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Recurrent Extraskeletal Ewing Sarcoma of the Upper Limb: A Surgical Case Report

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1. Abstract

Extraskeletal Ewing sarcoma (EES) is a rare, aggressive soft tissue tumor. We present the case of an 11-year-old girl with a recurrent EES of the right upper arm. The lesion required extensive surgical resection boosted with intraoperative radiotherapy, and reconstruction utilizing a pedicled latissimus dorsi flap. Approximately one year after, the patient experienced a humeral fracture that failed to heal over time, ultimately progressing to a pseudoarthrosis. Definitive management required reconstruction through a vascularized fibular graft. Multidisciplinary planning was central to achieving long-term disease control and functional recovery. Nearly a decade post-recurrence, the patient remains relapse-free with preserved arm mobility. This case underscores the complexity of EES management and highlights the importance of tailored, team-based approaches in pediatric oncologic surgery and reconstruction.

2. Introduction

Extraskeletal Ewing sarcoma (EES) is a rare soft tissue malignancy, representing 20–30% of Ewing sarcoma family tumors (ESFTs). With an incidence of 0.4 cases per million, EES mainly affects children under 5 and adults over 30. The extremities are the most involved anatomical sites [1]. ESFTs are characterized by recurrent chromosomal translocations, most often t(11;22), producing the EWS-FLI1 fusion protein. Histologically, they show a uniform population of small round cells with a high nuclear-to-cytoplasmic ratio.

EES management requires a multidisciplinary approach, involving typically a combination of chemotherapy, surgical resection, and radiotherapy. The 5-year survival ranges around 60–70% in localized cases, but falls below 30% in metastatic disease, which occurs in approximately 25% of patients [3].

This case highlights the surgical management of pediatric EES, focusing on decision-making, operative strategy, and perioperative care within a complex clinical setting. A written informed consent for publication of the surgical case has been obtained.

3. Case Report

We report the case of an 11-year-old girl with a recurrence of EES. The patient, already known for her congenital cardiopathy, was diagnosed at age of 5, with an EES of the right upper arm with secondary involvement of the humerus. The tumor was treated with definitive chemoradiotherapy.

The recurrence presented as a progressively enlarging mass over the past three months in the same area. The girl reported no symptoms except mild discomfort only on deep palpation.

An MRI revealed a 4.2×4.7×7.9 cm heterogeneous soft tissue mass occupying the anterior compartment of the arm, infiltrating the biceps brachii, the distal portion of the deltoid, and surrounding subcutaneous tissues, including encasement of the cephalic vein. The mass displaced but did not invade the neurovascular bundle (median and ulnar nerves, brachial vessels). No metastatic spread was found. Histopathological examination confirmed EES recurrence.

Surgical approach comprised wide resection of the soft tissue of the anterior portion of the limb (Figure 1), including the affected muscles, the cephalic vein and the musculocutaneous nerve. After the excision, intraoperative radiotherapy was performed. A latissimus dorsi pedicled my cutaneous flap, harvested (Figure 2) and tunneled through the axilla, was anchored proximally to the coracoid process and distally to the biceps tendon and served

to cover the soft tissue defect (Figure 3). Neoadjuvant and adjuvant chemotherapy supported the treatment. About one year after surgery, the patient developed a pathological fracture of the humeral diaphysis. Despite its management, over follow-up the fracture failed to show radiographic and clinical signs of consolidation and progressed to a state of pseudoarthrosis. Ultimately, a reconstructive surgical approach was planned.

Following the optimization of the recipient bed with debridement of the humeral stumps (Figure 4), a 5-cm vascularized osteocutaneous fibular graft was harvested (Figure 5) and inserted at the level of the defect (Figure 6), with periosteum sutured circumferentially at both graft-host interfaces. Definitive fixation was achieved using a Pedifrag locking plate system, in conjunction with the external fixator. Microvascular reconstruction involved an end-to-side anastomosis of the peroneal artery to the brachial artery and end-to-end anastomosis of the peroneal vein to the brachial vein. Surgical follow-up was uneventful and with optimal results (Figure 7).

Currently, nearly 10 years after the recurrence of EES, there has been no further relapse, and arm mobility is well preserved. Now an adult, the patient is living her life to the fullest while awaiting heart transplantation.

