Case report

LEIOMYOSARCOMA OF THE FEMORAL ARTERY: RARE LOCATION, WHAT MANAGEMENT OF ADVANCED STAGE, REPORT OF A CASE.

Abstract:

Vascular leiomyosarcomas are extremely rare tumours, accounting for less than 1% of all malignant tumours. Their treatment, whatever the stage, requires multidisciplinary management. Surgery with bloc resection remains the treatment of choice for localised tumours; in patients with unresectable locally advanced or metastatic disease, systemic treatment with essentially palliative aims may be proposed. Anthracycline-based treatment is the standard first-line therapy.

We report a clinical case of a 50-year-old female patient with pulmonary and bone relapse of an operated left femoral artery leiomyosarcoma in whom we initiated palliative mono-chemotherapy.

Key words: leiomyosarcoma, femoral artery, chemotherapy, doxorubicin.

INTRODUCTION :

Adult soft tissue and visceral sarcomas (excluding GIST) are rare tumours, with an estimated average incidence of 4-5/100,000/year in Europe. The most common types of soft tissue sarcoma are liposarcoma and leiomyosarcoma, each with an incidence of less than 1/100,000/year, while the majority of sarcoma histotypes have an incidence of less than 2/1,000,000/year. **[12]**

Vascular leiomyosarcomas are extremely rare tumours, accounting for less than 1% of all malignant tumours.Venous leiomyosarcomas occur five times more frequently than arterial leiomyosarcomas. Venous leiomyosarcomas are most often found in large vessels and less than 50% occur in the peripheral circulation. **[1]**

To date, there are very few reported cases of arterial leiomyosarcoma involving the femoral artery, especially in medical oncology. The treatment of leiomyosarcomas, whatever their stage, requires multidisciplinary management.

In this article, we report the case of a 50-year-old patient with stage 4 leiomyosarcoma of the left femoral artery.

CASE PRESENTATION :

This is a 50-year-old hypertensive grade 2 patient on Amlodipine 10mg daily, who consults for a hard, progressively enlarging and slightly painful mass on the medial aspect of the left groin.

An MRI angiography of the thigh showed an endovascular tumour process in the left femoral vein, suggesting in the first instance a leiomyosarcoma involving the femoral artery**(Figure 1).** Angio scan of the aorta and lower limb confirmed the malignant nature of the mass and its locally advanced status, with involvement of the superficial femoral artery and superficial left femoral vein.Anatomopathological study of the biopsy of the mass showed a morphological appearance and immunohistochemical profile in favour of an intermediate grade 11 (3+1+1) leiomyosarcoma according to the FNLCC histological grading. (Figure 2, Figure 3)

A thoracic-abdominal-pelvic CT scan carried out as part of the extension work-up was otherwise unremarkable.

The patient was initially managed by vascular surgeons who performed a complete resection of the tumour mass, then she was put under surveillance with a follow-up scan every 3 months.

Six months after surgery, a follow-up CT scan revealed secondary pulmonary and bone lesions, prompting a consultation with medical oncology for further treatment.

Clinically, she was in good general condition (OMS1). On physical examination, she had a surgical scar on the inside of her left thigh.

In conclusion, we have a 50-year-old patient, hypertensive on treatment, presenting with a metastatic relapse in the lung and bone of a leiomyosarcoma of the femoral artery that had been operated on. The PCR decision concludes that she should receive single chemotherapy with doxorubicin at a dose of 75mg/m^2 every 3 weeks.

DISCUSSION:

Soft tissue sarcomas comprise approximately 80 entities defined by the World Health Organization (WHO) classification on the basis of a combination of distinctive morphological, immunohistochemical and molecular features. **[5]**

First described in 1871 by Perl, vascular leiomyosarcomas are rare tumours. Their prevalence is difficult to quantify: it is estimated at 0.001%. The mean age of onset was 59.5 ± 13.2 years. Leiomyosarcomas affect women 3 times more than men (73% versus 27%). **[2]**

In 2008, an estimated 300 cases were described in the medical literature **[3]**. Peripheral arterial locations are rare. **[1]**

Leiomyosarcomas can give rise to metastases, but few series have been described. In Penel's study of 8 patients, 6 were metastatic. All had pulmonary metastases, two also had bone metastases, two had brain metastases and one patient had an adrenal

metastasis.Median survival is poor for patients with advanced leiomyosarcoma: 8 months median survival with survival ranging from 5 to 20 months. **[4]**

