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Case Report

Lumbar Vertebra Epithelioid Hemangioendothelioma: A Case Report

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Abstract

Epithelioid hemangioendothelioma is a rare vascular tumor originating from the vascular endothelium. This tumor can manifest asymptomatically and be incidentally discovered. However, the most frequent presentation in the context of spinal involvement is localized pain. Primary spinal epithelioid hemangioendothelioma is an exceptionally infrequent occurrence, with only a limited number of cases documented in medical literature. In our article, we present a case involving a 48-year-old male diagnosed with epithelioid hemangioendothelioma located in the L2 vertebral body within the lumbar spine. The condition was first identified due to the emergence of right lower back pain. To alleviate pain and reduce the risk of potential pathological fractures, a posterior fixation procedure involving L1 and L4 was carried out. The patient's condition improved significantly following radiotherapy, resulting in a positive outcome.

Keywords: *Epithelioid Hemangioendothelioma; Imaging; Histology; Vascular Tumor; Spine*

Introduction

Epithelioid hemangioendothelioma is an angiocentric vascular tumor with metastatic potential [1]; it was first described in 1982 by Weiss., *et al*. as a vascular tumor of bone and soft tissue [2]. Primary epithelioid hemangioendothelioma of bone accounts for only 1% of all malignant tumors of bone [3]. The behavior of epithelioid hemangioendothelioma is intermediate between hemangiomas and conventional angiosarcomas [4]. Spinal epithelioid hemangioendothelioma is a rather rare disease with few reported cases and series reported in the literature [3]. Clinically, it usually manifests itself by local pain [5]; imaging data are non-specific [6], the diagnosis of certainty of epithelioid hemangioendothelioma remains essentially histological [6]. We present a case of primary epithelioid hemangioendothelioma of the lumbar spine.

Case Report

Information patient

We present a case involving a 48-year-old male patient who was admitted to the emergency department due to an acute exacerbation of lower back pain. The patient had a four-year history of right-sided low back pain. His past medical history, medication usage, and family medical history did not reveal any significant findings.

Clinical findings

Upon admission, the patient was alert, conscious, and displayed regular vital signs. He reported experiencing low back pain, and the physical neurological examination did not uncover any abnormalities.

Timeline

A CT scan of the lumbar spine was performed showing osteolytic lesions with peripheral bone sclerosis involving the body of L3, blowing the bone cortex which is thinned and broken in places (Figure 1).

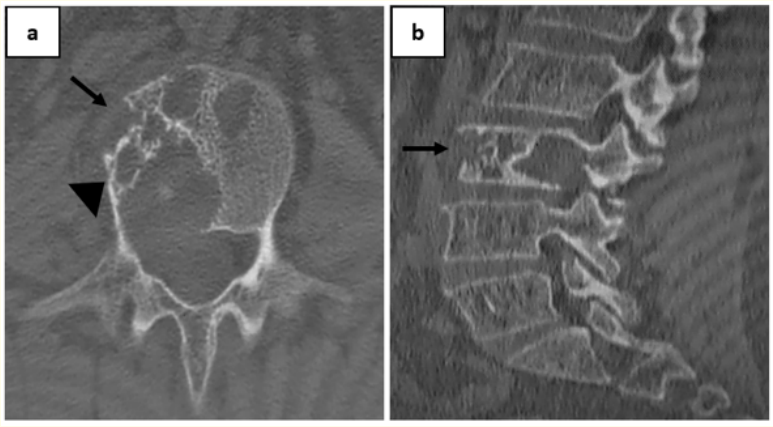

Figure 1: *CT scan of the lumbar spine in axial (a) and sagittal (b) sections: osteolytic lesions with peripheral bone sclerosis (arrowhead) involving the body of L3, blowing the bone cortex which is thinned and broken in places (arrow).*

To reduce pain and prevent possible pathological fracture a posterior fixation of L1 and L4, was performed.

An MRI of the dorsolumbar spine was performed, showing a lesional process of osteolytic tissue signal in L3 without structural abnormality of the vertebra, blowing out the supporting bone and breaking in places the anterior and posterior cortices with signs of endosteal resorption. This process presents an anterolateral right epidural extension with the envelopment of the L4 root and presents in T1 hyposignal, frank T2 hyper signal, STIR, intensely enhancing after injection of Gadolinium, presenting a flaky aspect in STIR and includes some vermicular structures in T1 and T2 hyposignal (Figure 2).

