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Rare Location of Ewing Sarcome: A Case Study

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Abstract **Case Report**

Introduction: Ewing's sarcoma is also known as a primary peripheral neuroectodermal tumour and is a primary malignancy accounting for 5-15% of all malignancies. It can involve all skeletal bones and more rarely soft tissue. The area of predilection of this tumour is the long bones 50-60%; the flat bones mainly the pelvis 20%, while the bones of the foot and the hand constitute an exceptional localization (1), hence the interest to report our case. ***Clinical presentation***: We report the case of an 18-year-old girl who presented with a painful swelling of the right heel evolving for 14 months in a febrile context. The diagnosis of Ewing's sarcoma of the calcaneus was suspected by radiological assessment and confirmed by a histological study after a biopsy. We discuss the clinico-radiological and histological diagnosis of this tumour and its management. ***Discussion*:** Ewing's sarcoma described by James Ewing in 1921(2), since then Cook reports only 29 cases of Ewing's sarcoma of the calcaneus in the literature (3), which shows the paucity of the literature concerning this type of calcaneal lesions, and therefore clinicians are unfamiliar with these tumours and therefore a delay in diagnosis, morbidity and amputations unnecessary. It is a tumour with a rapid potential for metastasis, particularly in the lungs, which determines the prognosis of this lesion. ***Conclusion*:** Ewing's sarcoma remains a rare disease (4) of which the calcaneus is an exceptional location. It may be misdiagnosed with other lesions, notably calcaneal osteomyelitis.

Keywords: Ewing's Sarcoma - Calcaneus – Chemotherapy.

INTRODUCTION

Ewing's sarcoma is also known as a primary peripheral neuroectodermal tumour [5], and is a primary malignancy accounting for 5-15% of all malignancies. It can involve all skeletal bones and more rarely soft tissue and the nervous system [1]. The area of predilection of this tumour is the long bones 50-60%; the flat bones mainly the pelvis 20%, while the bones of the foot and the hand constitute an exceptional localization [1], hence the interest to report our case. The treatment of this tumour consists of a combination of chemotherapy and surgery which has improved its prognosis [6].

CASE PRESENTATION

An 18-year-old Moroccan girl with no previous history of the disease was admitted to our clinic with a painful swelling of the right heel and ankle that had been present for 14 months and fever for 4 months. Local examination revealed a tense and firm swelling with very visible subcutaneous veins (figure 1).

Standard radiography showed poorly limited and mitted lytic lesions of the right calcaneus, with a periosteal reaction and spiculated soft tissue infiltration (Figure 2). The CT scan showed multiple geodense hypodense lesions with sunburst soft tissue infiltration (Figure 3).

Magnetic resonance imaging (MRI) detected a poorly limited lesion process of irregular cones with cortical disruption, hyposignal in T1 and hypersignal in T2, heterogeneously enhanced after gadolinium injection.

The study did not reveal any metastatic skips in the surrounding bones (Figure 4). The biological work-up revealed an inflammatory syndrome and an anaemia of 9g/dl. The extension work-up was unremarkable.

For a confirmatory diagnosis, a histological study associated with immunohistochemistry was performed after a biopsy. It was in favour of a Ewing's sarcoma with a strong expression of the anti CD99 antibody (figures 5 and 6).

After a multidisciplinary consultation meeting, and the consent of the patient and her family, the

management was neoadjuvent chemotherapy with a mid-leg amputation. 8 weeks later, the patient benefited from a limb prosthesis with good social integration. 15 months follow-up, no local or distant recurrence.

