Ewing's Sarcoma in a young Malian

Sarcome d'Ewing chez un jeune Malien

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RÉSUMÉ

Le sarcome d'Ewing est une tumeur osseuse maligne primitive rarement observée chez les populations noires. Nous rapportons un cas de sarcome d'Ewing chez un garçon de 20 ans, se manifestant par un gonflement douloureux de l'épaule gauche avec impotence fonctionnelle évoluant depuis 2 ans. Le patient a été transféré pour soins au service de rhumatologie de l'hôpital universitaire du Point G le 13 février 2012. Le diagnostic de sarcome d'Ewing avec métastase vertébrale (L1) a été posé après un examen pathologique de la biopsie osseuse.

Mots-clés : sarcome d'Ewing, Humerus, Malien

ABSTRACT

Ewing's sarcoma is a primary malignant bone tumour rarely observed in black populations. We report a case of Ewing sarcoma in a 20-year-old boy, manifested by painful swelling of the left shoulder with functional impotence evolving for 2 years. The patient was transferred for care to the Rheumatology Department of Point G University Teaching Hospital on February 13, 2012. The diagnosis of Ewing's sarcoma with vertebral (L1) metastasis was retained after a bone biopsy pathological examination.

Keywords: Ewing's sarcoma, Humerus, Malian

Introduction

Described by James Ewing in 1921, Ewing's Sarcoma is the second most common primary bone tumour after osteosarcoma, accounting for 3 % of all childhood malignancies [1], typically affecting the trunk and long bones. It usually affects children mostly male of 5-30 years of age with a peak of incidence between 10 and 15 years. Ewing's sarcoma is rarely observed in black populations [2]. Rarely, 1-2% of Ewing's sarcomas may involve epiphysis [3]. We present a case of Ewing's sarcoma of the proximal humeral epiphysis in a 20 -year- old boy.

Case report

O.F, a 20-year-old farmer, with no known surgical history, treated for 3 weeks with anti-tuberculosis drugs for a probable humeral mycobacterial osteomyelitis. He was referred and admitted to the Rheumatology Department of Point G University Teaching Hospital on February 13, 2012, for a pyretic painful swelling of the left shoulder with functional impairing, evolving for 2 years. He also suffered from low back pain and deterioration of the general condition.

On examination, there was a marked increase in the soft tissue swelling in the left shoulder with severe tenderness (Figure 1) on the spine. We have noted a dorsal gibbosity and marked lumbar spinal syndrome with a schöber index at 10 + 2 cm without neurological deficiency. There was weight loss of 3 kg as well. Biology studies revealed inflammatory anaemia, CRP > 15 mg/L, accelerated ESR at 80 mm, hypercalcemia at 2.7 mmol/L,and hyperuricemia at 449 µmol/L. Myelogram was normal. Tuberculosis research was negative. Liver and renal functions were normal.

Radiographs revealed a lytic lesion within the right proximal humeral epiphysis, a periosteal reaction taking a grass fire aspect with soft tissues invasion (Figure 2); and vertebral (L1) osteolytic images were also noted. Chest X-ray was normal. Ultrasound of the left shoulder shows collected deltoid myositis. Bone biopsy was performed and histological study showed clusters of small hyperchromatic round cells of lymphocytic appearance and oval vesicular nuclei (Figure 3). Immunohistochemical staining is not feasible in Mali. The diagnosis of Ewing's sarcoma with vertebral (L1) metastasis was retained. Prior to chemotherapy, oncologists had planned disarticulation, which was rejected by the patient. He received an infusion of zolendronic acid and he did not come at the next appointment.

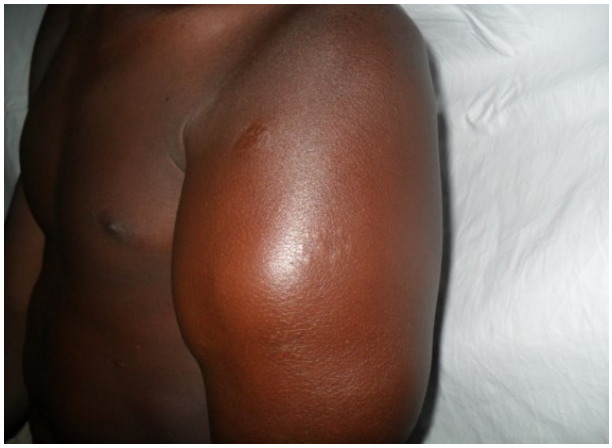

Figure 1. Soft tissue swelling in the left shoulder with severe tenderness.

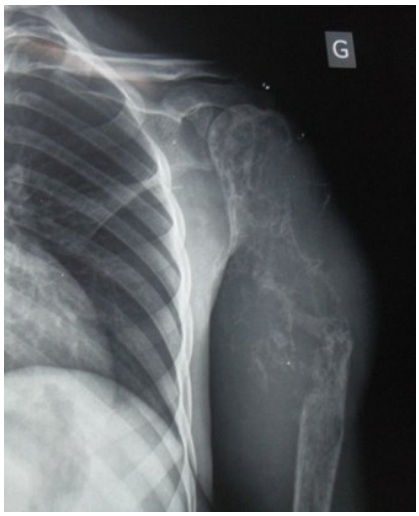

Figure 2. Lytic lesion within the right proximal humeral epiphysis, a periosteal reaction in grass fire with soft tissues invasion.