When managing patients with advanced or metastatic soft tissue sarcoma, decision-making is complex, depending on the various histological presentations, and must always be multidisciplinary. Resectable metachronous lung metastases (disease-free interval ≥ 1 year) without extrapulmonary disease are managed by surgery as standard treatment, if complete excision of all lesions is feasible, taking into account all prognostic factors. **[6]**

In patients with unresectable locally advanced or metastatic disease, systemic treatment with essentially palliative aims may be proposed. Anthracycline-based therapy is the standard first-line treatment. There is no formal demonstration that polychemotherapy is superior to single chemotherapy with doxorubicin alone in terms of overall survival. However, according to several randomised clinical trials, but not all, a higher response rate and longer progression-free survival can be expected in a number of sensitive histological types. **[7] [8]**

Consequently, polychemotherapy with an adequate dose of doxorubicin + ifosfamide may be the treatment of choice, particularly in histological types sensitive to ifosfamide, when a tumour response is considered potentially advantageous and the patient's performance status (PS) is good. Doxorubicin plus dacarbazine is an option for first-line polychemotherapy for leiomyosarcoma, in which the activity of ifosfamide is much less convincing, and for soft tissue sarcoma. **[9] [10]**

A phase III study compared doxorubicin monotherapy with the gemcitabine-docetaxel combination as initial treatment in patients with advanced soft tissue sarcoma of all types. The combination showed no improvement in progression-free survival and objective response rate and is not recommended as first-line therapy for patients with advanced soft tissue sarcoma including leiomyosarcoma. **[11]**

In the case of our patient, in the light of all these recommendations, we opted for a protocol of single chemotherapy with doxorubicin at a dose of 75mg/m2 every 3 weeks after a cardiac ultrasound in which the LVEF was 65%. This treatment was combined with zoledronic acid 4mg every 3 weeks after dental treatment and monitoring of renal function.

Therapeutic evaluation after the 3rd cycle of treatment showed stable lesions in the pulmonary and bone metastases on CT scans, and clinical benefit with a reduction in symptoms (bone pain, cough and asthenia). However, during the course of treatment she experienced a few episodes of grade 2 neutropenia, which had no impact on the continuation of treatment.

CONCLUSION :

Leiomyosarcomas are rare tumours with an unfavourable prognosis. Surgery with en bloc resection remains the treatment of choice for localised cases and certain

metastases. However, cases of recurrence are still very common, and if they are unresectable, they should be managed with palliative chemotherapy, preferably a single chemotherapy, to spare the patient the toxicities of systemic drugs which can affect their quality of life.

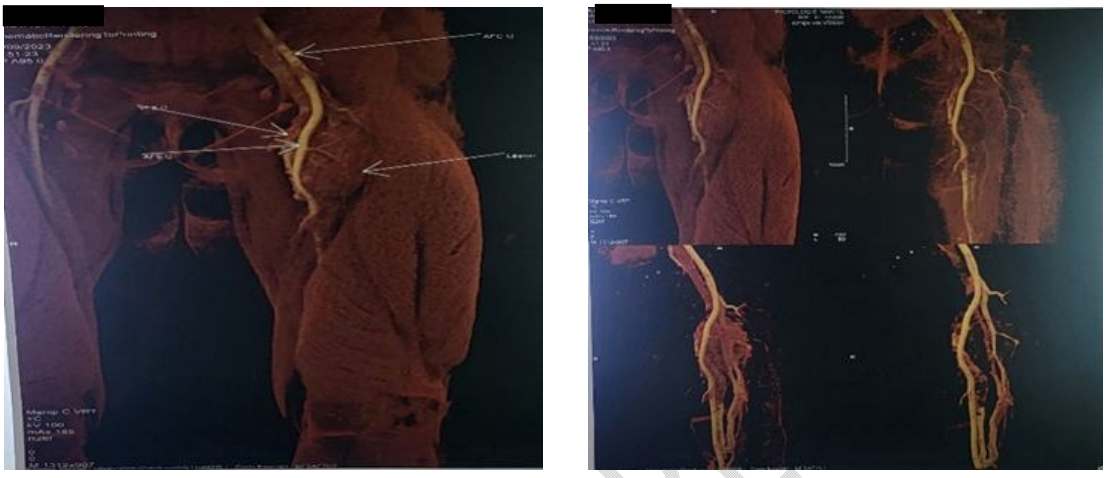

Figure 1 : MRI angiography: endovascular tumour process in the left femoral vein, suggesting in the first instance a leiomyosarcoma involving the femoral artery

