

## A rare case of a very aggressive angiosarcome of the tibia: A case report and review of the literature

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### Abstract

**Introduction and importance:** Angiosarcoma is a malignant vascular tumour of bone composed of endothelial cells. It accounts for 1% of all malignant bone tumours. The interest of this observation is to show the particularly aggressive and devastating nature of this type of tumour, which remains very rare and whose evolution is totally unpredictable.

**Case presentation:** This was an angiosarcoma of the tibia in a 29-year-old patient with no particular pathological history, which according to the patient had appeared 3 months previously. A radiological work-up confirmed the malignant nature of the lesion, hence the need for a biopsy, which confirmed the diagnosis of angiosarcoma.

**Clinical discussion:** Angiosarcoma is defined by its high histological degree of anaplasia, its radiological aggressiveness and its rapid clinical course with a very high metastatic potential. It is a rare tumour that occurs mainly in adults aged between 20 and 60, with a predominance of males. CT and MRI are used to define the limits of the tumour, biopsy confirms the diagnosis, and treatment is surgical.

**Conclusion:** Angiosarcoma is an angioendothelioma with a high grade of malignancy, the course of which is usually lethal whatever the treatment used.

**Keywords:** Bone; Tumour; Angiosarcoma; Metastasis; Surgery

### 1. Introduction

Angiosarcoma is the accepted term for high-grade malignant vascular tumours. These tumours are rare, accounting for only 1% of malignant bone tumours. The majority of these tumours are primary. However, there are also so-called secondary forms [1, 2] (the disease occurs at any age, with a peak in frequency at 60 and 80 and a median age of 51 [3]). There is a clear male predominance, and the disease is most often found in the long bones and short bones of the extremities [4]. It is unique in 2/3 of cases [5]. The data in the literature are poor, and there are no consensus criteria for differentiating angiosarcoma from other vascular tumours, in particular HEE [7]. Angiosarcoma is most often manifested by a chronic painful mass [8].

Microscopically, bone angiosarcomas give rise to heterogeneous lesions, ranging from a well-differentiated tumour with vascular formation to a solid tumour that may mimic a metastatic carcinoma [9]. Genetically, a recent study showed a t(1;14)(p21;q24) translocation in a bone angiosarcoma [10]. The radiological presentations of bone angiosarcomas are aspecific, with both CT and standard radiography showing an invasive, multi-centric appearance, while MRI is useful

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for showing the extension of neighbouring structures. Scintigraphy is still useful for differentiating bone angiosarcomas from other vascular processes in the bone [11].

The prognosis remains guarded, with a high risk of metastasis [12]. Surgical treatment with monobloc excision remains the gold standard in single forms [13].



**Figure 1** X-ray of the left leg (AP and lateral) showing the malignant appearance of the lesion



**Figure 2** X-ray of the left leg after one month = spectacular evolution of the tumour



**Figure 3** Clinical aspect after mid-thigh amputation left leg

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## 2. Clinical observation

The authors report the case of a 29-year-old patient who had been complaining of intermittent pain in his left leg for 3 months, with no downstream vasculo-nervous disorders. A radiological work-up showed (figure 1) a diaphyseal-metaphyseal-epiphyseal osteolytic lesion of the left leg with cortical blowing without opacification of the soft tissues. We then requested a CT scan, which unfortunately could not be carried out due to lack of funds. Given the malignant appearance of the lesion, a biopsy was performed, confirming that it was a grade III angiosarcoma of the tibia. After consultation with the oncologists, the patient underwent radiotherapy and chemotherapy 1. A follow-up radiograph taken 1 month later showed (figure 2) a spectacular worsening of the lesion, with extensive lysis over almost the entire diaphyseal-metaphyseal-epiphyseal bone surface and virtual disappearance of the cortical bone. A mid-thigh amputation was performed (figure 3). The extension work-up, consisting of a chest X-ray, thoraco-abdominal CT scan and scintigraphy, revealed a pulmonary lesion that was probably metastatic. A new chemotherapy protocol was initiated. After 1 month of treatment, the patient died of pulmonary complications.

I declare that this work has been reported in accordance with SCARE criteria [6]. (The patient is not informed by this work because of his death, but the family's agreement has been obtained)

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## 3. Discussion

Bone vascular tumours are a heterogeneous group of tumours with a spectrum ranging from benign to malignant. Angiosarcoma is the accepted term for high-grade malignant vascular tumours [7].

These tumours are rare, accounting for less than 1% of malignant bone tumours. The majority of these tumours are primary. However, secondary forms have been described as developing after radiotherapy [1, 2]. Or after bone infarction [14], or Paget's disease [15], or overload disease [16] or contact with orthopaedic implants and metallic materials [17, 18]. These tumours can occur at any age, peaking at 60 and 80, with a clear male predominance, and are most often found in the long bones [3, 4]. Bone angiosarcomas are clinically manifested by chronic pain followed by the appearance of a rapidly progressive mass at the time of diagnosis.

### 3.1. Imaging

- Radiographs usually show a purely lytic image, sometimes heterogeneous with a mixture of lysis and sclerosis [11].
- On CT scans, bone angiosarcoma has a multicentric and invasive appearance, as on standard X-rays [11].
- On MRI, bone angiosarcoma is hyposignal in T1 with a signal of variable intensity, and hypersignal in T2 [11].
- CT and MRI can be used to determine the limits of the tumour and whether it has invaded neighbouring soft tissue. A CT scan of the lung and a bone scan must be included in the extension assessment [11].

### 3.2. Microscopy and genetics

Microscopically, angiosarcoma is a malignant vascular tumour of bone composed of endothelial cells with a high histological degree of anaplasia [9].

Genetically, recent studies have described a translocation t(1;14)(p21;q24) in bone angiosarcomas [10].

### 3.3. Surgery

Surgical treatment must be discussed collegially at a multidisciplinary consultation meeting, and multifocal involvement must be sought before any therapeutic decision is taken. (The gold standard is a monobloc excision that is as carcinological as possible (R0)). Amputation is questionable in locally advanced forms [13].

### 3.4. Chemotherapy

Both chemotherapy and radiotherapy have no advantage in terms of overall survival, but they do improve local control in order to avoid mutilating surgery or positive margins, and reduce the rate of recurrence. It may be a solution, particularly in severe cases, as part of palliative treatment [19].

### 3.5. Prognosis

Angiosarcoma has a poor prognosis, progressing rapidly and very aggressively. The 5-year survival rate does not exceed 20% on average [13, 20], and the preferred sites for metastases are bone and lung [12].

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## 4. Conclusion

Angiosarcoma is an angioendothelioma of high grade malignancy, the course of which is usually lethal regardless of the treatment used. Even if radiotherapy and chemotherapy are sometimes used as a complement to surgery, the best guarantee of a chance of survival would be surgery that is as extensive as possible, or perhaps simply radical. The rarity of this bone entity explains the paucity of data available to us for evaluating treatments. The question then arises: should severe angiosarcoma be treated radically from the outset?

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## Compliance with ethical standards

### *Disclosure of conflict of interest*

The authors declare no conflict of interest in relation to the writing of this article.

### *Statement of ethical approval*

The present research work does not contain any studies performed on animals/humans subjects by any of the authors'.

### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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