CASE REPORT

Vertebral Bone Primary Angiosarcoma: A Case Report

Alfredo E Romero-Rojas, MD¹, Julio A Diaz-Perez, MD^{1,3}, Lina M Ariza-Serrano, MD³, Fabian E Neira-Escobar, MD²

¹National Institute of Cancer, ²Department of Radiology, National University of Colombia, Bogota, Colombia, and ³Moore's Cancer Center, University of California San Diego, La Jolla, California, USA

Introduction

Primary vascular tumors of the vertebral bones are extremely unusual¹, comprising <1% of all bone neoplasms^{1,2}. Their diagnosis is often challenging because of their clinical presentations vary from totally asymptomatic to extremely aggressive². Another problem associated with these tumors is the great confusion around their terminology; this problem has been resolved with the restriction of the term "hemangioendothelioma" to soft tissue lesions with a low degree of malignancy only^{1,3}. The term "epithelioid hemangioendothelioma", which was first introduced by Weiss and Enzinger to denote a group of soft tissue tumors, has been used many times since to incorrectly categorize bone tumors¹. The correct pathological diagnosis of these neoplasms is a priority because it is essential to the rapid selection of an effective management plan. We here describe a case of a primary vertebral angiosarcoma and present a concise review of the more relevant diagnostic aspects.

Case Report

A 69-year-old woman presented with a 3-year history of lower extremity paresthesiae at the T₁₀ level that limited her ability to walk. Previous cervical and lumbar MRI and dorso-lumbar radiographs had shown degenerative changes without any evidence of fractures or invasive lesions of the spinal cord. On physical examination, the patient had mild palpebral ptosis with facial symmetry. In addition, she had spasticity in both legs and decreased T₁₂ level sensation. No other clinical neurological manifestations were found. Vertebral MRI showed reduced height and low intensity on the T₁ sequence and mild hyperintensity of the T₁₁ vertebral body on the T₂ sequence. A short time inversion recovery contrasted sequence showed increasing signal intensity. The vertebral lesion involved adjacent soft tissues and dura mater, compress-

ing the spinal medulla to the right and posterior aspect of the spinal canal at the T₁₀ level and involving 80% of the canal at T₁₁ (Fig. 1).

A vertebral bone biopsy was performed. Histopathological examination showed a vascular neoplasm characterized by a proliferation of endothelial cells with atypical nuclei distributed in anastomosed intra-trabecular channels filled with abundant erythrocytes (Fig. 2). The endothelial cells were positive for Friend leukemia integration 1 transcription factor (FLI-1), cluster of differentiation (CD) 34 and CD31 and negative for CD10, osteopontin, epithelial membrane antigen, cytokeratin (CK) 7, CK20 and AE1/AE3 (Fig. 2). A primary angiosarcoma of the vertebral body was diagnosed.

The patient received palliative care and died 2 months later.

Discussion

Because bone vascular lesions can be reactive processes or benign or malignant (highly aggressive) neoplasms⁴, the terminology used to describe these tumors is variable. In 1943, Stout first described a case of vascular tumor of bone and denoted it as "hemangioendothelioma"¹. Later on, with discovery of the wide behavioral spectrum of those neoplasms, they have been classified as both benign and malignant tumors, the latter being variously called angiosarcomas, angioendotheliomas, hemangioendotheliomas, hemangioendothelial sarcomas, hemangiosarcomas or anaplastic vascular tumors. These capricious terminologies have been used because of the variability in their clinical and morphological features, which are a product of their complex and irregular architecture combined with a high degree of pleomorphism and cellular atypia^{3,4}. Some authors have suggested that the ones that behave innocuously are really bone hemangiomas (the most common bone tumor, with an autopsy prevalence around 10.7% in the

Address for correspondence Julio A. Diaz-Perez, MD, Moore's Cancer Center, University of California San Diego, 9500 Gilman Dr, La Jolla, California, USA 92037 Tel: 001-858-8223334; Fax: 001-858-405-8697; Email: jdiazperez@ucsd.edu or patologia@udes.edu.co

Disclosure: The authors state that they have no actual or potential conflicts of interest.