Anatomopathological study of bone fragments from a scanno-guided biopsy of the lytic lesions revealed a vascular tumor proliferation organized in patches without a proper capsule with an anastomotic network of fine congestive capillaries dissociated by larger vessels. The endothelium is turgid on the surface without cytonuclear atypia or mitoses. Some vessels are quite large with a slightly thickened wall. The interstitial tissue is dissociated by hemorrhagic foci and scattered with mono- and polynuclear inflammatory elements without calcifications (Figure 3).

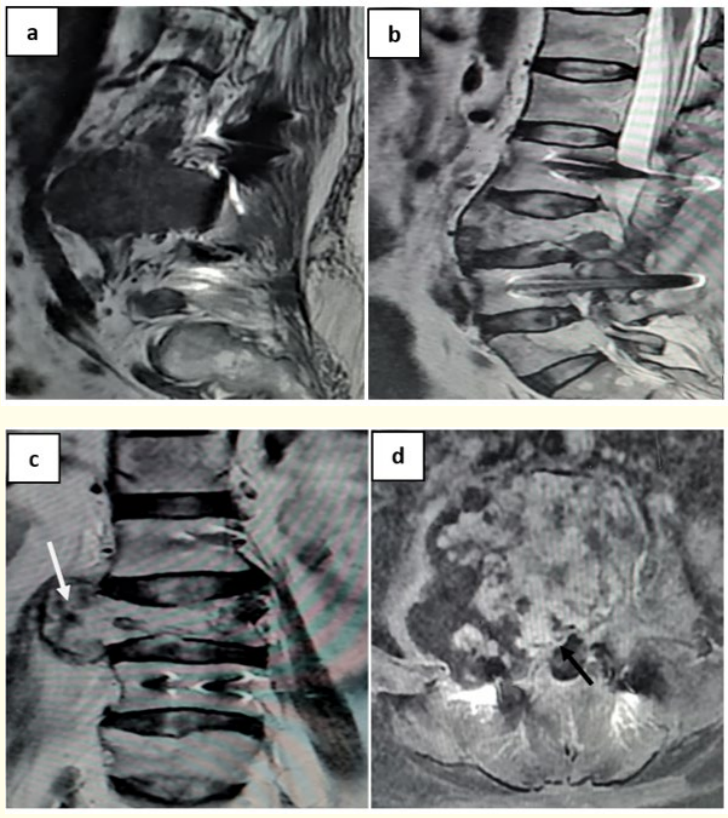

Figure 2: *MRI of the dorsolumbar spine: Osteolytic tissue lesion process of the body of L3, blowing and breaking in places the anterior and posterior cortices with evidence of endosteal resorption. This process presents a right anterolateral epidural extension (black arrow) with the envelopment of the L4 root (white arrow) presenting in T1 hyposignal (a), frank T2 hyper signal (b, c), intensely enhancing after injection of Gadolinium, presenting a flocculent aspect in STIR (d).*

Immunohistochemically, the epithelioid endothelial cells were positive for the endothelial markers CD31 (Figure 3).

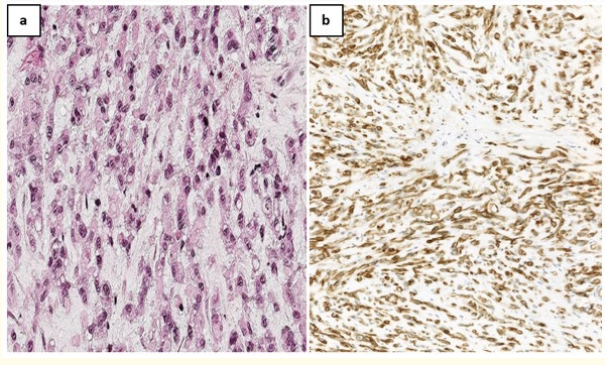

Figure 3: *(a) Anatomopathological study of bone fragments from a scanno-guided biopsy of the lesion: vascular tumor with cords and groups of epithelioid cells. (b) Immunohistochemically, the epithelioid endothelial cells were positive for the endothelial markers CD31.*

Diagnostic assessment: The CT and magnetic resonance imaging (MRI) results did not provide specific findings. Nevertheless, the morphological features and immunohistochemical profile were indicative of an epithelioid hemangioendothelioma diagnosis.

Therapeutic intervention and following up

The choice was made to solely administer radiotherapy for tumor treatment. The patient underwent a total of 30 fractions of local radiation therapy (RT). During the most recent follow-up, which occurred approximately 3 months after completing the radiotherapy, there was a significant improvement in spinal pain.

Informed consent: Written informed consent was obtained from the patient for participation in our study.

