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Epithelioid Hemangioendothelioma of the Proximal Portion of the Left Tibia about a Case

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Abstract Case Report

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor, of intermediate malignancy, its evolution is unpredictable and its treatment is essentially surgical. We report a case of an EHE involving the proximal portion of the left tibia in a 32-year-old woman, which caused a diagnostic issue. And in whom, the first step of the surgical treatment has been established up to now.

Keywords: Epithelioid hemangioendothelioma, vascular tumour, osseous.

INTRODUCTION

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumour considered to be of intermediate malignancy, that can affect bones, soft tissue, lungs and the liver. We report in the following, a clinical case of an EHE involving the proximal portion of the left tibia, with a review of the literature.

OBSERVATION

This is Mrs L.E, 32 years old, with a history of treated rheumatic fever, and who has been reporting for a year inflammatory pains of the left knee, gradually increasing in intensity and resisting medical treatment. The orthopedic examination finds pain caused by the pressure of the proximal portion of the left leg, normal walking without lameness or axial deformation, with retained articular amplitudes of the left knee (150 degrees of flexion and 180 degrees of extension). An X-ray of both knees showed a metaphysial and diaphysial lytic image with blurred and polycyclic contours blowing the posterior cortical of the upper end of the left tibia without rupturing it, and blurred opaque images of the distal ends of the two femurs (figure 1 left). At that time, several diagnoses were mentioned, including a benign or malignant tumor origin, an infectious, hormonal, rheumatological or hematological origin, etc.

To better study these images, a Computed Tomography (CT) of the two knees was carried out initially, and confirmed the presence of these images of epiphysial, metaphysial and diaphysial osteolysis with cortical rupture (Figure-1 right). In a second step, a magnetic resonance imaging (MRI) of the left knee was performed, showed the presence of a 12-cm tumoral process involving the proximal end of the left tibia with the soft tissue broken in (Figure-2).

A surgical biopsy was performed in the operating room, then an anatomopathological study, who favoured an Epithelioid hemangioendothelioma (Grade II (intermediate) according to the CRLCC, the immunohistochemical study was positive for CD31 expression.

A spreading assessment was carried out, it included:

- A thoraco-abdominopelvic CT scan, which came back normal.
- A skeletal scintigraphy: showing an heterogeneous hyper-fixation at the proximal third of the left tibia and the external side of the distal part of the left femur (Figure-3 left).
- A positron emission tomography (Pet scan): showing a hyper-metabolism of the proximal part of the left tibia and the distal end of the left femur (Figure-3 right).

In front of this chart, a decision was made to perform a left knee resection-arthrectomy (Figures 4-6), with the initial installation of a spacer (made from a nail of femur+antibiotic cement) (Figure 6 & 7), a massive prosthesis will be installed later in a second operating time.

