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Extraosseous Ewing's Sarcoma in the thigh A *case report and review of the literature*

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ABSTRACT

The extraosseous Ewing's sarcoma is a rare tumor. The authors reported a case on the thigh level, and discussed; in the light of a review of the literaturethe etiopathogenic, diagnosis and therapy aspects of this tumor.This case is about a 13 years old child, who suffered from a posttraumatic pain in the right thigh. The Standard radiography was normal. MRI showed a tumor mass extending to the bone. A surgical biopsy and histological examination confirmed a diagnosis of an extraosseous Ewing's sarcoma. The patient died after chemotherapy.Ewing's sarcoma remains a relatively rare disease. Its diagnosis is, currently, based on histology and molecular biology's data. Its Treatment is based on the combination of chemotherapy surrounding the complete surgical resection of the tumor. His prognosis was significantly improved with the help of modern therapeutics.

Key words: Extra-skeletal Ewing's; sarcoma; Immunohistochemistry.

INTRODUCTION

Extraosseous Ewing's sarcomas (EES) are rare tumors (5% of soft sarcomas tissue) that occur predominantly in adolescents and young adults between 10 and 30 years. Paravertebral area and the extremities are the locations most frequently encountered. We report one case with review of literature.

MATERIALS AND METHODS

Case report

This concerns a child of 13 years old without significant pathological history, seen in consultation for painoriginally post-traumatic in the right thigh and persistent at rest. Clinical examination noted a predominant firm and sensible swelling in the anterolateral surface of the thigh. The patient was apyretic, and generally, in goodhealth condition.

A standard X-ray was normal (fig 1). MRI highlighted an important tumoral mass in the soft parts of the thigh extending to the bone (fig 2). A surgical biopsy was performed and the histology favored extraosseous Ewing's sarcoma. The patient died from complications of chemotherapy.

RESULTS AND DISCUSSION

Ewing's sarcoma is a variety of sarcoma originally called reticuloendothelial. It may evolve in the bone marrow: bone sarcoma develops among children from 10 to 14 years old. There are also extraosseous localizations [1]. The first case of EES was reported by Tefft[2]. The boys are more often affected than girls. Allam[3] reported a sex ratio of 2.4: 1 in a set of 24 patients.
Contrary to osseous form where the average age of bone sarcoma occurrence is around 20 years old. EES is diagnosed at an average age of 16.5 years old [3]. However cases have been reported among patients aged 14 months and 63 years old [4, 5]. In most cases the EES affects paraspinal area, lower limbs, chest wall and pelvis, rarely the head and neck region [5, 6, 7].
The EES usual revealing symptom is pain, almost always present, which can occur in a context of trauma as in our case and therefore gives a misleading picture.The second symptom which can motivate the consultation is the existence of a swelling mass, rapidly increasing size. It can become very important [8], but it is not uncommon to discover the disease at the metastasis stage.
The paraclinical record includes standard radiography, ultrasound which highlights a well-circumscribed lesion, Hypoechoic. The CT scanconfirms an extra bone mass character and absence of osteolysis. ButMRI is more accurate; the MR images on T1W1 appearhypointense to isointense and hyperintense on T2W1. Howeverthese pictures are nonspecific [9].

The certain diagnosis is brought by histology which highlighted that small round cells with oval nuclei and cytoplasm extremely reduced [8].

The differential diagnosis is discussed with other small round cell tumors in particular rhabdomyosarcoma, primitive neuroendocrine tumors peripherals (PNET), carcinoma Merkel cell and lymphoma [10].

Recent advances in immunohistochemistry and cytogenetics have helped in the diagnosis of EES.Identification of the MIC2 gene with CD99 marker and positive monoclonal antibody O13 staining to cell surface glycoprotein p30/32 is highly suggestive of EES, mainly distinguishing it from osseous Ewing's sarcoma [11, 12, 13]. Even though;neuroblastoma, embryonic rhabdomyosarcoma and lymphoma may also occasionally show positive staining for O13, they are excluded by other immunohistochemical stains [11, 12, 13]. Another diagnostic feature of EES is the demonstration of a reciprocal cytogenetic translocation involving an exchange of chromosomal material and creation of novel chimeric genes between chromosomes 11 and 22 [t(11;22) (q24,q12)] which is present in 85-90% of Ewing's sarcoma [14,15].

EES usually follows an aggressive course with a high incidence of recurrence. Regional nodal spread rarely occurs while distant metastasis most commonly affects the lung and bone. Because of its rarity, there is a paucity of data on the optimal management of EES. The traditional "surgical excision only" treatment leads to a high mortality rate whencompared to combined modality therapy, which has a disease-free survival rate of 83% in patients with truncal lesions [4, 5, 16] Current evidence suggests wide surgical resection followed by chemo-irradiation offers the best chance for the disease control [5,16,17]. Younger age at diagnosisand wide surgical resection margins appear to be significant positive prognostic factors for EES whilst size, location and stage of the tumor do not seem to influence survival [17].