Received 26 December 2012; accepted 21 January 2013

Fig. 1 MR Images showing (a) T₁ low intensity signals in the vertebral bodies of T₈ and T₁₁ with (b, c) hyper-intensity in the T₂ and STIR sequences in sagittal views. (d) MRI showing multiple lesions between T₅ and L₁ in bone window. (e and f) These lesions were biopsied under CT guidance, posterior wall rupture with spinal canal compromise and rejection/compression of the medulla cord is visible.

general population, although many cases diagnosed as heman-
giomas are actually vascular malformations)^{2,3}.

Angiosarcomas are malignant vascular neoplasms with endothelial cell differentiation^{3,4} and atypical features such as polyhedral and fusiform cytology. On clinical evaluation, the most distinctive characteristic of bone angiosarcomas is the presence of multiple lesions in the same bone: solitary multifocal multicentric lesions (1/3 of all cases)⁵; such as were visible in our case (Fig. 1). Another typical feature is that the mass is highly destructive with poorly defined margins^{2,5,6}. Bone angiosarcomas occur more often in men than women, at an average age of 62 years (range, 26–83 years) and average 5 cms in diameter^{6,7}. Previous exposure of affected subjects to external factors such as radiation, thorotrast and polyvinyl chloride has been described⁸. No such exposure had occurred in our case.

Macroscopically, these neoplasms are soft and hemorrhagic. Histopathological examination shows neoplasms with highly complex architecture composed mainly of anastomosing vascular cords bounded by atypical endothelial cells and showing alveolar or pseudo-glandular formations. Desmoplastic reactions and papillary formations are also sometimes found in bone angiosarcomas. Furthermore, trabecular osteoid with osteoclastic activity typical of bone neoformation is frequently seen^{1,3,4}. The endothelial cells within angiosarcomas can resemble the malignant cells observed in epithelial neoplasms; for this reason, they are called “epithelioid”^{3,5}. Because of this last, epithelial neoplasm is a frequent differential diagnosis. Other entities that can be confused with bone angiosarcomas are fusiform sarcomas. These possibilities can be excluded by immunohistochemical staining: angiosarcomas are characteristically positive for CD34, CD31, FLI-1, factor

Fig. 2 Histopathological examination showed a malignant neoplastic lesion composed of a vast quantity of anastomosing vascular channels with atypical prominent endothelium that contained erythrocytes and invaded adjacent bone structures (HE, a: $\times 10$, b and c: $\times 40$). The neoplastic cells were positive for (d) CD34 ($\times 4$), (e) CD31 ($\times 40$) and (f) FLI-1 ($\times 40$), and negative for (g) AE1/AE3 ($\times 4$).

VIII, thrombomodulin and *Ulex europaeus* agglutinin 1 and have a high proliferation (Ki67) index.

In our case, because the disease presented at such an advanced stage, it was impossible to provide any curative therapeutic strategy. In these highly aggressive tumors, the initial treatment of choice is total en bloc resection. Because this approach minimizes intralesional invasion, it is also recommended for primary malignant spine tumors in instances where the expected surgical margin is definitive⁹. However, recurrence after resection is very common and the prognosis notoriously poor because metastases tend to develop early¹.

Metastases to the lungs and other parenchymal organs are found in about 66% of cases. Radiotherapy is effective for local control of the disease in some patients and in others offers dramatic palliative benefits. Angiosarcomas can disseminate to surrounding tissues or, frequently, to extra-osseous sites. Chemotherapy regimens offer variable results in such cases and the ideal combination remains undefined^{1,2,10}. Chemotherapy regimens, including cisplatin with doxorubicin, cisplatin plus paclitaxel and cisplatin plus doxorubicin and paclitaxel, have been studied¹⁰. However, prolonged responses are very uncommon and better therapies are urgently needed^{9,10}.

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