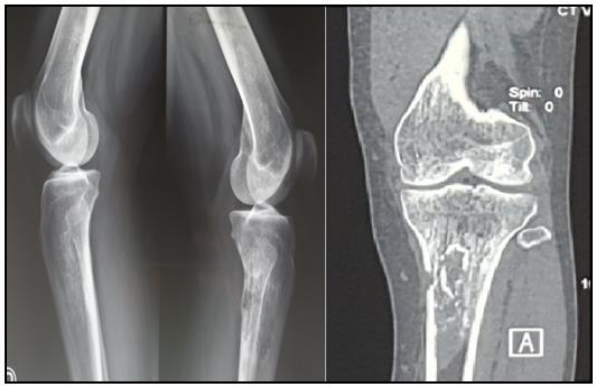

Fig-1: X-ray and Computed Tomography

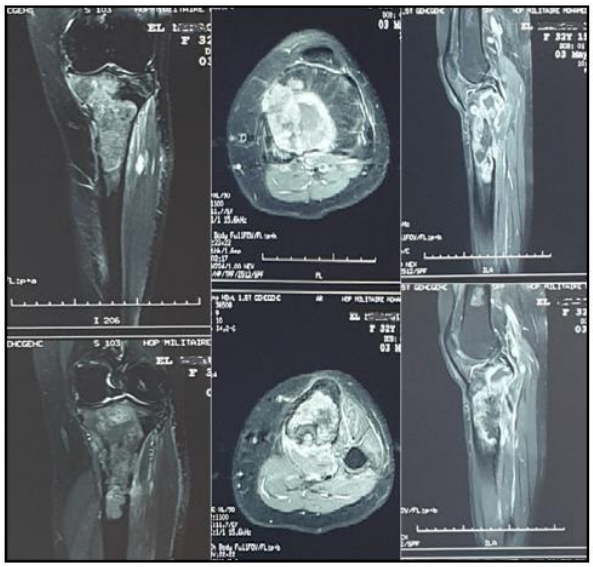

Fig-2: MRI

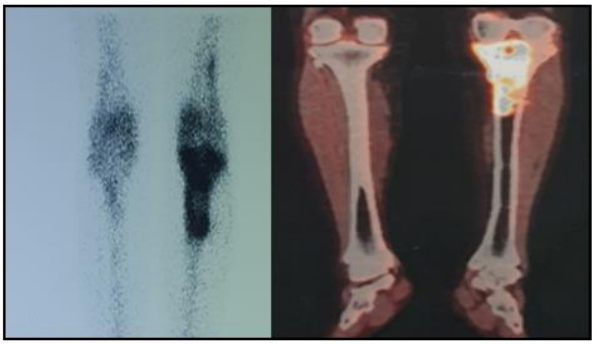

Fig-3: Skeletal scintigraphy and Pet scan

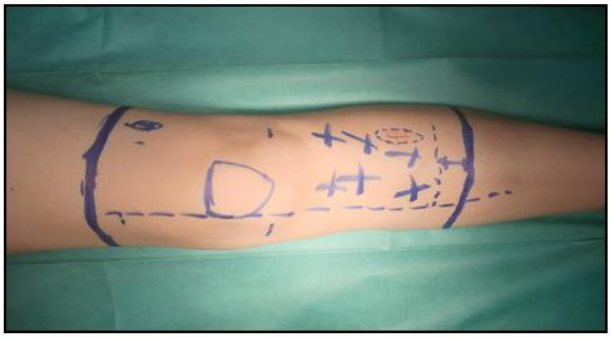

Fig-4: Anterior Approach

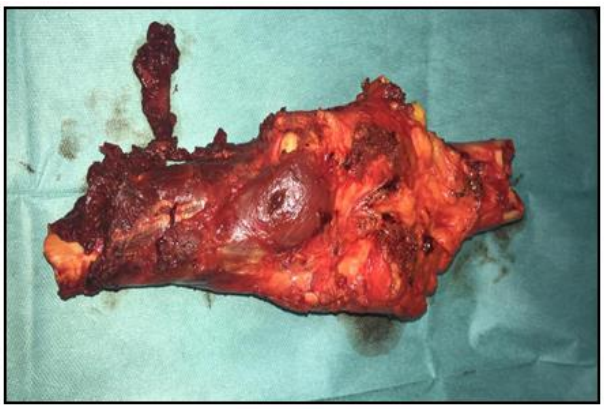

Fig-5: Resection piece

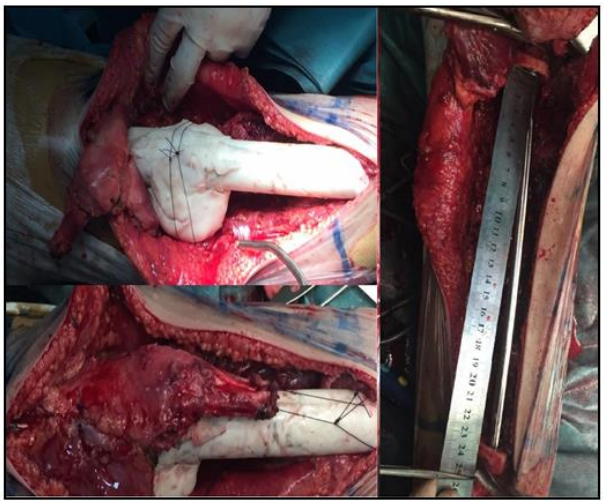

Fig-6: Resection-arthrectomy (25cm) and installation of a spacer

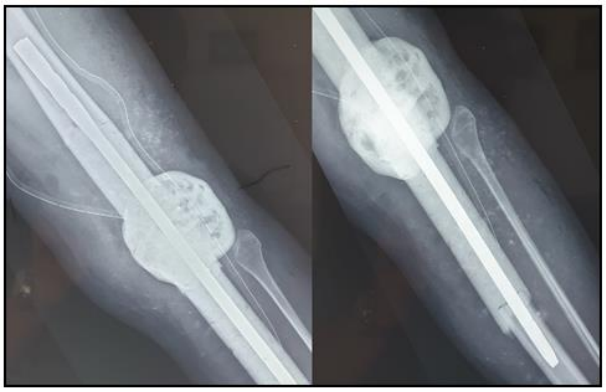

Fig-7: Post-operative radiography

DISCUSSION

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor, of recent description, its initial diagnostic criteria were established in the 1970s–1980s. It is a ubiquitous tumour that can develop in bones, soft parts or viscera (liver and lung) [3, 4, 5]. This term «Epithelioid hemangioendothelioma» was first introduced by Weiss and Enzinger in 1982 [1], to describe a bone or soft parts tumor, of intermediate malignancy "Border line" [2, 3, 4], that may simulate benign 'hemangioma' tumors or show signs of malignancy such as 'angiosarcomas' [6]. In the 1970s, Corrin *et al.*, demonstrated that these tumours were derived from endothelial progenitors [7]. They can be diagnosed at any age, but the average age of diagnosis is usually 20 to 30. There are as many cases among men as there are among women.

In the current classification of the World Health Organization (WHO), EHEs are classified as locally aggressive tumors with metastatic potential [9]. They can be bone-isolated or integrated into a multifocal shape. Most bone EHEs mainly affect long limb bones (vertebrae in less than 10% of cases), either by single or multiple damage to a single piece of bone, or by affecting several bone segments in an anatomical region, or in a diffuse way over the entire skeleton.

EHE is characterized by clinical latency, with a very slow evolution. The main symptoms are pain, swelling and compressions, especially neurological ones in case of vertebral location. These lesions rarely cause changes in the colour of the skin opposite said lesion. Vascular symptomatology such as oedema and thrombophlebitis may be associated [8]. Lymphadenopathy can be found in the drainage territories of the tumor site.

Standard radiography and computed tomography (CT) are not specific, showing osteolytic lesions with clear or multilobed contours, sometimes with cortical rupture, without periosteal reactions. Calcifications and pathological fractures are possible. MRI with gadolinium injection shows heterogeneous contrast of lesions; and allows a better study of the tumor's anatomical relationships and the search for a possible extension in the soft parts.

On the anatomical-pathological level:

- Macroscopy: Tumour mass developed at the expense of the vessel wall, with vessel obliteration and centrifugal extension in surrounding tissue starting from the vessel [6, 9].
