Extra Osseous Ewing's Sarcoma: A Case Report

Corresponding Author:
Dr. Najib Alidrissi,
Orthopedic and trauma surgeon, University Mohammed V - Souissi, 10000 - Morocco

Submitting Author:
Dr. Najib Alidrissi,
Orthopedic and trauma surgeon, University Mohammed V - Souissi, 10000 - Morocco

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Author(s): Alidrissi N

Abstract

We report a case of extraosseous Ewing's sarcoma localized in the gluteal region. The treatment consisted of chemotherapy to reduce tumor size followed by surgery.

Introduction

The extra-osseous Ewing's sarcoma (EES) is a rare tumor, which is derived from neural crest cells and integrates within the peripheral neuroectodermal tumors. (1,2) These tumors have common cytogenetic features. (2, 3)

Case Report(s)

Mr. A., aged 24 has a swelling on the right buttock, painless at rest, pain with walking. The examination was unremarkable. Plain radiographs of the hip and pelvis revealed no abnormalities in the bone (Fig. 1). The examination of the pelvis CT scan can visualize a hypodense expansive process, developed in the right gluteal muscles, without osteolysis next (fig2). The assessment of extension is normal. The anatomopathological study performed after surgical biopsy of the mass confirmed the diagnosis of extra-osseous Ewing's sarcoma. (Fig. 3) Chemotherapy was administered before the surgical treatments in the patient then underwent surgery two months later after discontinuation of chemotherapy and has received wide resection (Fig. 4), the sciatic nerve was independent of tumor mass. The immediate evolution was marked by a postoperative infection consequent to immunosuppression of the patient was under chemotherapy and has been controlled by appropriate antibiotic therapy and local care.

Discussion

Unlike Ewing intraosseous, which is common in males, no predisposition in terms of sex is present. Two thirds of tumors diagnosed occur in young adults. On the etiopathogenic, q12 translocation of chromosome 22 is found in 95-100% of cases. (4, 5, 6) Clinical signs are nonspecific. The topography is ubiquitous but preferentially reached the paravertebral region, the thoracic wall, retroperitoneum and lower extremities rather than higher. (7, 8, 9, 10) Radiologically: nonspecific mass. Most lesions are encapsulated, hypoechoic on ultrasound, hypodense on CT, it is often hyper-vascularized, so hyperdense after injection of contrast material. In MRI, the signal intensity of the tumor is iso-hypointense on T1, hyperintense on T2. (3, 11, 12) The diagnosis is the cytogenetics and immunohistochemistry, particularly through in situ hybridization by immunofluorescence. The treatment combines surgery, chemotherapy and radiotherapy. (14, 15) The Ewing's sarcoma have developed rapidly, neoadjuvant treatment (radiotherapy and chemotherapy) is made ??of this very useful in reducing tumor size and allow complete removal. The 10-year survival is possible, it would be at around 62 to 77%. The intensification of chemotherapy more aggressive may explain this difference. The term remission is possible. (16)

Conclusion

The EES is a rare, interesting young adults, the diagnosis is difficult even on the histology. It is very close neuroectodermal tumors in terms of ultrastructure, cytogenetics, biochemistry and immunology. In the absence of specific radiological signs, it seems necessary to include in the differential diagnosis of any primary tumor of soft parts Imaging, MRI, in particular, allows an assessment of the lesions and monitor therapy. Ewing sarcoma bone deserves extra-early diagnosis, which can offer the best chance of survival.

Abbreviations(s)

EES: extra-osseous Ewing's sarcoma
MRI: Magnetic resonance imaging

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Illustrations

Illustration 1

Fig1: normal radiograph of the hip

Illustration 2

Fig2: CT of the pelvis revealing a hypodense expansive process, developed in the right gluteal muscles, without osteolysis
Illustration 3

Fig 3: Surgical specimen.

Illustration 4

Fig 4: wide resection of the tumor, suture the rest of the gluteus maximus over the greater trochanter.
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