

## Ewing's Sarcoma of the Scapula in an Infant: About a Case

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### Abstract

### Case Report

Ewing sarcoma (ES)/primary neuroectodermal tumor (PNET) is the second most common malignant bone tumor of childhood and adolescence. It can occur at central or peripheral sites, at skeletal or extraskeletal sites, and is more common at the femurs, ilium, and tibia. We report a rare case of Ewing's sarcoma of the scapula in a 2-year-old infant diagnosed with pain and swelling of the left shoulder. Histopathological confirmation of Ewing's sarcoma was possible following ultrasound-guided biopsy. Treatment consisted of initial chemotherapy, followed by surgical tumor resection after good initial response, followed by adjuvant therapy. The current post-treatment follow-up is 2 years without incident. This case is reported to highlight the rarity of localization and therapeutic modalities, particularly surgical.

**Keywords:** Ewing's sarcoma (ES), Scapula, Scapulectomy, child.

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## INTRODUCTION

Ewing sarcoma (ES) is the second most common bone tumor in childhood and adolescence [1]. It usually occurs in the metaphysis, the diaphysis of the long bones of the extremities [2] or flat bones especially the pelvis. The lungs, bones, and bone marrow are the most common sites of metastasis [2]. The prognosis of ES has improved in recent years, thanks to multidisciplinary management involving surgery, radiotherapy, and chemotherapy as multimodal therapies [1]. The scapular localization of the ES is exceptional [3], we present a case of left scapula ES in a 2-year-old infant and discuss the relevant literature on this subject. To our knowledge, fewer than 20 cases of scapula ES have been reported in the literature so far [3-5].

## CASE REPORT

HY, 2 years old, is a male infant and only child of his parents, who has consulted in our facility following the progressive appearance of a swelling of the left shoulder that has been evolving for two months. The swelling is accompanied by mild pain that worsens during the night, with episodes of fever resolved by the antipyretic intake. The medical and surgical history is unremarkable, including no history of loss of appetite, recent weight loss or previous scapular injuries, there are no similar cases in the family. Parents are not inbred.

Clinical examination revealed swelling at the level of the left scapula, supraspinal non-inflammatory

of 5cm long axis, tenderness above the scapular area and an obvious restriction of movement of the left shoulder joint (Fig 1), a limitation of abduction to 90° and external rotation abolished. Absence of mucocutaneous pallor for the rest of the clinical examination without particular.

The radiological assessment included: A standard X-ray of difficult interpretation suggesting a lytic left scapular lesion. An ultrasound objectifying a tumor process with signs of aggressiveness due to muscle invasion and bone irregularity. Magnetic resonance imaging (MRI) was completed showing a lesional process of the left shoulder centered on the scapula, iso-intense in T1 with areas of discrete hypersignal, hyper-intense in T2, moderately enhanced after injection of paramagnetic contrast medium in a relatively homogeneous manner and measuring (50\*52mm) transverse axes over a height of (62mm). Process responsible for bone lysis of the scapula with periosteal reaction, invasion of the soft tissue and myosseous detachment of the infraspinatus which is laminated. Absence of extension to the supraspinatus, deltoid and trapezius muscles. Evoking a PNET in the first place (Fig 2). An ultrasound-guided biopsy was performed to diagnose ES on the morphological and immunohistochemical appearance of a round-cell tumor. 18-FDG POSITRON EMISSION TOMOGRAPHY (PET-CT) showed a hypermetabolic process (SU Vmax: 5.65) centered on the left scapula with scapular lysis in situ and extension to adjacent parts without other metastasis sites (Fig 3). After multidisciplinary

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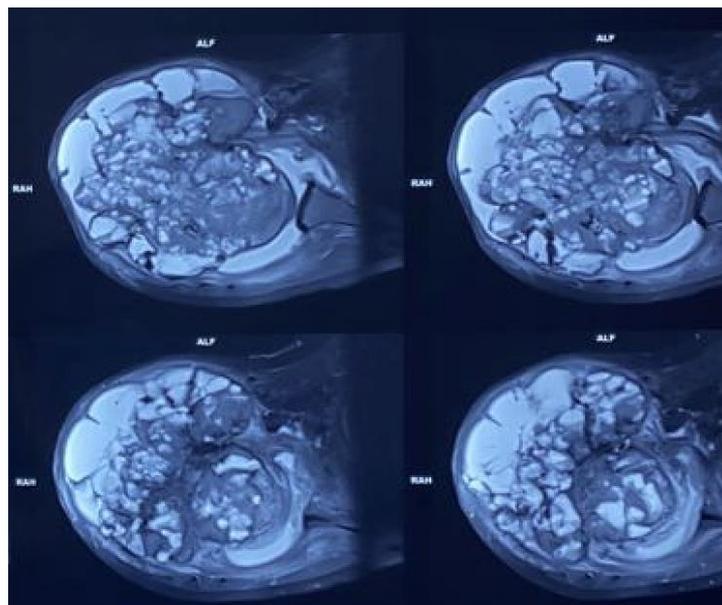
consultation, the patient started chemotherapy according to the Euro Ewing 99-version 8/2003 protocol. Regular clinical and radiological evaluation was undertaken. Follow-up imaging after the 6th course of VIDE showed a reduction in the lesional process centered on the left scapula at 48\*10 mm versus 51\*16 mm which was judged to be resectable in the Medico-surgical staff, the indication for a total left scapulectomy thus placed and then performed. According to the following surgical procedure:

