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Intractable low-back pain in a patient with history of multiple neoplasms

Christelle Haikal¹ · Nathan Beucler² · Stéphane Fuentes² · Corinne Bouvier³ · Thomas Le Corroller^{1,4}

Question

A 45-year-old female patient, with a history of surgically treated tonsil squamous cell carcinoma, thyroid cancer treated by thyroidectomy and radioactive iodine therapy, and high-grade cervical intraepithelial neoplasia, presented with intractable low-back pain (Figs. 1, 2, and 3).

Answer

Epithelioid hemangioendothelioma of the third lumbar vertebra.

Discussion

MRI (Fig. 1) demonstrated a single, not sharply demarcated lesion of the third lumbar vertebra presenting with intermediate T1W signal intensity (SI), high T2W SI, and inhomogeneous enhancement. The lesion appeared confined to the vertebral body, causing a discrete focal superior endplate impression, and did not show a mass effect or

spinal canal infiltration. CT (Fig. 2) disclosed a lytic and well-circumscribed lesion involving the left aspect of the vertebral body, with focal cortical destruction. On PET-CT, the tumor showed a maximum standardized uptake value of 7 mg/mL, with no other lesion. At this point, a diagnosis of bone metastasis was suggested, and the patient underwent a CT-guided bone biopsy. Histological examination (Fig. 3) revealed cords of epithelioid cells (arrows) and vascular structures (arrowheads) embedded within spongy tissue. Immunohistochemistry demonstrated diffuse cytoplasmic expression of CD31 and CD34 endothelial markers and nuclear expression of CAMTA1 (Fig. 3), confirming a diagnosis of epithelioid hemangioendothelioma (EHE). The patient underwent en bloc resection of L3 vertebral body and spine reconstruction using an expandable cage and posterior osteosynthesis.

With the advent of diagnostic molecular pathology, the classification of vascular bone tumors (VBT) has been clarified into three different tumor types: hemangioma, hemangioendothelioma and angiosarcoma [1–4]. Except for its quite uncommon epithelioid variant, hemangioma of bone is constantly benign even in its radiologically “aggressive” expansile presentation [5]. Malignant VBT include EHE and angiosarcoma that are considered respectively low- to intermediate-grade and high-grade malignant neoplasms [5].

EHE of bone is rare, with most patients diagnosed during the second to third decades of life. The tumor can affect long tubular bones, especially in the lower extremities, followed by pelvis, ribs and spine. EHE is multifocal in more than half of cases, either within a single bone or involving separate bones, and tends to cluster in an anatomical region [3–5]. It manifests with localized pain and swelling. On imaging, EHE of bone appears as a lytic lesion that originates in the medullary cavity and grows in an infiltrative and destructive fashion. The tumor may be expansile and erode the cortex, even if soft tissue extension remains uncommon [3–5].

Histologically, EHE is a solid mass composed of cords and nests of large epithelioid endothelial cells in a

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Fig. 1 Magnetic resonance imaging of the lumbar spine demonstrating a single, not sharply demarcated lesion of the third lumbar vertebra presenting with intermediate T1W signal intensity (a), high T2W signal intensity (b), and inhomogeneous enhancement (c)

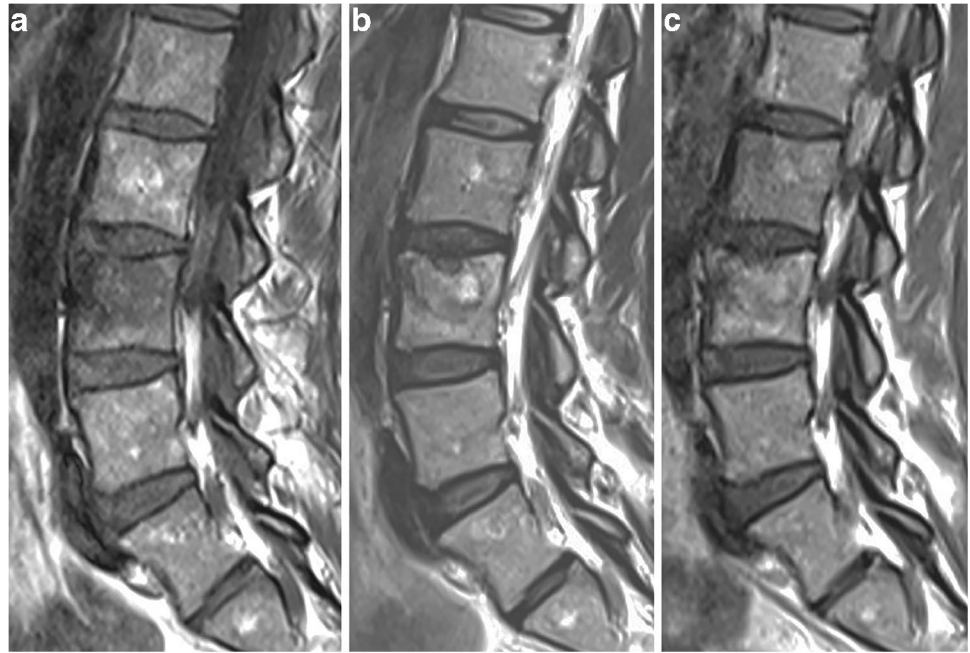
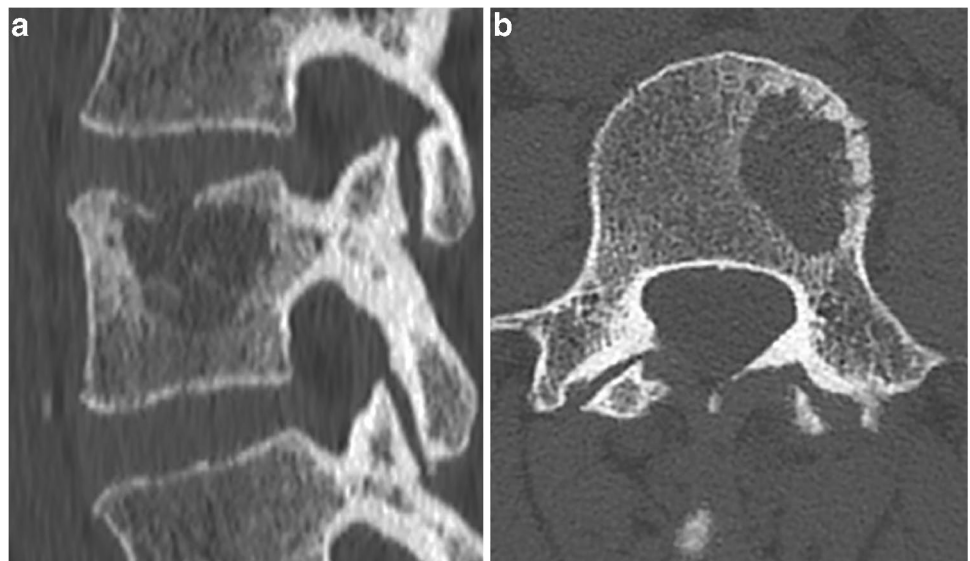


