Oxford City Publishers

Case Reports in Orthopaedic Surgery Journal

Case Report | Vol 3 Iss 4

Free Vascularized Fibular Graft: The Solution for a Return to Elite Volleyball After Humeral Ewing's Sarcoma

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Received: September 25, 2024; **Accepted:** October 13, 2024; **Published:** November 15, 2024

Abstract

Case: A 13-year-old girl, volleyball player, presented with right arm pain after minor trauma. She was diagnosed with Ewing's Sarcoma of the proximal humeral shaft. An en bloc resection and reconstruction were performed using a free vascularized fibular graft and carbon-fiber plate. She returned to sports activity at 12-months postoperative, achieving complete functional recovery. At 5-years follow-up, she is a junior volleyball athlete, with no signs of tumor recurrence.

Conclusion: Free vascularized fibular autograft is a viable option to reconstruct large bone defects after tumor resection of the upper extremity in young athletes. Complete return-to-practice may be achieved, even in high-demand sports.

Keywords: Ewing's Sarcoma; Humerus sarcoma; En bloc resection; Free vascularized fibular graft; Carbon-fiber plate

Introduction

Ewing's sarcoma is a malignant tumor that mainly affects children and young adults [1,2]. The most important prognostic indicator is the presence of metastases at the time of presentation, which happens in 25% of patients [3]. Survival rate is approximately 60% in 10 years for localized disease and 30% for metastatic disease [2].

The current standard treatment of Ewing's Sarcoma is a combination of multi-agent chemotherapy and local surgical control (which may be combined with radiotherapy). This should be managed by a multidisciplinary team. The local approach to the tumor can be done with limb salvage procedures or, if not feasible, amputation [1,4].

For limb sparing procedures, there are multiple options for reconstruction after tumor resection: endoprosthetic replacement; vascularized bone graft; nonvascularized bone grafts (autograft or allograft); or a combination of both. The choice between these reconstruction techniques depends mainly on the anatomical location of the tumor and patient's age [5,6].

In this article we report the use of a vascularized free fibular autograft for humeral reconstruction following an en bloc resection of a diaphyseal Ewing's sarcoma.

Case Presentation

A healthy 13-year-old girl, volleyball player, presented with right arm pain after minor trauma. She described a six-month history of right arm discomfort, mainly after intense physical exertion and that was alleviated by ibuprofen. On physical examination, she had tenderness and swelling on the middle third of the arm, with no palpable mass. The neurologic examination of the right upper extremity was normal. Plain radiographs showed a poorly defined permeative lesion, with cortical erosions, involving the right proximal humeral shaft (Figure 1). Magnetic resonance imaging (MRI) showed a heterogenous lesion in diaphysis, measuring 134 x 18 mm, with periosteal reaction and cortical disruption and a soft tissue component of 67 x 13 mm (Figure 2). An Orthopedic Oncology surgeon performed a core needle biopsy, with extemporaneous analysis. Histological examination showed small round blue cells, mostly apoptotic, with areas of necrosis. Molecular analysis demonstrated an EWSR1 rearrangement, confirming the diagnosis of Ewing's sarcoma. Whole body computed tomography (CT) was negative for metastatic dissemination. The patient received neo-adjuvant chemotherapy comprising of seven cycles of VIDE (vincristine, ifosfamide, doxorubicin and etoposide).

Preoperative Planning and Surgical Technique

After completing neoadjuvant chemotherapy, MRI demonstrated a similar intramedullary extension but a reduction of Gadollinium enhancement and a complete soft tissue mass involution. So, the tumor board deemed the patient to be a suitable candidate for limb salvage procedure. Preoperative planning determined an en bloc excision of 14 cm, sparing all major neurovascular structures and the proximal humeral physis, with primary reconstruction using free vascularized fibular autograft (Figure 3), fixed with a radiolucent plate.