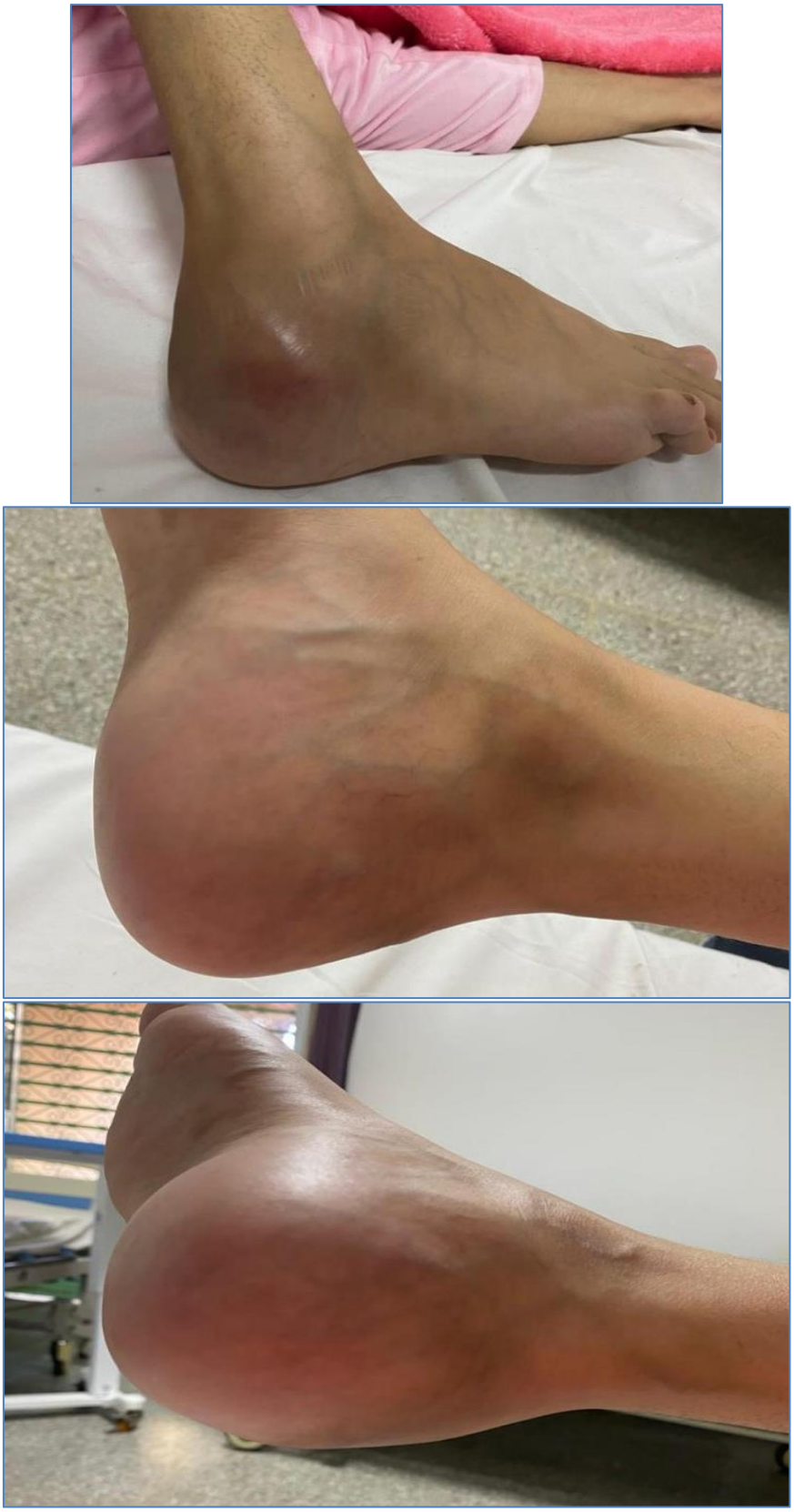


Figure 1: Clinical appearance of the lesion

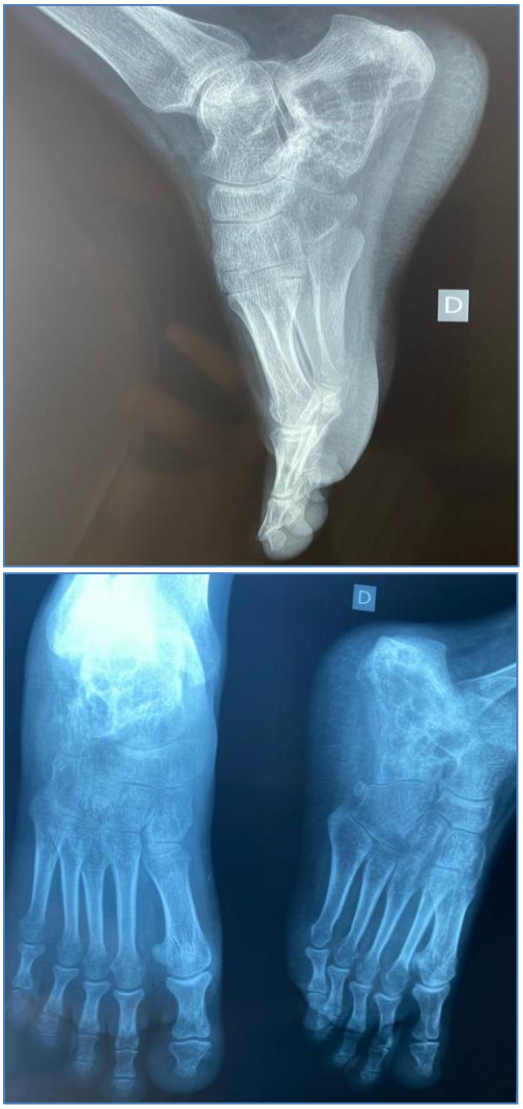