Discussion

Epithelioid hemangioendothelioma is an extremely rare malignant vascular tumor that exhibits an intermediate behavior, falling between locally aggressive hemangiomas and angiosarcomas [3,4]. It accounts for just 1% of primary bone tumors [2,3], with spinal involvement being particularly uncommon [4]. Its first documented description dates back to 1982 by Weis., *et al*. [2]. According to the WHO 2020 classification, bone epithelioid hemangioendothelioma is classified as a low to intermediate-grade malignancy [7]. This condition can develop at any age but tends to have a preference for individuals aged 20 to 30 years [7]. It occurs more frequently in men, with a male-to-female ratio of 2:1 [7]. In over 50% of cases, it presents as multifocal lesions [4]. Primary spinal epithelioid hemangioendothelioma possesses an inherent potential for metastasis [2], with estimated rates of distant metastasis ranging between 20% and 30%, along with local recurrence rates of up to 13% [4].

Epithelioid hemangioendothelioma originates from vascular endothelial or pre endothelial cells [5,7]. Pathologically, it is manifested by epithelioid endothelial cells arranged in nests or cords [5], it does not show well-formed vascular structures such as those seen in hemangioma nor cytological atypia that is present in angiosarcoma [6]. Immunohistologically, epithelioid hemangioendothelioma cells usually express endothelial markers, such as RME, CD31, CD34, and FVIII-Rag [5].

In our case, the epithelioid endothelial cells were positive for the endothelial markers CD31.

Epithelioid hemangioendothelioma has no specific clinical signs [5], it most commonly manifests as local pain or neurological symptoms resulting from the spinal cord or nerve root compression [5,7]. Our patient presented with right low back pain.

Imaging is necessary to obtain a better morphological and topographical characterization of epithelioid hemangioendothelioma, it also allows the detection of complications including fracture or spinal cord compression which are frequently reported [8].

The radiological characteristics of epithelioid hemangioendothelioma are not specific [5,6]. On standard radiography and CT scan, it usually appears as osteolytic lesions surrounded by sclerotic margins [3]. Periosteal reaction and calcifications are rare [3], and there may be focal areas of calcifications described as “ice floes” sign [2]. A typical epithelioid hemangioendothelioma is characterized by low to intermediate intensity on T1-weighted images and high intensity on T2-weighted images, restricted diffusion, with clear homogeneous or peripheral enhancement [3,7], destruction of the bone cortex and soft tissue invasion may occur in some cases [3].

The differential diagnosis of primary spinal epithelioid hemangioendothelioma arises primarily as a solitary tumor with the following diagnoses:

- Giant cell tumors: Which most often manifests as expansive osteolytic “soap bubble” lesions with T2 hyposignal whereas the typical form of epithelioid hemangioendothelioma usually shows T2 hypersignal [9].

- Epithelioid hemangioma: It occurs mainly in the dorsal spine, preferentially in the transverse process, and appears as multifocal expansive lytic lesions with T1 isosignal and T2 hypersignal [9].
- Chondrosarcoma: Usually located in the dorsal spine, it appears as expansive lytic lesions with diffuse ring or arc calcifications, in T1 hyposignal and T2 hypersignal [9].
- Hemangioma: Is a most common vascular tumor of the spine, it is characterized by a gridded appearance on CT and a T1 and T2 hyperintense signal on MRI [5,9].
- Skeletal aneurysmal bone cyst: Rare, it is mostly found in young subjects (< 30 years), it appears as an eggshell osteolytic lesion, heterogeneous, in hyposignal T1, hypersignal T2 with presence of a liquid-liquid level [2,9].

Moreover, in elderly patients with multifocal involvement, the primary differential diagnoses include metastases, lymphoma and myeloma [6]. For young patients with multifocal involvement, the differential diagnosis encompasses brown tumors, Langerhans cell histiocytosis, and fibrous dysplasia [6].

Due to its rarity and unpredictable course, there are no established treatment guidelines for epithelioid hemangioendothelioma [7,10]. Treatment options for primary spinal epithelioid hemangioendothelioma encompass surgical intervention, radiation therapy, and chemotherapy [5,7]. The overall survival rate for patients with unifocal epithelioid hemangioendothelioma was 89%, whereas it dropped to 50% for patients with the multifocal form [7].

Conclusion

Primary spinal epithelioid hemangioendothelioma is an exceptionally rare form of low- to intermediate-grade malignant vascular tumor, typically manifesting as localized pain. Imaging findings are non-specific. Typically, it appears as an expansive osteolytic lesion with sclerotic borders and a matrix resembling soap bubbles. However, a definitive diagnosis relies primarily on morphological and immunohistochemical histological data.

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