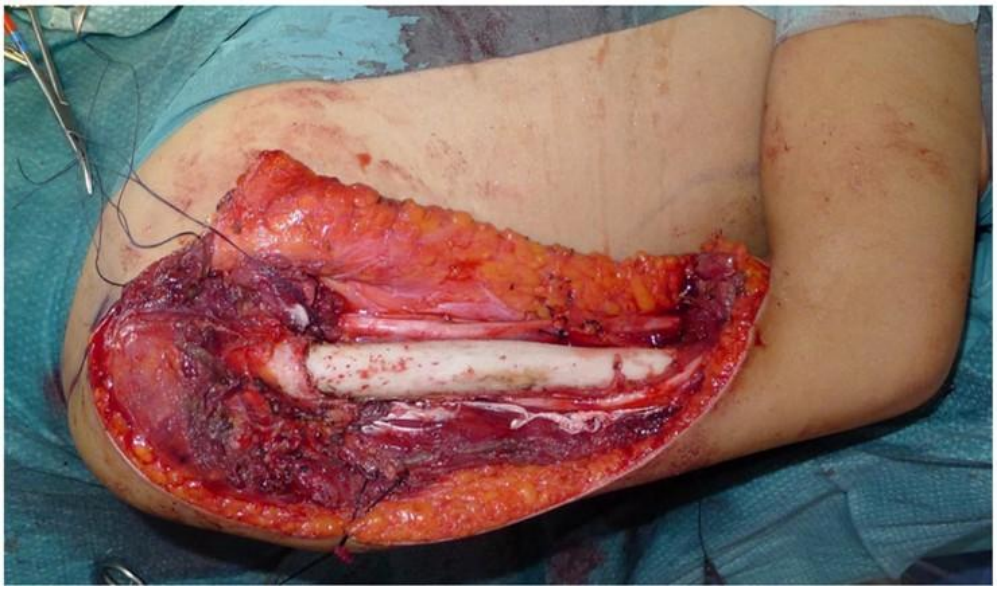

Figure 1: Intraoperative view after the wide resection of the anterior compartment of the right arm.

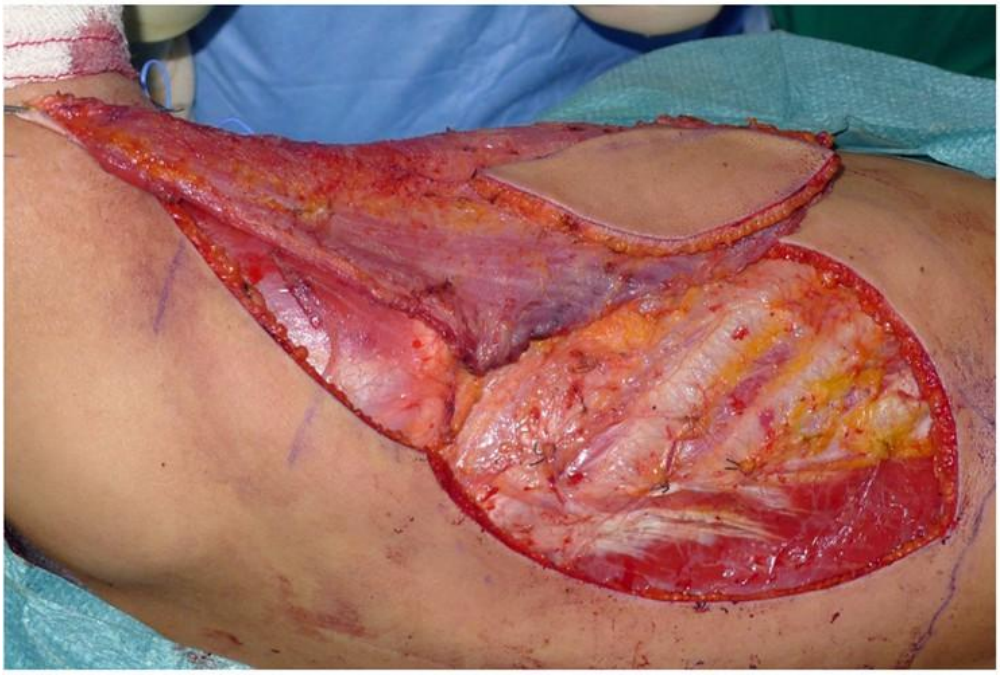

Figure 2: The latissimus dorsi flap was harvested and mobilized on a narrow vascular pedicle.