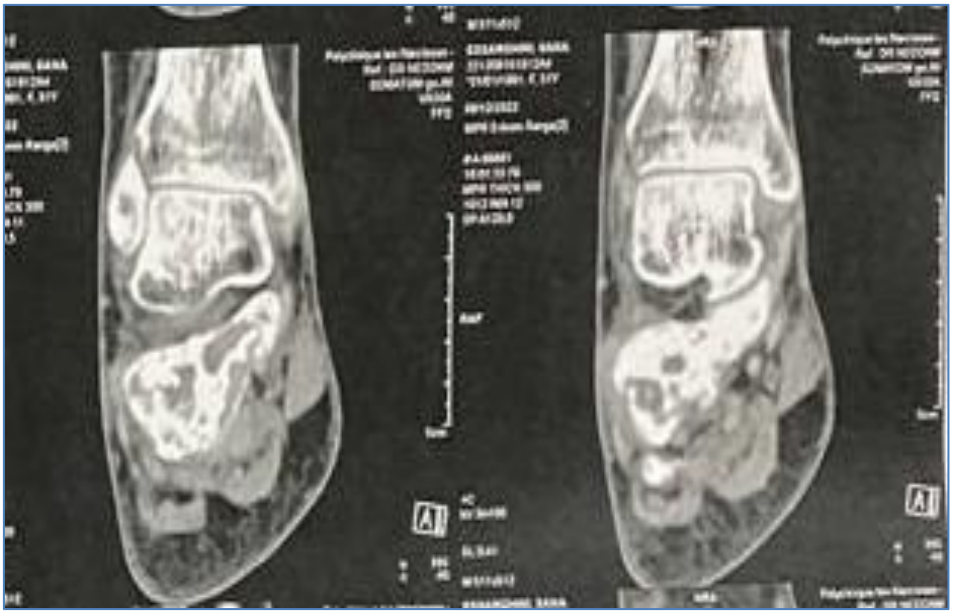


Figure 2: tandard radiograph showing mitted lytic lesions with spiculated soft tissue infiltration