The patient is under general anesthesia, intubated, ventilated, in the right lateral recumbency, T-shaped approach to the spine of the scapula and the inner edge of the scapula, subcutaneous dissection and lifting of the fasciocutaneous flaps, Scapulectomy begins at the inner edge by disinserting the trapezius, rhomboid, angular muscle of the serratus major with respect for their vascularization and then that of the latissimus dorsi. At the outer edge, section of the deltoid muscle outside the long portion of the triceps muscle, the major and

minor round muscles inside, identification and control of the posterior circumflex pedicle of the axillary artery, the left subclavian artery and the brachial plexus which are placed under the lake allowing their control throughout the operation, Dissection, then rotator cuff section, Disinsertion of the long portion of the biceps and the pectoralis minor, coracobrachial and short portion of the biceps at the level of the coracoid process. Cleidohumeral, acromioclavicular and coraco acromioclavicular disarticulation with section behind the omohyoid muscle Verification of the integrity of the noble structures of the left arm 2nd step: repair by suspension of the humeral head from the clavicle by a suspension ring with an external hook, Reinforcement by neighborhood muscles and suspension of the biceps inwards, Closure plane by plane on Redon drain, dressing, Dujarier bandage elbow to body. The aftermath is simple, removal of Redon on the 3rd day dressing on the 15th day, Chemotherapy resumed on day 21 without incident. The child recovered partial mobility in his arm, with a follow-up of 2 years after chemotherapy (Fig 4).



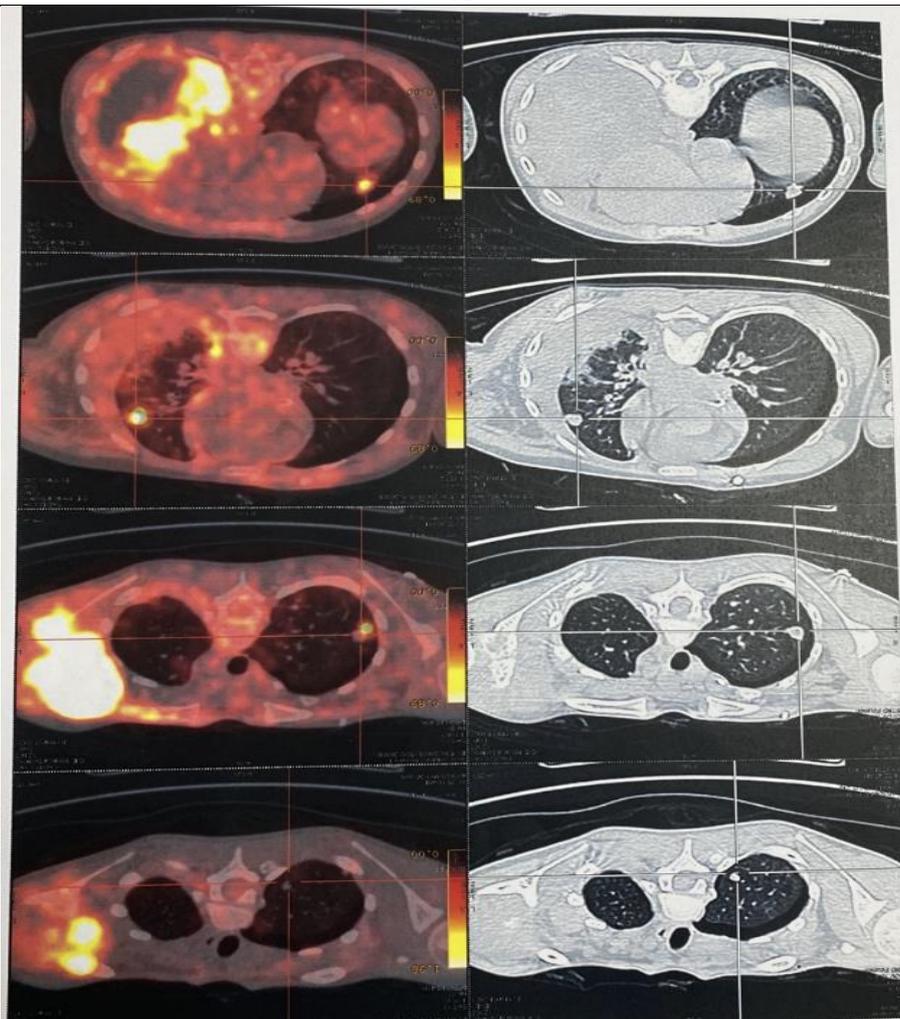
**Fig 1: Inspection during the clinical examination of the patient shows a large swelling in the left scapular region**



