Hemangioendothelioma of bone

Anshul Agarwal, MD; Ashish Shah, MD; Chhaya Patel, MD; Asha Mary George, MD; Chandralekha S. Tampi MD

Department of Histopathology, Lilavati Hospital and Research Centre, Mumbai, India

Abstract

Introduction: 360 degree circumferential occurrence of spinal tumor is extremely rare.

Clinical material: A middle aged man presented to us with this tumor causing spinal cord compression, deformity and pain locally in the region of T12 and L1 from where the tumor was **arising.**

Past history: In 1996, he was operated for the same tumor elsewhere and stabilization was done. Details are not known.

Present history: Patient presented with above symptoms without bladder and bowel involvement, and weakness in right leg with some difficulty in walking.

On clinical examination plantars were flexors, jerks were elicited but not brisk and there was no wasting or sensory loss. Right side quadriceps was weak.

X-rays and MRI showed a multilobulated tumor arising mainly from T12 and partly L1 vertebrae with involvement of all the elements. It was moderately vascular on DSA and embolization was carried out.

The surgery: This was carried out in two stages. In the first stage through a transthoracic approach the anterior and anterolateral part of the tumor was excised and reconstruction was done by using implants. At a later stage, the posterior and posterolateral part of the tumor was excised and stabilization was carried out uneventfully.

Results: Patient has stood the surgery well. There were no complications. The histopathology of tumor suggests it to be hemangioendothelioma of the spine.

Key Words

- 360 degree circumferential tumor
- Spinal tumor
- Hemangioendothelioma

Introduction

The term hemangioendothelioma (HE) is used for a group of vascular tumors with intermediate malignancy, originating from the vascular endothelium and showing a histopathological appearance and a borderline aggressive biological behavior, i.e., between that of a hemangioma and angiosarcoma.[1] It is a rare well-differentiated endothelial tumor with variable presentation and behavior which may develop at different sites, such as in soft tissue, lung, pleura, spleen, heart, liver, as well as intracranial. Primary bone HE accounts for less than 1% of malignant bone tumors; cases occurring in the spine region are, especially rare.[2] Hemangioendothelioma primary in bone is an unusual neoplasm, it occurs in the second and third decades; and chiefly affects lower limbs, but it may involve any bone.[3–5]

Here, we report a unique case of spinal HE and present surgical treatments, pathologic findings, and imaging characteristics in a comprehensive review of the literature.

Case report

A 53-year-old male was admitted for back pain, since 3 years. He was operated 15 years ago for giant cell tumor spine. A high resolution CT & MRI revealed a large expansile lytic lesion involving D10-L1 vertebrae (size 9x9x8 cms) (Figure 1 A,B&C). The surgery was performed in two stages – At first debulking was done posteriorly and then a month later from the anterior aspect with stabilization.

On histological examination, a spindle cell tumor with destructed bony trabeculae, pieces of cartilage and fibrous

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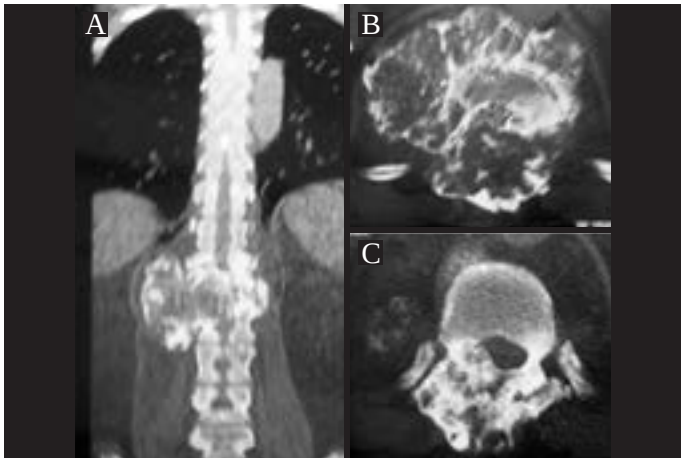

Figure 1 A,B&C
CT scan showing large expansile lytic lesion involving D10-L1 vertebrae

