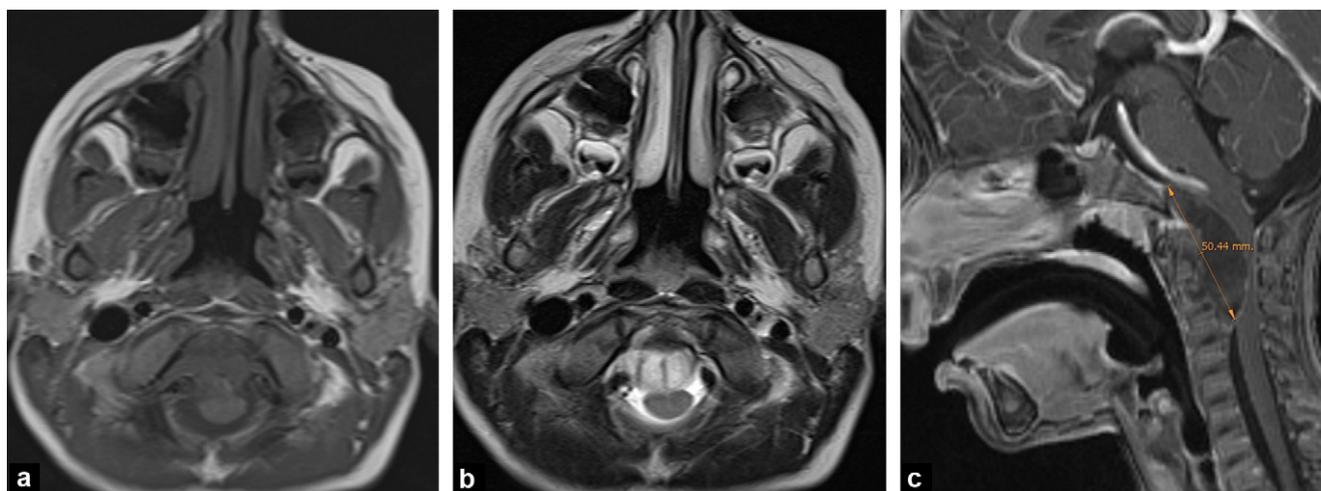


Figure 1. Brain and medullary MRI. a) Axial T1 sequence without contrast. b) Axial T2 sequence. c) Sagittal T1 sequence after injection of gadolinium.

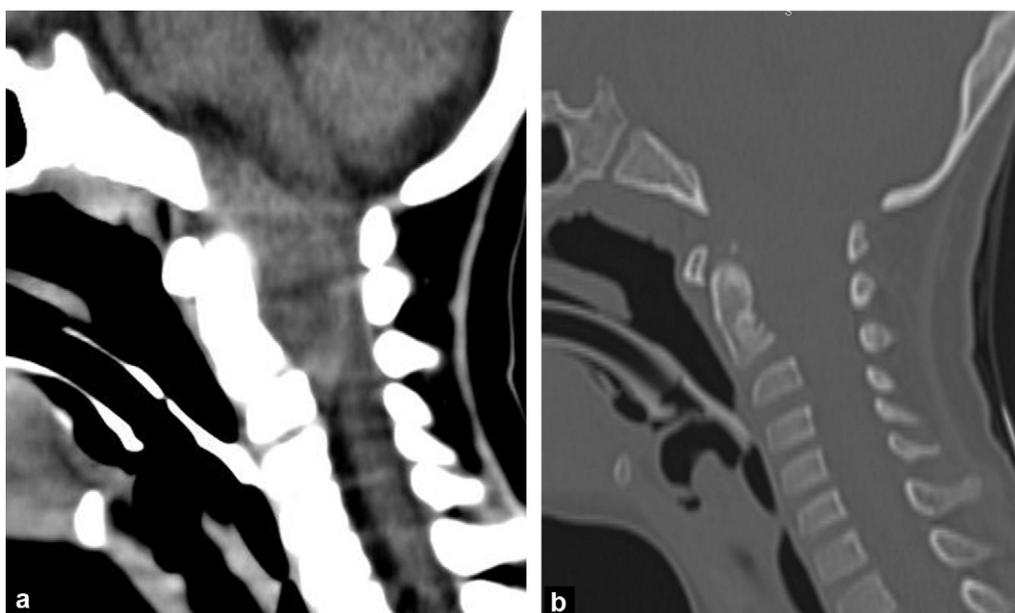


Figure 2. CT scan without injection, sagittal sections. a) Soft window, b) Bone window.

What is your diagnosis?

After reading the case report, which diagnosis would you choose from the following proposals:

- epidermoid tumour;
- chordoma;
- chondrosarcoma;
- lymphoma;
- metastasis.

Diagnosis

Cervical chordoma with moderate bone impairment in a child.