CONCLUSION

EES is a rare form of soft tissue sarcoma.The results of the present study show that EES is an aggressive disease with a high incidence of local recurrence and distant metastases. Metastases at presentation and bulk of disease are among the most important prognostic factors influencing the treatment results. Therefore, Non-mutilative surgical treatment with negative resection margins, together with the use of aggressive combination chemotherapy can give the best chance of cure in thisrare type of diseas.

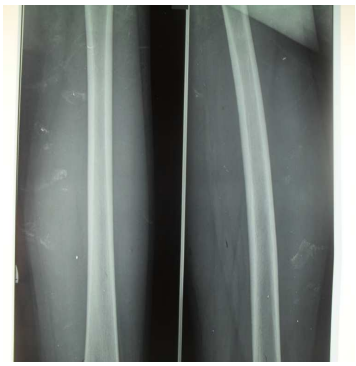

Figure 1: standard radiography of the right thigh was normal

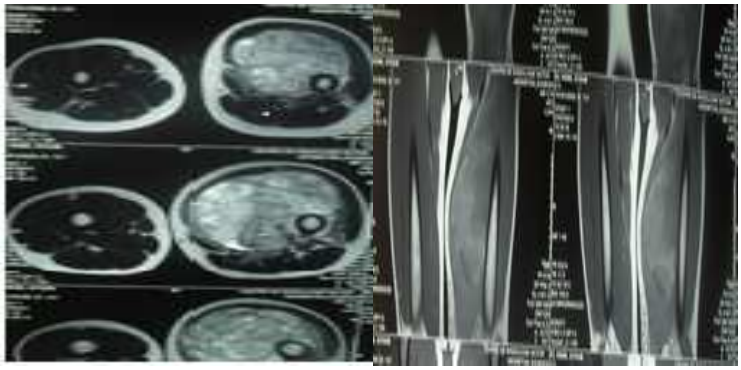

Figure 2: MRI of the right thigh highlighted an important tumoral mass in the soft parts extending to the bone

REFERENCES

[1]. François Natali , Gwenaël Couchouron, Nicolas Paleiron, Michel Andre, Philippe Mondine Sarcome d'Ewing extraosseux. Situation clinique douloureuse **2008** ; 9 : 34-39

[2]. Tefft, M., Vawter, G., Mitus, A. Paravertebral "round cell" tumors in children. Radiology 92:1501-1509, **1969**.

[3]. A. Allam, G. El-Husseiny, Y. Khafaga, A. Kandil, A. Gray, A. Ezzat, et al., Ewing's sarcoma of the head and neck: a retrospective analysis of 24 cases, Sarcoma 3 (**1999**) 11–15.

[4]. L. Angervall, F.M. Enzinger, Extraskeletal neoplasm resembling Ewing's sarcoma, Cancer 36 (**1975**) 240–251.

[5]. N.P. Rud, H.M. Reiman, D.J. Pritchard, F.J. Frassica, W.A. Smithson, ExtraosseousEwing's sarcoma: a study of 42 cases, Cancer 64 (**1989**) 1548–1553.

[6]. Angervall L, Enzinger FM. Extraskeletal neoplasm resemblingEwing's sarcoma. Cancer **1975**;36:240e51.

[7]. Soule EH, Newton Jr W, Moon TE, et al. ExtraskeletalEwing's sarcoma: a preliminary review of 26 cases encounteredin the Intergroup Rhabdomyosarcoma Study. Cancer **1978**;42:259e64.
[8]. Encyclopédie médico-chirurgicale, appareil locomoteur, sarcome d'Ewing,14-756, P :2 les localisations inhabituelles des sarcomes chez l'enfant
[9]. F.ElOunani, R Dafiri – Service de radiologie – hôpital d'enfant – Maternité CHU, Rabat – Maroc
[10] Gillespie JJ, Roth LM, Wills ER, et al. Extraskeletal Ewing's sarcoma: histological and ultrastructural observations in three cases. Am J SurgPathol **1979**;3:99e108.
[11]. Boor A, Jurkovic I, Friedmann I, et al. ExtraskeletalEwing'ssarcoma of the nose.J LaryngolOtol**2001**;115:74e6.
[12]. Yang YS, Hong KH. Extraskeletal Ewing's sarcoma of thelarynx.J LaryngolOtol**2004**;118:62e4.
[13]. Weidner N, Tjoe J. Immunohistochemical profile of monoclonalantibody O13: antibody that recognizes glycoproteinp30/32MIC2 and is useful in diagnosing Ewing's sarcoma andperipheralneuroepithelioma. Am J SurgPathol**1994**;18:486e94.
[14]. Bridge JA. Cytogenetics and experimental models of sarcomas.CurrOpinOncol1995;7:333e9.
[15]. Aurias A, Rimbaut C, Buffe D, et al. Chromosomal translocations in Ewing's sarcoma.NEngl J Med **1983**;309:496e8.
[16]. Kennedy JG, Eustace S, Caulfield R, et al. Extraskeletal Ewing's sarcoma: a case report and review of the literature. Spine **2000**;25:1996e9.
[17]. Ahmad R, Mayol BR, Davis M, et al. ExtraskeletalEwing'ssarcoma.Cancer**1999**;85:725e31.