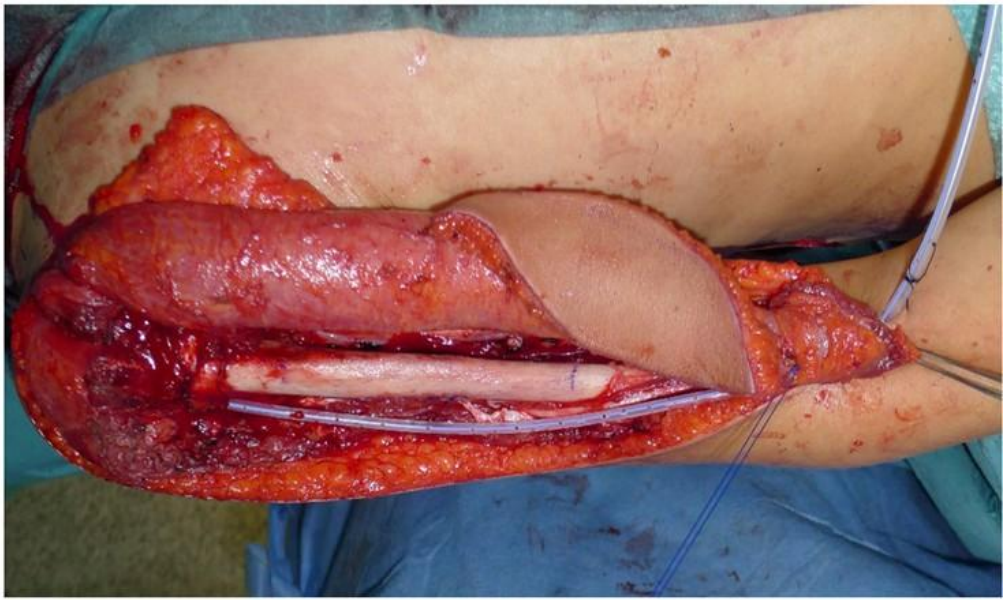

Figure 3: The flap was sutured onto itself to restore the volume of the resected anterior compartment of the arm and anchored proximally to the coracoid process and distally to the biceps tendon.

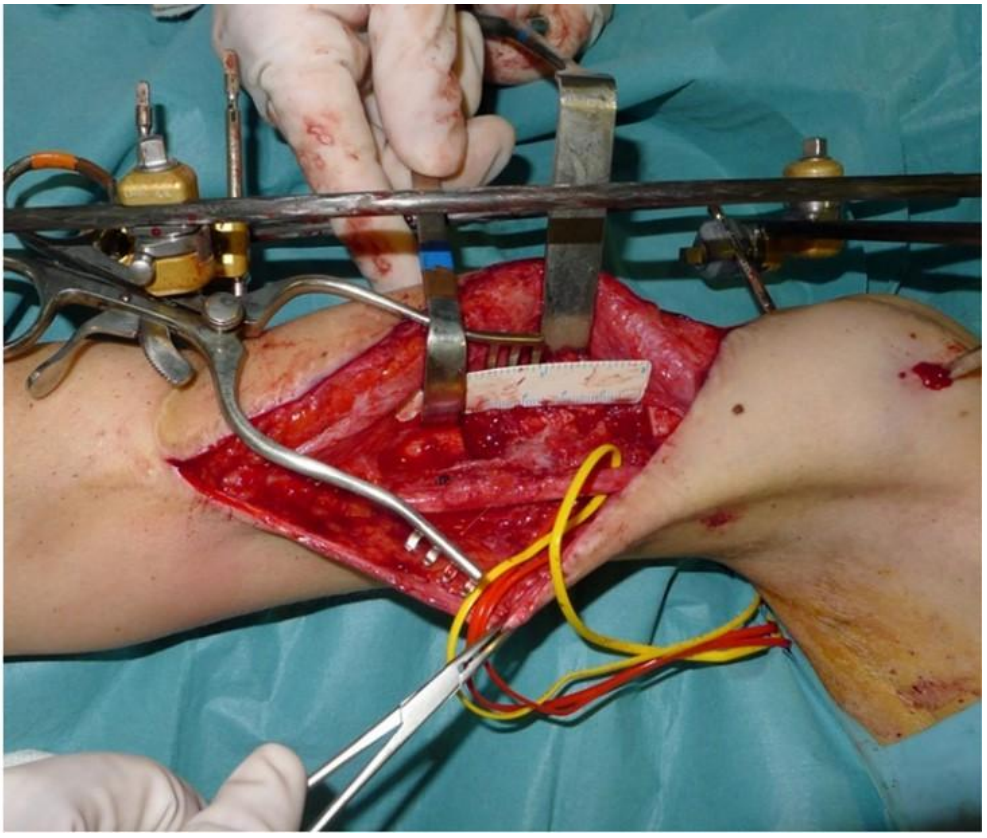

Figure 4 - 5^{cm}: Pseudoarthrotic gap in the humeral diaphysis.