Comments

The examinations carried out found a mass at the medullary canal extending from the clivus to vertebral body of C3, through the foramen magnum, and moderate impairment of vertebral body of C2. This mass is extradural anterior: it pushes the spinal cord back, reduces the sub-arachnoid spaces and enlarges the medullary canal. It is responsible for the compression of the cervical spinal cord and the medulla oblongata, without sign of myelopathy on the MRI. On the MRI (Fig. 3), the mass moderately hypointense on T1-weighted images, hyperintense on T2-weighted images, without restriction of the diffusion and is moderately enhanced at the periphery after injection of the contrast product. Heterogeneity of the signal from the adjacent bone is noted. On CT scan (Fig. 4), the tumour is well

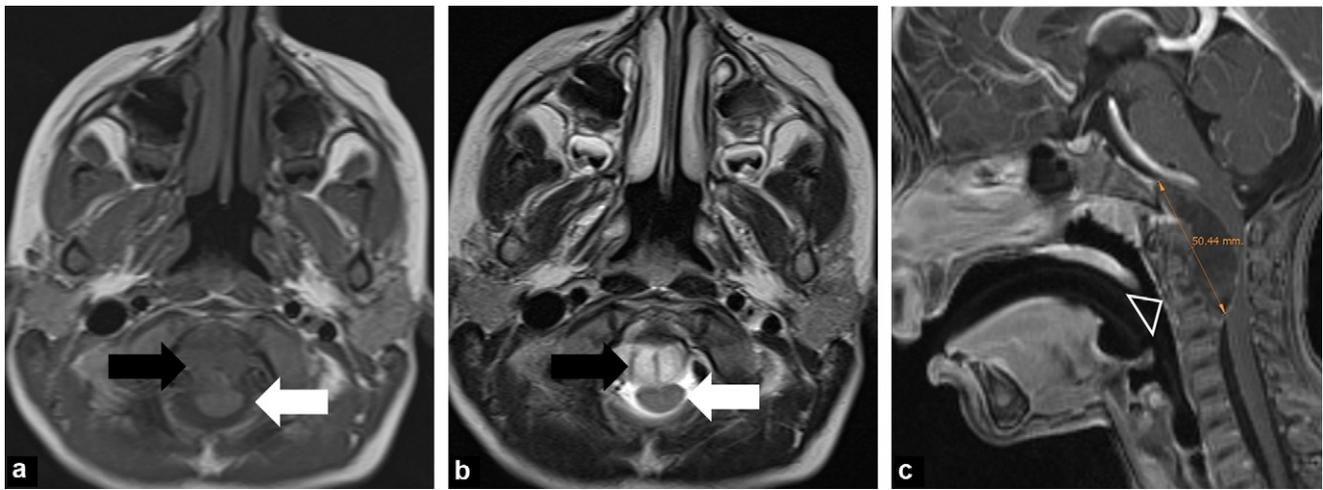


Figure 3. Brain and medullary MRI: anterior extradural intracanal mass (black arrow), pushing the spinal cord back (white arrow) and in contact with C2 vertebra (tip of arrow).

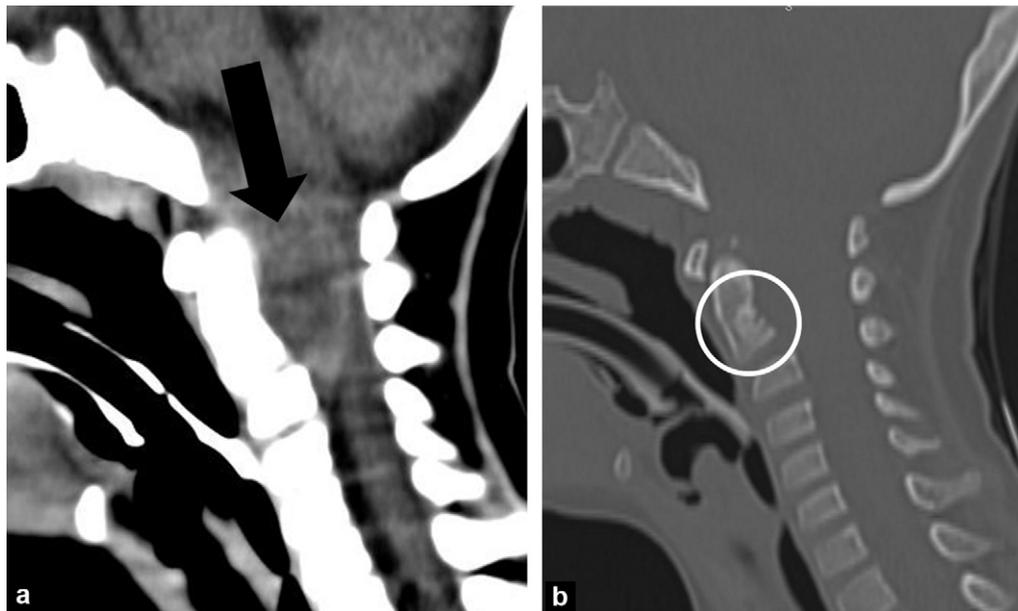


Figure 4. Sagittal unenhanced CT scan: the same heterogenous intracanal mass is found (black arrow) with erosions of C2 cortical bone and several osteocondensent zones (white circle). Enlargement of the medullary canal.

defined, heterogeneous, with hypo and hyperdense zones. A deformation of C2 vertebral body and dens is noted with cortical osteolysis and an adjacent osteocondensing zone. This bone impairment is very moderate compared with the size of the mass.