The surgical intervention was performed in a semi-beach chair position. An extended deltopectoral approach to the right humerus was used, isolating the biopsy tract (Figure 4). The deltoid, pectoralis major and latissimus dorsi were released. The proximal bone cut was confirmed with a needle and fluoroscopy images (Figure 5). Distal osteotomy was made 14cm distal, using a ruler to mark the distal end. The resected specimen was sent to anatomopathological analysis (Figure 6). The contralateral fibula was simultaneously harvested by a Plastic Microsurgeon for an intercalary vascularized bone graft. (Figure 7) The proximal and distal ends of the graft were burred down into the medullary canal on either end of the remaining humeral segments. Microvascular anastomoses were performed between the peroneal artery and vein and deep branches of the brachial artery and vein (Figure 8). Deltoid, pectoralis major and latissimus dorsi were reattached through transosseous tunnels (Figure 9). Fixation was achieved with a long carbon-fiber proximal humerus plate. Drains were placed and the wounds closed in layers (Figure 10). Final pathology confirmed Ewing Sarcoma with negative margins (minimum 7 mm) and complete response to chemotherapy. Staging by TNM system: ypT0 Nx R0.

Postoperative plain radiographs were obtained prior to discharge (Figure 11). The patient remained with a arm sling for 4 weeks and weight bearing as tolerated on the left lower extremity in a walker boot. She received 7 cycles of adjuvant chemotherapy, consisting of one cycle of IVA (ifosfamide, vincristine and actinomycin D) and six cycles of VAC (vincristine, actinomycin D and cyclophosphamide). There were no neurovascular impairment or wound complication during follow up. Radiographic control after five months showed bone union and complete incorporation of the graft, without hardware complications (Figure 12). After complete recovery of shoulder and elbow range of motion, the patient was allowed to return to low-impact sports activities after 12 months postoperative and to volleyball 6 months later. She maintained regular follow-up with plain radiographs (Figure 12), CT and/or MRI, to evaluate graft hypertrophy and exclude suspicious lesions. At 5 years postoperatively, there is no evidence of tumor recurrence, and she is a junior volleyball athlete, without pain or functional limitations and without upper limb discrepancy.

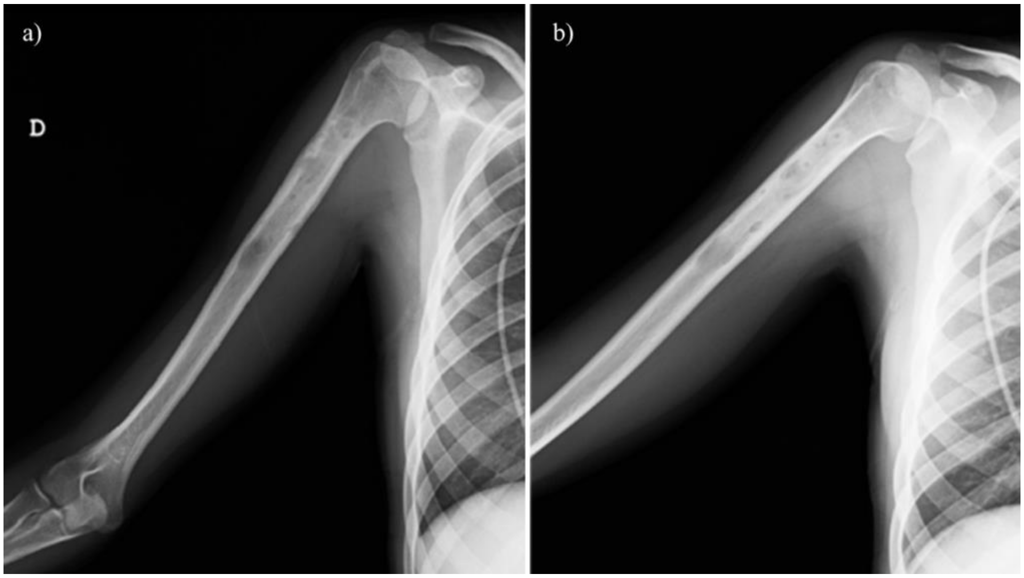

Figure 1: Right arm plain radiographs. Legend: a) anteroposterior view; b) side view.

Figure 2: Coronal view of T2-weighted magnetic resonance image.