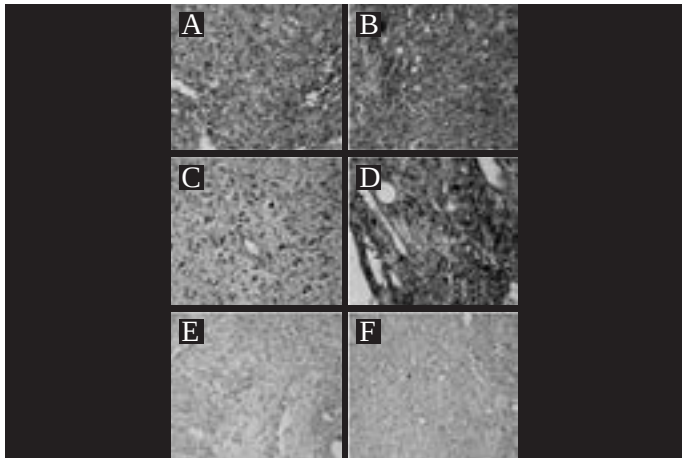

Figure 2 A,B,C,D,E&F
A,B – H & E section (x400)
C – Tumor cells immunoreactive for CD31
D – Tumor cells immunoreactive for vimentin
E – Tumor cells negative for CK. (F) MIB-1 index<1

tissue were seen. The tumor cells were haphazardly arranged in a myxohyaline background. Few areas of calcification and hemorrhage were also present. The individual tumor cells showed mild nuclear pleomorphism with intracytoplasmic vacuoles. Areas showing vascular lumina formation were noted. Focal collections of osteoclastic giant cells were seen. Features of a giant cell tumor were not seen.

Immunohistochemical profile confirmed the endothelial and vascular nature of the lesion with both the intervening as well as vascular space-lining cells showing positivity for the endothelial cell markers CD31 and vimentin. The tumor was immune-negative for cytokeratin (CK). The MIB-1 Index was less than 1%. The morphology and IHC confirmed hemangioendothelioma (intermediate grade Sarcoma) (Figure 2 AB,C,D,E&F).

The post-operative period was uneventful.

Discussion

Hemangioendothelioma is a rare tumor of vascular origin with an uncertain malignant potential. Hemangioendothelioma of the soft tissue was described in 1982 by Weiss and Enzinger6, and it has been described with lesser frequency in visceral organs and other sites.[7,8] Hemangioendothelioma of bone is rare.[2–5,9–11] The clinical behavior is variable; most are cured by excision, but upto 40% recur, 20–30% eventually metastatize, and perhaps 15% of patients die of tumor.

HE, typically, presents with non-specific signs and symptoms, most commonly painful lesions; which may be associated with a mass, thereby resulting in a loss of function. Vascular or neurological symptoms appear depending on the location. The typical radiographic and CT appearance of HE arising in bone is a lytic lesion without matrix mineralization; osseous expansile remodeling may be seen. Joint invasion is a common feature and homogeneous enhancement is present on contrast material-enhanced CT or MRI. The signal intensity characteristics on MR images are non-specific. There is low-to-intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images. The aggressive radiographic appearance of HE raises the suspicion of malignancy. Differential considerations for this radiographic presentation include Langerhans' cell histiocytosis, giant cell tumors, fibrous dysplasia, lymphoma, and metastatic diseases.[12]

Histopathological evaluation is central to the diagnosis of this tumor. At pathological analysis, the tumor may be well circumscribed or have indistinct borders. The surface is macroscopically light red or purple, with a soft consistency associated with bone fragments. The distinct well-lined anastomosing vascular channels often seen in hemangiopericytoma are absent in this tumor. Cells may be round or fusiform with a central nucleus and prominent intracytoplasmic vacuolation. The positivity of endothelial cell markers CD31 and factor VIII-related antigen is essential in the diagnosis, as in the specimen in our case. Classification of malignant potential relies on the degree of vasoformative activity, atypia of the endothelial cells, and the frequency of mitotic activity as predictive signs. Unlike other aggressive vascular tumors, such as hemangioendothelial sarcoma or angiosarcoma, the histological grading system is not useful for predicting prognosis.[12–14]

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Address for correspondence
Dr. Anshul Agrawal: Email id: dranshulagarwal@gmail.com