Skeletal Radiology (2020) 49:819–820
https://doi.org/10.1007/s00256-019-03311-z

TEST YOURSELF: ANSWER

Chronic low back pain

Rania Zeitoun[1] · Petra Balogh[2] · Amir Amiri[3] · Alex Gibson[3] · Asif Saifuddin[4]

Published online: 15 November 2019

Diagnosis

Lumbar vertebral body benign notochordal cell tumor presenting as an ivory vertebra.

Discussion

Benign notochordal cell tumors (BNCT) are vertebral body lesions of notochordal origin. The notochord is an embryonic midline structure that plays a pivotal role in vertebral development, contributing to formation of the nucleus pulposus. A variety of lesions of notochordal origin arise along the axial skeleton, consecutively described since the first recognition of such a lesion by Virchow in 1846, named 'chordomata'. He was also the first who applied the term 'physaliphorous' in reference to the characteristic vacuolated cells [1]. By now, from the skull base to the coccyx, the following lesions can be distinguished: notochordal nests, ecchordosis physaliphora spheno-occipitalis (EP)—distinctively present at this anatomical site—and other lesions such as BNCT and chordoma [2, 3]. All lesions show identical immunoprofiles regardless of their biological behavior and are positive with epithelial markers and brachyury.

Under microscopic examination, BNCTs show sheets of physaliphorous cells with adipocytic-like features, lacking intercellular myxoid matrix, significant cytological atypia, mitotic activity, and necrosis. BNCT do not have the permeative growth within bone that characterizes their malignant counterpart, chordoma [4].

BNCTs are becoming more commonly recognized on imaging studies, having previously been described as intraosseous giant notochordal hamartomas, or giant notochordal rests [3, 5]. BNCTs have been found in 20% of autopsy cases, mostly involving the sacro-coccygeal vertebrae.

BNCTs are typically focal lobular lesions located centrally within the vertebral body, measuring from a few millimeters up to 3.5 cm [6]. They typically show hypointense T1-weighted (T1W) and hyperintense T2-weighted (T2W) and short tau inversion recovery (STIR) signal intensity (SI) on magnetic resonance imaging (MRI). They usually show corresponding mild sclerosis on radiographs and computed tomography (CT) with preserved trabecular pattern [5–7]. Unlike their malignant counterpart chordoma, BNCTs do not enhance and are not associated with osteolysis, vertebral collapse or soft tissue extension [6]. However, the coexistence of BNCT and chordoma has been described [8].

Our case demonstrated diffuse sclerosis of the L4 vertebral body on radiographs (Fig. 1) resulting in an 'ivory vertebra'. CT confirmed diffuse medullary sclerosis without expansion or collapse (Fig. 2), with maintenance of the endplates and adjacent disc spaces. Bone scintigraphy showed no abnormal uptake. MRI performed at presentation and at 17-month follow-up showed diffuse heterogeneous hypointense SI on T1W SE and mild hyperintensity on T2W FSE and STIR images (Fig. 3a, b, and c).

Due to on-going symptoms and a lack of definite diagnosis, a CT-guided biopsy was performed. Histological examination demonstrated a core of bone with the medullary spaces filled with large, polygonal cells with abundant clear and focally vacuolated cytoplasm, with bone remodeling (Fig. 4a). On immunohistochemistry, the cells were diffusely positive for MNF116 and brachyury (Fig. 4b). A diagnosis of benign notochordal cell tumor was made, with no evidence of transition

The case presentation can be found at https://doi.org/10.1007/s00256-019-03310-0

✉ Rania Zeitoun
raniazeitoun@gmail.com

[1] Imaging Department, Kasr Al-Ainy Faculty of Medicine, Cairo University, Cairo, Egypt

[2] Histopathology Department, Royal National Orthopaedic Hospital NHS Trust, Stanmore, UK

[3] Spinal Surgery Department, Royal National Orthopaedic Hospital NHS Trust, Stanmore, UK

[4] Imaging Department, Royal National Orthopaedic Hospital NHS Trust, Stanmore, UK

to chordoma. The currently described case would appear to represent a very rare presentation of BNCT as an 'ivory vertebra', with only a single case previously described in a literature review of all published cases to 2011 [1].

Atypical vertebral hemangioma and Paget's disease are major considerations in the differential diagnosis. BNCTs and hemangioma share high T2W SI and sclerotic trabeculae. However, hemangioma is characterized by the 'polka dot' sign representing the reinforced sclerotic trabeculae against the adjacent fatty marrow and vascular elements [9]. Paget's disease shows expansion, squaring, loss of the anterior vertebral body concavity, cortical thickening, and trabecular coarsening due to the excessive abnormal bone remodeling [10].

In conclusion, we present a case of BNCT presenting as an 'ivory vertebra' on radiography, and diffuse vertebral body involvement on both CT and MRI. This condition should now be included in the differential diagnosis of 'ivory vertebra'.

Acknowledgements The authors would like to acknowledge Dr. Roberto Tirabosco, Consultant Histopathologist, RNOH for his help with the manuscript.

Compliance with ethical standards

Conflict of interest The authors have no conflict of interest to declare.

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