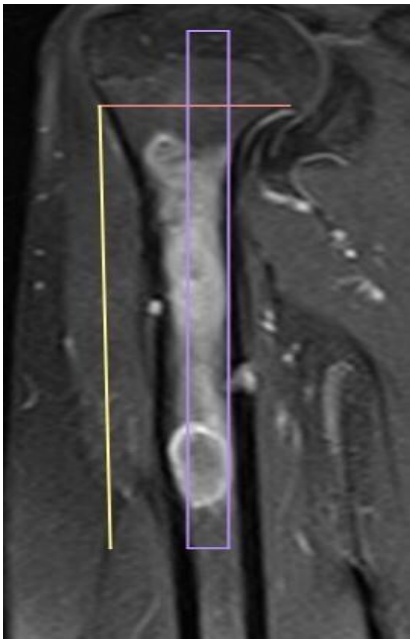

Figure 3: Preoperative planning, with representation of humeral en bloc resection distal to proximal physis (red and yellow lines), and reconstruction with fibular graft (purple rectangle).

Figure 4: Extended deltopectoral approach, isolating the biopsy tract.

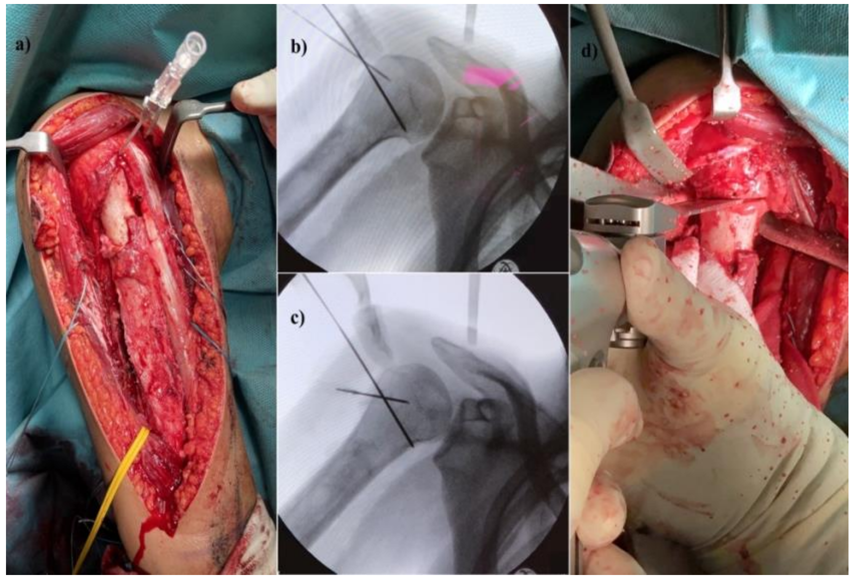

Figure 5: Steps of proximal humeral osteotomy. Legend: a) inserting a needle to mark the proximal osteotomy cut; b) and c) fluoroscopy images to confirm the needle was distal to proximal physis; d) proximal osteotomy cut.

Figure 6: Resected specimen after en bloc resection, with "P" marking the proximal end.

Figure 7: Harvesting vascularized left fibula autograft. Legend: a), b) and c) lateral approach elevating peroneal muscles and performing a 360º subperiosteal dissection; d) free-vascularized graft, showing the vascular pedicle.