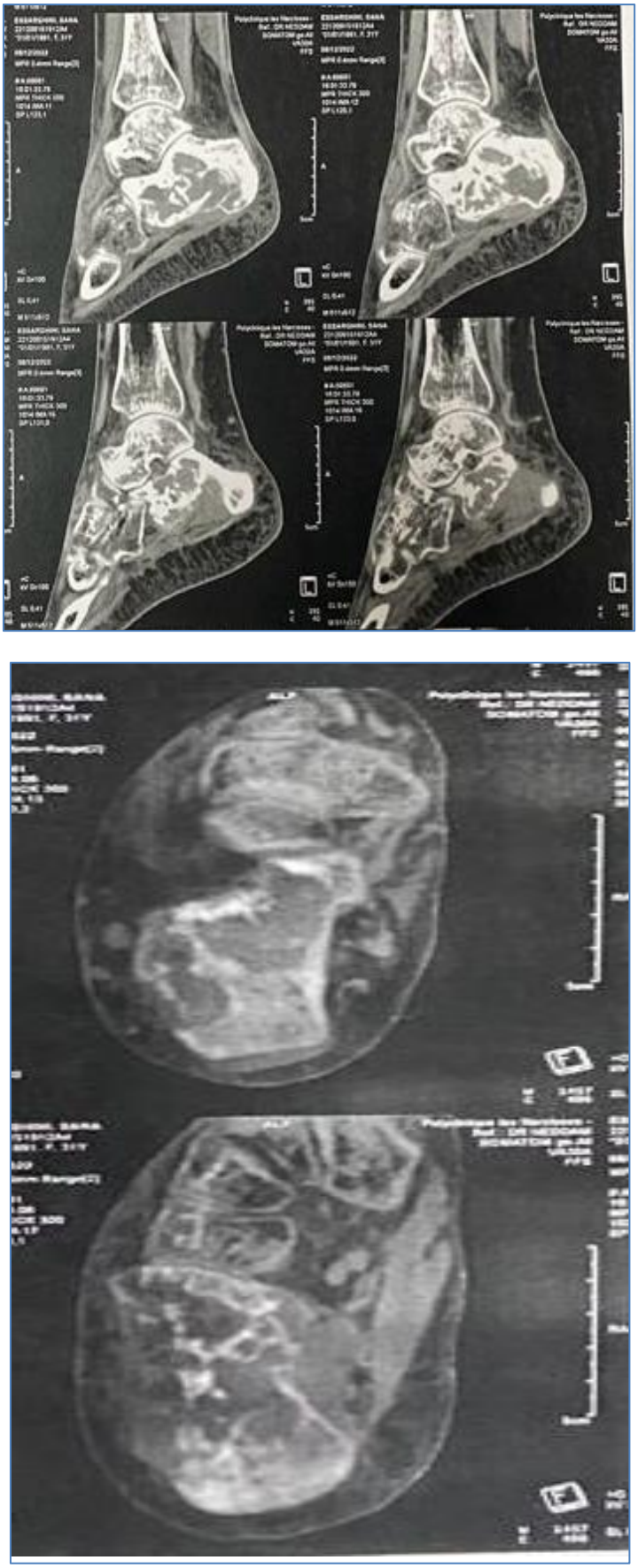


Figure 3: CT scan of right calcaneus showing hypodense lesions with sunburst appearance