Fig. 2 Sagittal (a) and axial (b) computed tomography images showing a lytic and well-circumscribed lesion involving the left aspect of the vertebral body, with focal cortical destruction



myxohyaline stroma [5]. Owing to its epithelioid morphological features, EHE can easily be confused with metastatic carcinoma, melanoma, mesothelioma, or epithelioid angiosarcoma, especially when multifocal [3, 5]. Evidence of CAMTA1 expression using immunohistochemistry or identification of a WWTR1-CAMTA1 fusion transcript confirms the diagnosis of EHE [6–8]. Rarely, YAP1-TFE3 fusion can be found in a small subset of cases that exhibit a distinct morphology with solid growth or well-formed vascular channels lined by epithelioid cells with nuclear atypia and abundant pale cytoplasm [5].

Treatment of EHE of bone involves wide surgical resection. The overall survival rate is 92% at 10 years [9]. Involvement of two or more bones is associated with a worse prognosis [5].

Because there are no specific CT or MRI features in EHE of bone, this tumor can easily be misdiagnosed as metastatic carcinoma, especially in its multifocal presentation or when there is past history of primary tumor as in our patient. As with other malignant VBT, the only diagnostic clue is the possible multifocal clustering in an anatomical region [3–5]. Histological sampling is crucial. Wide resection is the treatment of choice.

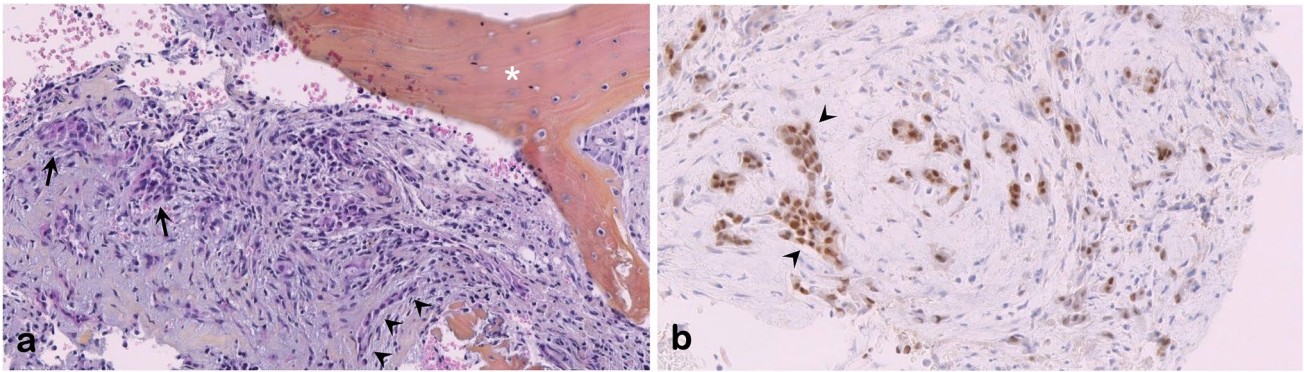


Fig. 3 Microscopic features. **a** Cords (black arrows) and vascular structures (black arrowheads) embedded within loose tissue and surrounded by bone (white star); hematoxylin–eosin staining. **b** Diffuse nuclear expression of CAMTA1 by immunohistochemistry (black arrowheads)

Declarations

Conflict of interest The authors declare no competing interests.

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