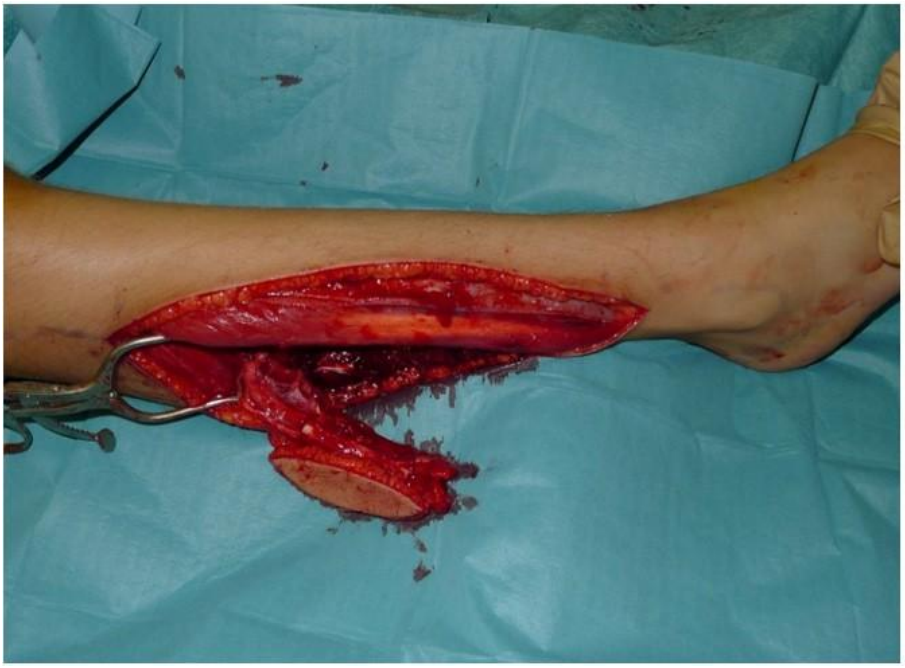

Figure 5: Harvesting of the vascularized osteocutaneous fibular graft.

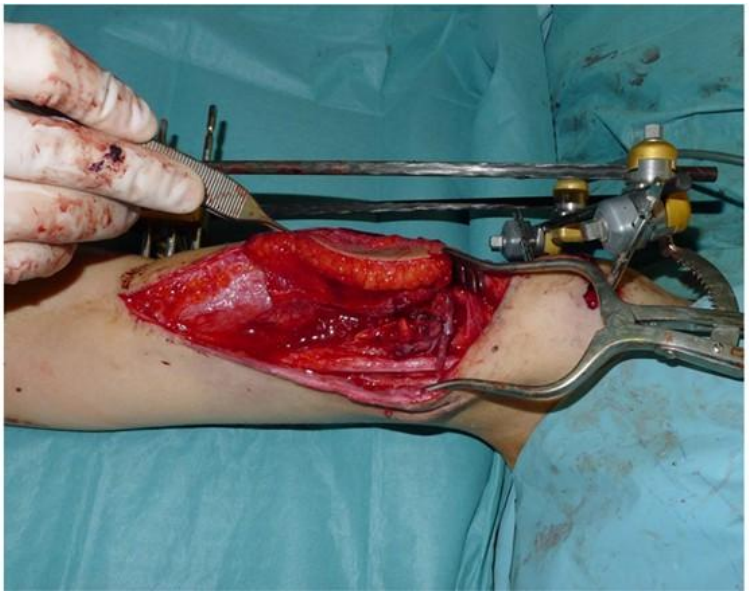

Figure 6: Placement of the free fibular graft into the humeral defect.

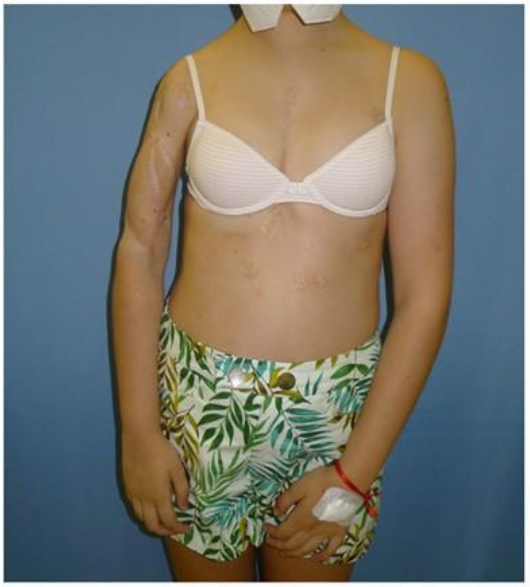

Figure 7: 18 months after free fibular graft intervention.