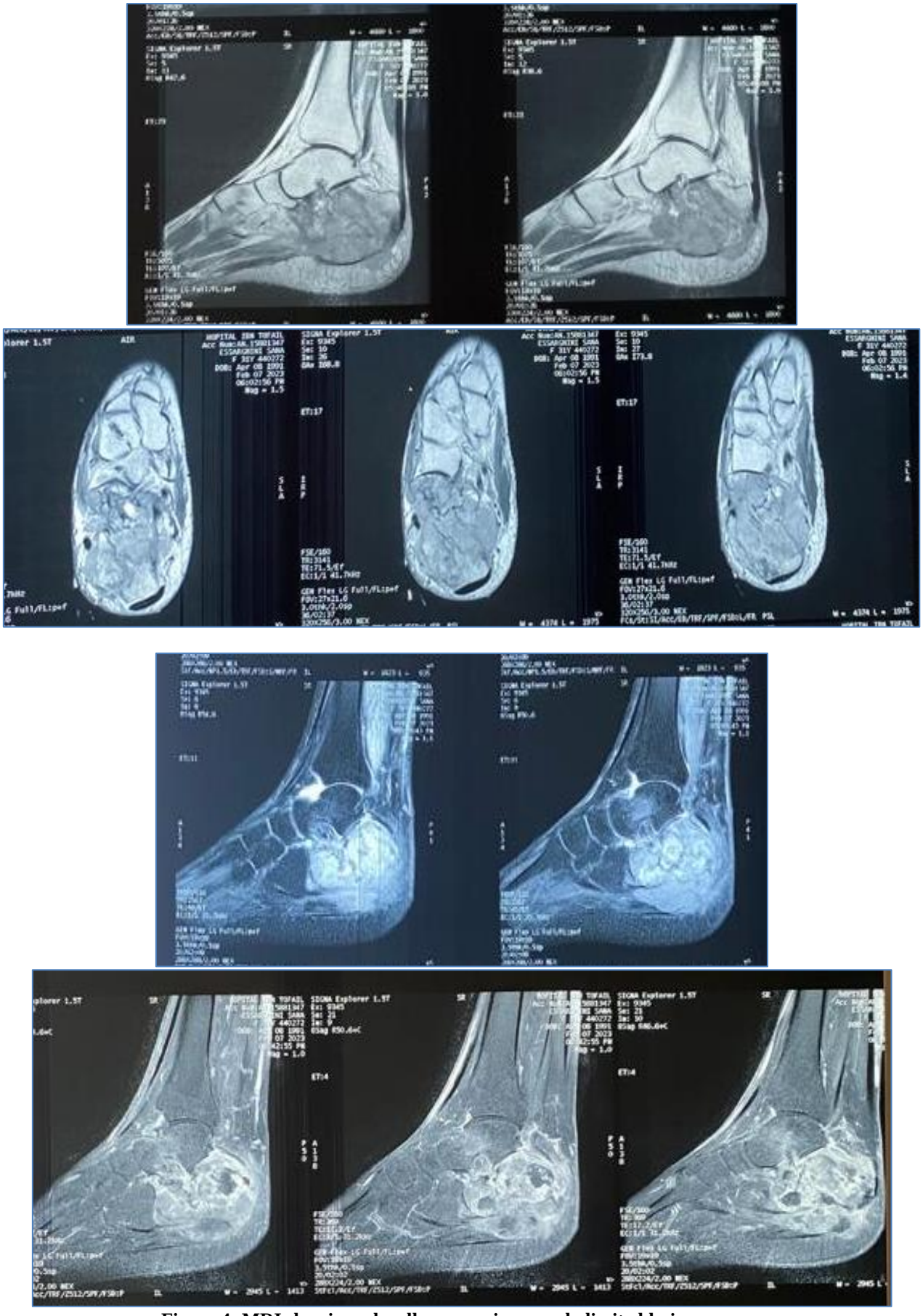


Figure 4: MRI showing a locally aggressive, poorly limited lesion process

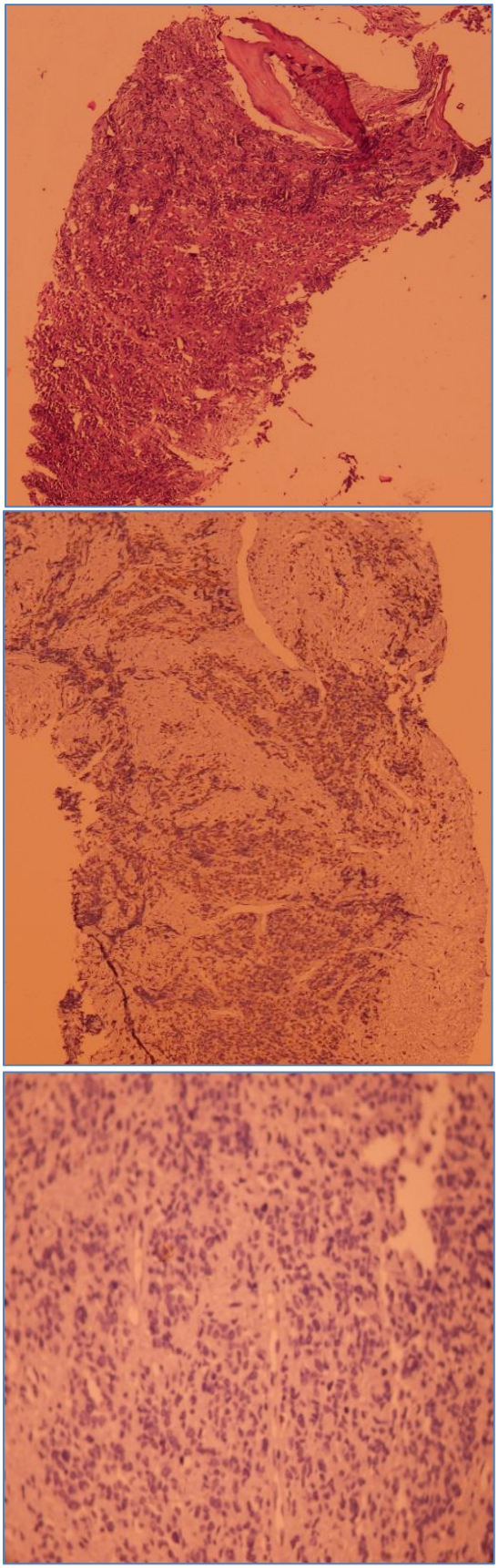


Figure 5: Histological appearance in favour of Ewing's sarcoma

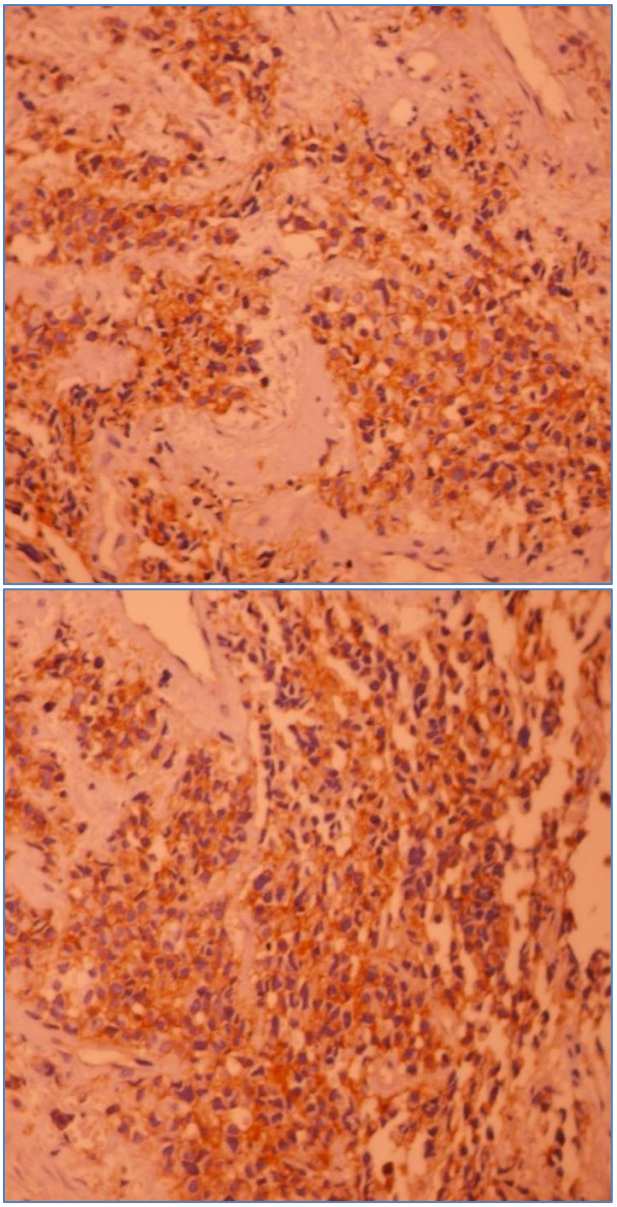


Figure 6: Immunohistochemistry with high expression of anti-CD99 antibody

DISCUSSION

Ewing's sarcoma is a primary malignant bone tumour, described by James Ewing in 1921[2], since then Cook reports 29 cases of Ewing's sarcoma of the calcaneus in the literature [3], which shows the paucity of the literature on this subject, and therefore clinicians are unfamiliar with these tumours, so we still see diagnostic delays, unnecessary morbidity and amputations, in addition to the use of traditional medicine. The foot remains a rare location for malignant tumours accounting for less than 1% of all bone and soft tissue sarcomas, in particular primary malignant tumours of the calcaneus are extremely rare [7].

Ewing's sarcoma is the second most common type of primary malignancy [8], often affecting children and adolescent males with a sex ratio of 1.5 [9]. The aetiology of this tumour is still unknown, however some genetic factors are incriminated such as the translocation involving the EWS gene carried on chromosome 22 and the FLI-1 gene carried on chromosome 11 [10]. This may explain the frequency of this lesion in Caucasians and its rarity in Africans, testifying in addition to the rarity of our case.

It is a tumour of neuroectodermal differentiation characterised by a proliferation of small round cells, with a rapid metastatic potential where the lung is the first location, hence the interest of an extension assessment. It is also characterised by aggressive invasion of the surrounding soft tissue. Current treatment is based on a combination of multidrug therapy and surgery, which may be amputation or extensive resection. The advent of chemotherapy has improved the prognosis of this tumour, which used to survive less than 10% due to metastases despite radical

surgery or irradiation [1]. If left untreated, the outcome is rapidly fatal.

Its prognosis depends essentially on the presence or absence of metastases, with certain factors such as fever, biological inflammatory syndrome and anaemia being found to have a poor prognosis [11]. The probability of recurrence-free survival at 5 years is 78% if there is no metastasis; 50% if there is an isolated lung metastasis, and 30% if there are multiple metastases [12]. There is always a risk of lung metastases even if an amputation is performed, hence the importance of good surveillance [13].

CONCLUSION

The calcaneus is an atypical location for Ewing's sarcoma, which may be misdiagnosed as osteomyelitis. Early recognition of this atypical feature is necessary for its proper treatment and successful outcome.

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