**Scholars Journal of Medical Case Reports**

OPEN ACCESS

Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

# Extra-Skeletal Ewing Sarcoma: Report of Two Cases

Rim Mahad*, Meriem Ouali Idrissi, Lkbir Abidine, Badre Boutakioute, Najat Cherif Idrissi El Ganouni

Radiology Department, Arrazi Hospital, Mohammed VI University Hospital of Marrakech, Morocco

**DOI:** 10.36347/sjmcr.2021.v09i05.001 | **Received:** 21.03.2021 | **Accepted:** 29.04.2021 | **Published:** 05.05.2021

***Corresponding author:** Rim Mahad

**Abstract** **Case Report**

An extra-skeletal Sarcoma (EES) is a rare and rapidly progressive mesenchymal cell tumor in young patients. Histological characteristics are similar to those of Ewing bone sarcoma. Imaging is essential for early diagnosis and evaluation of the preoperative response. Here, we report two cases of young patients in which imaging and histology determined that extra-skeletal sarcoma was involved.

**Keywords**: Ewing's sarcoma, extra-skeletal, magnetic resonance imaging.



## INTRODUCTION

Extra-skeletal Ewing's sarcoma is a rare entity. It is part of the Ewing family of tumors (EFTs), which also includes bony Ewing's sarcoma and Askin's tumor. It is a round cell tumor of high grade of malignancy, which preferentially affects adolescents [1]. MRI is the modality of choice in the assessment of regional and local extension and evaluation of preoperative response [2]. Treatment of localized forms is based on chemotherapy, surgery and/or radiation therapy.

## CASE REPORTS

### Case1

A 14-year-old child, without any particular pathological history, who has been presenting for 3 months with a swelling of the right leg that is progressively increasing in volume. The clinical examination showed a huge firm, insensitive mass, fixed in relation to the superficial plane, without inflammatory signs opposite. Standard radiography showed a densification of the soft parts of the right leg. Additional magnetic resonance imaging revealed an intra-compartmental spindle-shaped lesion in the anterior compartment of the leg with a discrete T1 hypersignal, T2 hypersignal and heterogeneous enhancement after contrast. The biopsy with anatomopathological study concluded to a Ewing's sarcoma. The extension workup was negative. The patient was put on chemotherapy [Fig 1].

### Case2

This is a 21-year-old girl, with no particular pathological history, who consulted for a mass in the right shoulder that had been evolving for one year, painless, with no associated signs. The biological workup was normal. Ultrasound examination of the soft tissues showed a large mass in the right supraclavicular fossa, measuring 7x8 cm, oval, with heterogeneous hypoechoic lobulated contours, lighting up in places on color Doppler. A complementary magnetic resonance imaging study showed a large necrotic mass in the right supraclavicular fossa, with well-defined lobulated contours, surrounded by a pseudo capsule, intensely and heterogeneously enhanced by gadolinium, delimiting areas of liquid signal within the mass, with liquid-liquid levels. The biopsy with anatomopathological study concluded to a Ewing's sarcoma [Fig 2].

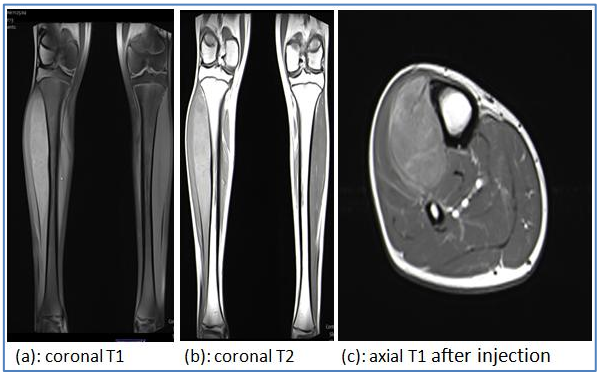


**Fig-1: Knee MRI: Deep subaponeurotic tissue lesion in discrete T1 hypersignal, T2 hypersignal and heterogeneous enhancement after contrast**