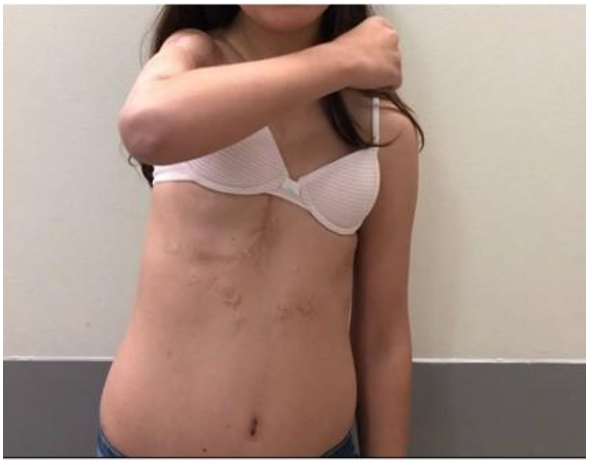

Figure 8: Almost four years after functional reconstruction with a pedicled latissimus dorsi flap, and two years after free fibular grafting, elbow flexion is completely preserved.

4. Discussion

Systemic chemotherapy is a key component in the treatment of Ewing sarcomas, often administered prior to surgery or radiotherapy to shrink the tumor mass and address potential micro metastases. Standard regimens, such as VIDE (vincristine, ifosfamide, doxorubicin, and etoposide), are commonly employed. Individuals with EES are more frequently managed with a combination of chemotherapy and surgical excision alone, reserving radiotherapy for situations involving positive surgical margins or when the tumour is deemed inoperable [4]. In our clinical case, at the first diagnosis of EES, a multidisciplinary team opted to pursue definitive chemoradiotherapy. This strategy was chosen due to the presence of secondary humeral involvement, as radical surgical resection would have required highly mutilating procedures with significant potential functional impairment.

Upon the recurrence, the team, decided on a course of neoadjuvant chemotherapy followed by surgical intervention with intraoperative radiotherapy, aiming at reducing the risk of pathological fractures [5]. In the adult population, the latissimus dorsi flap has consistently proven to be a reliable and versatile option for upper limb reconstruction following oncological resection, especially when both soft tissue coverage and restoration of elbow or shoulder function are required [6]. Conversely, reports concerning paediatric applications remain limited [7, 8]. In our patient, the choice of this flap not only facilitated a relatively swift postoperative course but also enabled meaningful functional recovery of arm mobility (Figure 8). As feared, a pathological fracture eventually occurred and failed to heal. This prompted the tumour board to recommend reconstruction of the pseudoarthrosis segment of the humerus using a free vascularized fibular flap. This flap was first described in 1998 for proximal humeral reconstruction in paediatric patients [9] and has since been employed with favourable outcomes demonstrating a key role in functional and biologically active bone reconstruction [7, 10]. As previously mentioned, every therapeutic decision regarding the patient was made within a multidisciplinary team composed of oncologists, paediatric surgeons, plastic surgeons, and orthopaedic surgeons. We believe this was fundamental to the successful outcome of the patient in the long battle against the tumour.

6. Conclusion

Extraosseous Ewing sarcoma is a rare tumor where integration and coordination of surgery, radiotherapy and systemic treatments play a central role. Despite advances, especially in microsurgery, post-oncologic reconstruction in children remains challenging. With them an individualized treatment plan is imperative, considering rapid recovery to enable timely potential adjuvant therapy, minimizing donor-site morbidity, and ongoing somatic growth. In this context, sharing experiences from complex clinical cases contributes to improving therapeutic approaches for rare tumors such as EES.

7. Author Contributions

We, the undersigned authors, hereby certify that we have participated sufficiently in the work to take public responsibility for appropriate portions of the content. Each author certifies that their contribution is as stated below.

• Antonio Massimo Spedale: Manuscript writing, Data Collection, literature review, submission

• Felicia Geanina Grosu: Manuscript writing, Data Collection, literature review

• Manuel De La Torre: Surgical case management, study design, methodology

• Beatriz Berenguer: Surgical case management, supervision, final approval

• Concepcion Lorca Garcia: Surgical case management, supervision, final approval

All authors have read and approved the final version of the manuscript and agree to be accountable for all aspects of the work.

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