Discussion

Chordomas are rare (4% of primary bone tumours), malignant, slow growing, locally aggressive although rarely metastatic [1–3]. They develop out of intraosseous remains of the primitive notochord [4], generally at its extremities in proportions estimated at 25–35% at the base of the skull, 50 to 60% at the sacrococcygeal level, 15–25% in the cervico-dorso-lumbar vertebral bodies [2,5–7]. However, one study

evaluates this distribution at a third at each level [1]. Chordomas are rarely only intraosseous and frequently extend to the epidural and perivertebral space by enlarging the intervertebral foramen, and less frequently the intervertebral discs [4,6,8]. The chordomas of the cervical spine account for 20 to 50% of the spinal locations, with impairment of C2 vertebral body most often reported, probably due to the larger size of the vertebra [6]. Rare locations outside of the axial skeleton (nasopharynx, intradural...) [2] and extraosseous locations [5,6,8] have been described. The mean age of the patients presenting chordoma is 30 to 50 years [2,6]. The sex-ratio is two men for one woman [1,2]. This tumour is extremely rare in the child, where it is generally located at the bottom of the skull, with an extension facilitated through the foramen magnum [7]. Signs and symptoms vary according to the location, but are often chronic [6]:

- cervical spinal location: cervical pain, pain at the shoulders, arms and muscular weakness, especially in case of associated myelopathy, dysphagia, respiratory difficulties [4,6];
- retroclival location: ophtalmoplegia (proximity of cranial nerves III, IV and VI) or chronic fronto-orbital headache, or even impairment of other lower cranial nerves [2].

The imaging assessment is based on the CT scan and MRI, both efficient in detecting the tumour. However, the MRI better detects the extension, especially at the level of the central nervous system [1]. Classically, the MRI finds a heterogenous mass with iso/hypo signal intensity on T1-weighted images (bone) and in hyperintense on T2-weighted images (with possible partitions in hyposignal). In T2*, hypodense elements may be visualised (blood, calcifications). After injection of gadolinium, an enhancement is generally visualised although of variable intensity, sometimes in a honeycomb pattern (related to the bone partitions) [1,2,4,7]. The retroclival locations achieve the classic "thumbprint" on the pons of the brain stem. The T1-weighted sequence with fat saturation better detect the contrast enhancement. CT scan shows, a central well-defined, globally hyperdense mass with hyper- and hypodense zones (fragments of bone within the tumoral matrix) and very often lytic lesions of bone [1,2,4,6]. The enhancement is heterogeneous [2,4,6]. For retroclival chordomas, vascular imaging should be carried out due to the risk of arterial invasion, by an angio-MR, CT arteriography or digital subtraction arteriography, if necessary [1,2,6]. The interest of our case resides in the atypical presentation of this tumour, first since it is exceptionally rare in children, and second due to its extradural location with very limited bone impairment. The differential diagnoses are:

- epidermoid tumours: they are generally intradural. They do not induce any modifications in the adjacent bone, are not enhanced after injection of gadolinium and have a high diffusion signal with drop in ADC [2];
- lymphoma: the intercanal spinal location of primary lymphoma is very rare. It generally consists of a more homogenous tumour, with homogenous enhancement and restriction of the diffusion;
- chondrosarcoma: it is generally not median on the clivus but develops along the lateral edges, in the petro-occipital fissure. However, the signal is identical to that of the chondroma and may present chondroid calcifications (arc-shaped, globular or linear) [1,2,7]. Like chordoma, chondrosarcoma is very rare in young children [2];
- plasmocytoma: the signal in T2 weighted sequence is generally lower and more homogenous than that of the chondroma. It is also osteolytic. The mean age is higher (50–90 years) [2];
- metastases: they generally are lesions that destroy the bone with extension to the soft parts, with known neoplasm [2];
- for chordomas with extension through the intervertebral foramen, it is also necessary to mention tumours of the neural sheath such as schwannomas [4,6];
- finally, ecchordosis phylasiphora is a benign ectopic notochordal tissue in intradural retroclival situation, found in 2% of the autopsy series [3,5], although the imaging is typical: T1 isosignal, T2 hypersignal without enhancement, with presence of osseous stalk [9].

The histopathologic features of chordoma are typical, showing phylaphorous cells [2,4]. Two types of histology are distinguished: the typical chordoma and the chondroid chordoma, which is more frequent in children and has a better prognosis [2,7]. The treatment is preferably surgical, often completed by proton therapy [1,2,4,7]. The prognosis: 60 to 70% survival for 5 years without recurrence after surgical treatment and radiotherapy [2].

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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