Discussion

We report a case of Ewing's sarcoma in a 20-year-old boy with no known family history. This is the only case in the department of Rheumatology in 15 years and first reported case in Mali, to our knowledge. It confirms the rarity of Ewing's tumour in black populations [2,4]. Ewing's Sarcoma is the second most common primary bone tumour after osteosarcoma, accounting for 3% of all childhood malignancies [1]. Ewing's sarcoma has a predilection for the male sex (male/ female ratio, 1.3–1.5:1) [4]. It occurs in a wide range of ages from infants to the elderly, although approximately 80% of patients are younger than 20 years [4]. Peak incidence is during the second decade of life, although 20–30% of cases are diagnosed during the first decade [4].

Ewing's sarcoma is ubiquitous, preferentially affecting flat bones and is most often associated with extensive soft tissue extension: pelvis (26%), thorax (rib, clavicle, scapula, 20%), femur (16%), tibia (9%), spine (8%), humerus (5%) and skull (1%) [5,6]. Rarely, 1%–2% of Ewing's sarcoma may involve epiphysis [3]. Ewing's Sarcoma has a strong potential to metastasize, most commonly occur in the lungs and bone and more than 10% of patients present with multiple bone metastases at initial diagnosis [4].

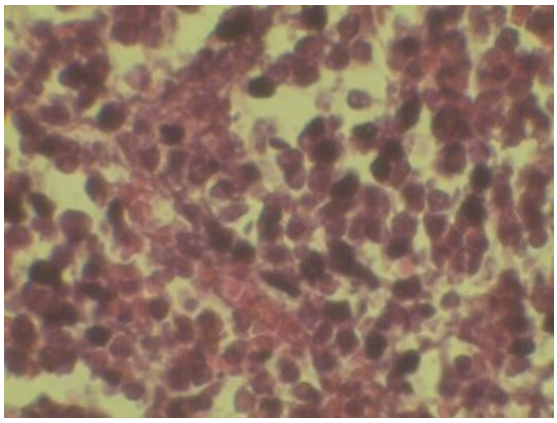

Figure 3. Clusters of small hyperchromatic round cells of lymphocytic appearance, oval vesicular nuclei.

The earliest symptom was a painful swelling of the left shoulder, accompanied by fever followed by low back pain and deterioration of the general condition. In the literature, the most common presenting symptoms in a patient with Ewing's sarcoma include pain, swelling, or a mass. Approximately 20% of patients present with fever, which may lead to the mistaken diagnosis of osteomyelitis [7].

Laboratory studies may reveal anaemia, leucocytosis, or an increased erythrocyte sedimentation rate [7]. The initial imaging investigation of a suspected bone tumour is a radiograph in two planes. It revealed lytic lesion within the right proximal humeral epiphysis and periosteal reaction, suggesting a diagnosis of primary malignant tumour. These lesions are also reported during eosinophilic granuloma, Burkitt lymphoma, fibrous dysplasia, aneurysmal cyst, giant cell repair granuloma [6]. Typically, Ewing's sarcoma appears as an ill-defined, permeative, or focally mouth-eaten, destructive intramedullary lesion accompanied by a periosteal reaction that affects the diaphysis of long bones [4].

The patient is staged for both local and metastatic disease. A vertebral (L1) metastasis was associated. Diagnostic staging should include a CT scan of the chest to determine pulmonary metastases and a technetium-99m whole-body radionucleotide bone scan to identify skeletal metastases [4, 6]. This could not be done in our patient because of the cost of these exams.

In Africa, the endemicity of tuberculosis may lead to mistaken diagnoses, as was the case with our patient. The definitive diagnostic method is biopsy. Pathological examination in our patient described small round cells suggestive of Ewing's Sarcoma [8]. Histologically, Ewing's sarcoma is composed of a homogeneous population of small round cells with high nuclear to cytoplasmic ratios that are arrayed in sheets [4]. Unfortunately, cytogenetic examination was not feasible on site.

The patient had only received an infusion of zolendronic acid. He had refused surgical amputation of the limb before the chemotherapy. Tremendous strides have been made in the treatment of Ewing's sarcoma [7], as approximately 70% of patients are long-term survivors. A multi-disciplinary team approach is necessary to combine chemotherapy, surgery, and/or radiation in the care of the patient. The presence of distant metastasis at diagnosis is the most unfavorable prognostic factor [4] in Ewing's sarcoma, and the patient had. Even with aggressive treatments, patients with metastases have only an approximately 20% chance of long-term survival [4].

Conclusion

Ewing's sarcoma remains a relatively rare disease in Mali. Histopathology confirms the diagnosis. Chemotherapy, surgery and/or radiotherapy remain the mainstay of treatment. Prognosis remains unfavorable in Africa, due to the delayed diagnosis and the expensive cost of treatment.

Conflicts of interest: None declared.

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