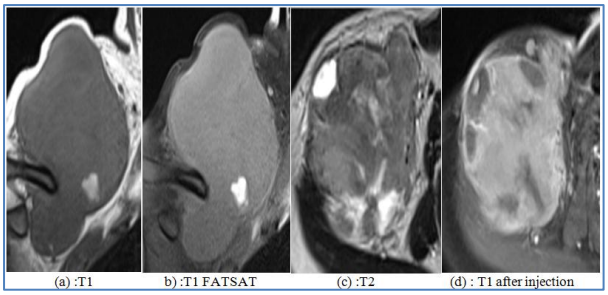


**Fig-2: MRI of the supraclavicular region: Subaponeurotic tissue lesion in T1 hypersignal with hemorrhagic areas in T1 hypersignal, heterogeneous T2 hypersignal and heterogeneous enhancement after contrast**

## DISCUSSION

Ewing's sarcoma is a malignant bone tumor of children and young adults, first described in 1921. It belongs to the family of neuro-ectodermal tumors that share a chromosomal translocation (22, 11) [2]. Preferentially located in flat bones and most often with significant soft tissue extension. Rarely from soft tissue [1-3]. Extraskeletal sarcoma can localize in all soft tissues, however it seems to have a predilection for the paravertebral region, the chest wall, the lower limbs and the pelvis [4]. Medical imaging plays a very important role in determining the topography of the tumor, its relationship with nearby organs, and in the assessment of its extension. In particular, on MRI, extraskeletal sarcoma is hyposignal T1 and hypersignal T2 and is heterogeneously enhanced after injection of contrast medium. The definitive diagnosis of EES is histological due to nonspecific clinical and radiological characteristics [4, 5]. Histologically it is a small round cell tumor with CD99 positive on immunohistochemistry. The genetic mutations described above can be observed by fluorescence in situ hybridization (FISH) or by reverse transcription [6, 7]. The Management of EES is based on chemotherapy, surgery and radiotherapy allowing a long life expectancy.

## CONCLUSION

Extra-skeletal Ewing's sarcoma is a rare tumor of children and adolescents with difficult diagnosis. Anatomopathology and cytogenetics remains the mainstay of diagnosis, and MRI is the modality of choice in the initial workup, the assessment of extension and the evaluation of the preoperative response. An early diagnosis and multidisciplinary management for better results.

## REFERENCES

1. Taylor M, Guillon M, Champion V, Marcu M, Arnoux JB, Hartmann O. La tumeur d'Ewing. Arch Pediatr. 2005; 12: 1383-91.
2. Manduch M, Dexter D, Ellis P, Reid K, Isotalo P. Extraskeletal Ewing's sarcoma/primitive neuroectodermal tumor of the posterior mediastinum with t(11;22)(q24;q12). Tumori 2008; 6:888-91.
3. Rose JS, Hermann G, Mendelson DS, Ambinder EP. Extra-skeletal Ewing sarcoma with computed tomography correlation. Skeletal Radiol. 1983;9(4):234-7.
4. El Weshi A, Allam A, Ajarim D, Al Dayel F, Pant R,Bazarbashi S. Extraskeletal Ewing´s sarcoma family of tumours in adults: analysis of 57 patients from a single institution. Clinical Oncology. 2010;22(5):374-381.
5. Kuzucu A, Erkal HS, Soysal O, Serin M. Extraskeletal Ewing's Sarcoma Presenting with Multifocal Intrathoracic Mass Lesions Associated with Mediastinal Shift. Ann Thorac Surg. 2006; 81:1487-8.
6. Ahmad R, Mayol BR, Davis M, Rougraff BT: Extraskeletal. Ewing's sarcoma. Cancer. 1999;85:725-31.
7. Penal N, Lartigau E, Fournier C, Vilain MO, Dansin E, Taieb S. Sarcomes des tissus mous thoraciques de l'adulte: étude rétrospective de 40 cas. Ann Chir. 2003;128:237-45.