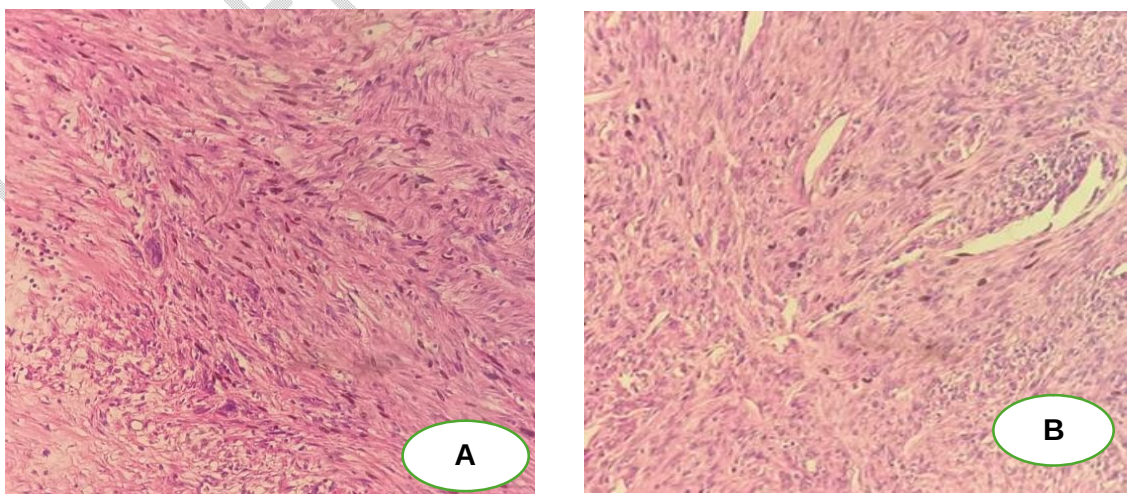

Figure 2 (A and B): Spindle cell tumour proliferation showing atypia and mitosis, HE 40X.

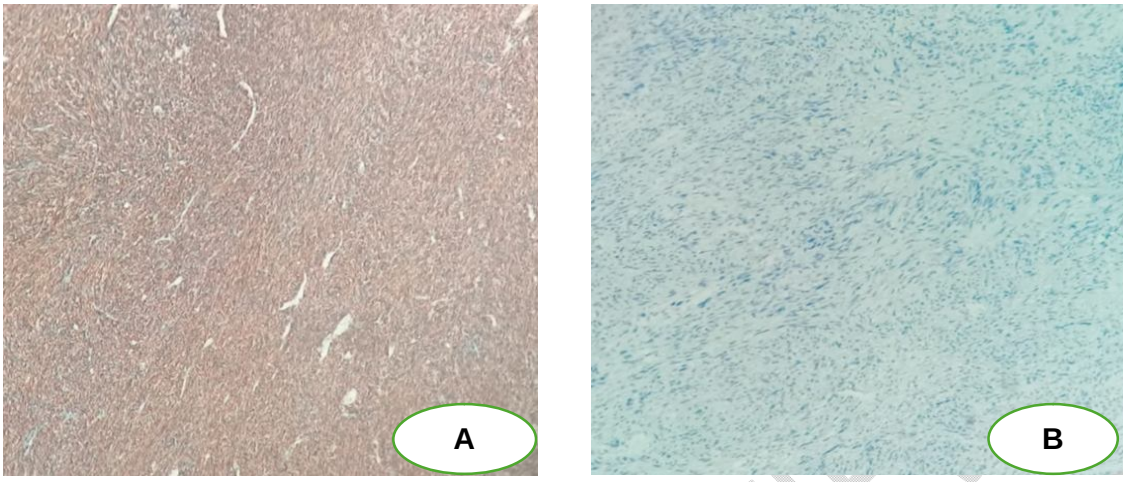

Figure 3 (A and B):

- Image A: positivity of Antibody Anti-AML, H-Caldesmone.
- Image A: absence of expression of Antibody Anti- PS100, Anti-CD31 & CD34.

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