- Microscopy: a) No aspect of mature vascular differentiation but limited phenotype, intracytoplasmic vacuoles containing erythrocytes [6, 9]. b) Epithelioid tumour cells arranged in chains or strings within a myxo-hyalin stroma. Most EHEs have a nuclear monomorphic aspect with low-grade aspects.
- Immunohistochemistry: Approximately 20% of EHEs express vascular markers (ERG, CD31), but CD34 expression is inconsistent [10]. 30% of EHEs express epithelial markers such as cytokeratin 7 or 8, cytokeratin 18, EMA [11]. 90% of EHEs express CAMTA1 [12, 13].

As for the extension test, one can recommend a Computed tomography of the thorax, abdomen and pelvis (CT-TAP) with intravenous injection of iodine contrast product. The place of the bone scan is not clearly established. This whole assessment is recommended before discussing any surgical options. The role of positron emission tomography is not clearly established because the fixation is inconsistent and the intensity is variable.

In case of an isolated lesion, accessible to a surgical act with a curative aim, surgery can be suggested. In case of inoperable or multifocal injury, a careful monitoring may be proposed as a first step. The surgical procedure consists of a wide resection or arthrectomy resection with joint reconstruction. Sarcomas were observed in irradiated territory after treatment of 8% of the bone EHE. Radiation therapy is therefore reserved for non-operable localised tumours [14]. The goal of systemic treatment is palliative [17], to slow the progression of the disease and maintain quality of life. There is no standard systemic treatment. Several chemotherapy protocols have been evaluated for this indication, even for hepatic or pulmonary EHEs, but none has showed superiority so far, with partial results.

EHEs are considered intermediate malignancy tumours in the WHO 2013 classification [9]. Their evolution is not predictable, between "benign" and "malignant" behaviour. The tumour may be spontaneously stable. One case of spontaneous regression has been documented [16].

After removing an isolated tumour, the risk of metastasis is increased if its size exceeds 3 cm and at least 3 mitoses per 50 fields. Disease-free survival is 100% for tumours without these criteria and 59 % for tumours with one of these two criteria [15]. Some multifocal forms remain stable for years or even decades.

CONCLUSION

Bone EHE is a rare tumour of vascular origin with intermediate malignancy, and of unpredictable evolution. Its clinical presentation is mild, and his histological diagnosis is difficult. Immunohistochemical positivity is found especially for CD31. Its treatment is surgical, and should be as conservative as possible. International multicentre studies are essential to get to

know this exceptional disease because clinical trials are still too rare.

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