**Fig 2: MRI performed in the three planes of space in T1 and T2 weighted sequences, without and with fat saturation, supplemented by an injection of paramagnetic contrast agent, supplemented by CT sections (shows a lytic tumor process centered on the scapula with extension to adjacent soft tissues suggestive of PNET)**



A



B

**Fig 3: A, B PET-CT shows left scapular hypermetabolism related to the primary tumor, absence of suspicious metabolic abnormality on the rest of the volume**



**Fig 4: Follow-up consultation of a 2- year postoperative**

## DISCUSSION

Ewing's sarcoma is a highly malignant tumor composed of small, round cells that primarily affects the skeletal system [1]. It is a neoplasm that mainly affects patients during the first two decades with a male predilection [4]. Similarly, the index case was a 2-year-old male child. It frequently sits in the pelvis, extremities such as the femur, tibia, humerus, and fibula, unlike in our case where it involved the scapula [4-6]. The scapula is a rare site for bone tumors (3%), and most scapular tumors are malignant [7]. The most common tumors affecting the scapula are chondrosarcoma and osteosarcoma. In a cohort study of patients diagnosed with ES between 1988 and 2018, only 29 cases were involved in the scapula [7].

The most common clinical symptoms are pain and a lump in the affected area. Fever, anemia, leukocytosis and increased erythrocyte sedimentation rate [8] are often observed, in our case the patient's laboratory work-up did not present anemia during admission and at the start of chemotherapy. X-rays usually reveal a lesion eaten by moths, as well as a subperiosteal reaction giving an appearance of onion skin [9, 11]. CT scans usually show the extent of bone destruction, and when it comes to detecting soft tissue extent and bone marrow involvement, MRI can be done. The examination should include a chest X-ray, chest CT scan, bone scan, and either a bone marrow biopsy or a positron emission computed tomography (PET-CT) scan [12]. The ES is often grey with necrotic and hemorrhagic areas. Surgeons may interpret the necrotic semi-fluid tissue of the lesion as pus [9].

Systematic therapy and local therapy are important for the treatment of ES. Heavy intraoperative systemic chemotherapy, including cyclophosphamide, adriamycin, vincristine, actinomycin, ifosfamide, and etoposide, is the primary treatment for ES [11].

However, radiotherapy as a local treatment can replace surgery in cases of surgical excision that is considered difficult or incomplete [12]. Surgical resection of the opular girdle is divided into six types according to Malawer. Partial scapulectomy (Malawer resection type II) preserves some of the rotator cuff muscles, including the infraspinatus, subscapularis as well as the anterior serratus muscle, so it has better results on shoulder function. Although the abduction movement may be particularly affected in total scapulectomy (type IIIA or B), the procedure still has a satisfactory level of function [13]. In our case, the patient underwent a total scapulectomy.

Surgery with neoadjuvant chemotherapy is considered better for overall survival in ES of the scapula compared to chemotherapy or radiotherapy alone. Similarly, our patient received induction chemotherapy followed by surgery. On the other hand, patients who underwent marginal resection and a chemotherapy response of <100% had the worst outcome [7]. Local recurrence did not have a significant effect on overall survival [7]. Long-term follow-up should be performed after primary management to detect any secondary malignancies and growth-related musculoskeletal complications. The Musculoskeletal Tumor Society (MSTS) score was used to assess scapula function, and according to Malik *et al.*, the median MSTS score was 67.4% [7].

The prognosis of ES depends on many factors: anatomical location, tumor stage and size, necrosis next to chemotherapy, presence or absence of metastases, elevated serum LDH levels, and older age [14]. Psychological counselling may be seriously needed after operations, especially in sarcoma patients, due to the psychological effects and depression [14], in the older subject.

## CONCLUSION

The scapula is a rare site for bone tumors, and the most common tumors affecting the scapula are chondrosarcoma and osteosarcoma. Ewing's sarcoma is extremely rare in the scapula and should be considered a differential diagnosis for any patient with inflammation above the scapular region. Surgical resection, multi-agent chemotherapy and radiotherapy are the mainstay of treatment for this rarity.

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