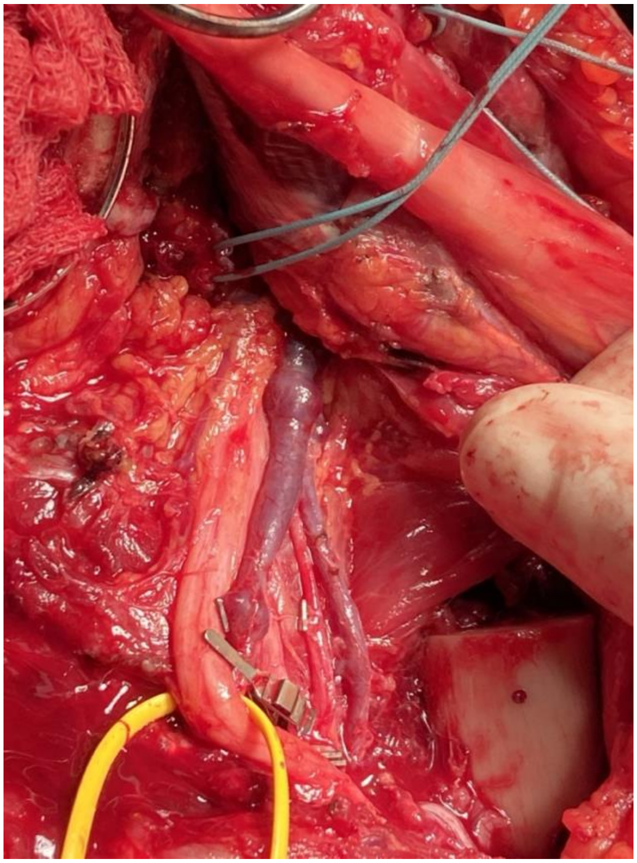

Figure 8: Deep branches of the brachial artery and vein, prior to microvascular anastomoses.

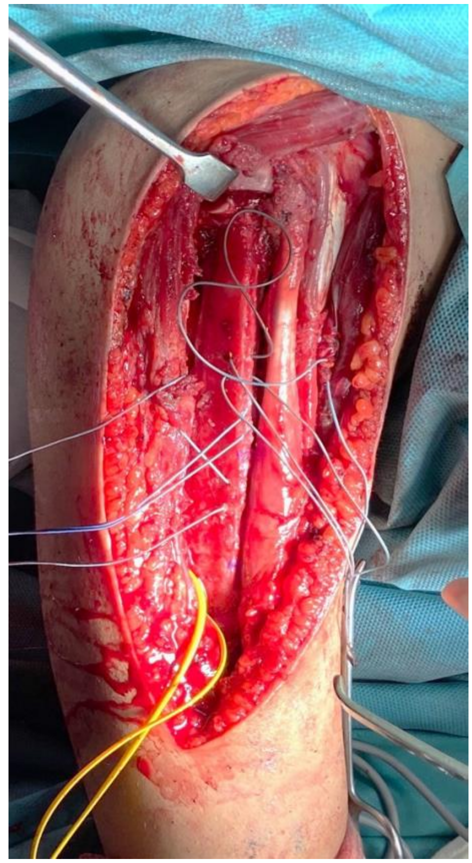

Figure 9: Transosseous tunnels to deltoid, pectoralis major and latissimus dorsi reattachment.

Figure 10: Wound closure after drain placement.

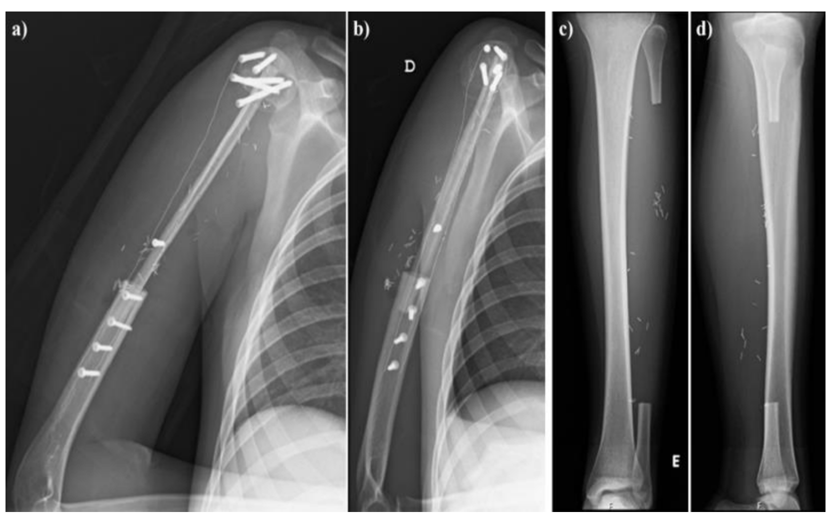

Figure 11: Postoperative plain radiographs. Legend: a) anteroposterior view of right arm; b) side view of right arm; c) anteroposterior view of left leg; d) side view of left leg.

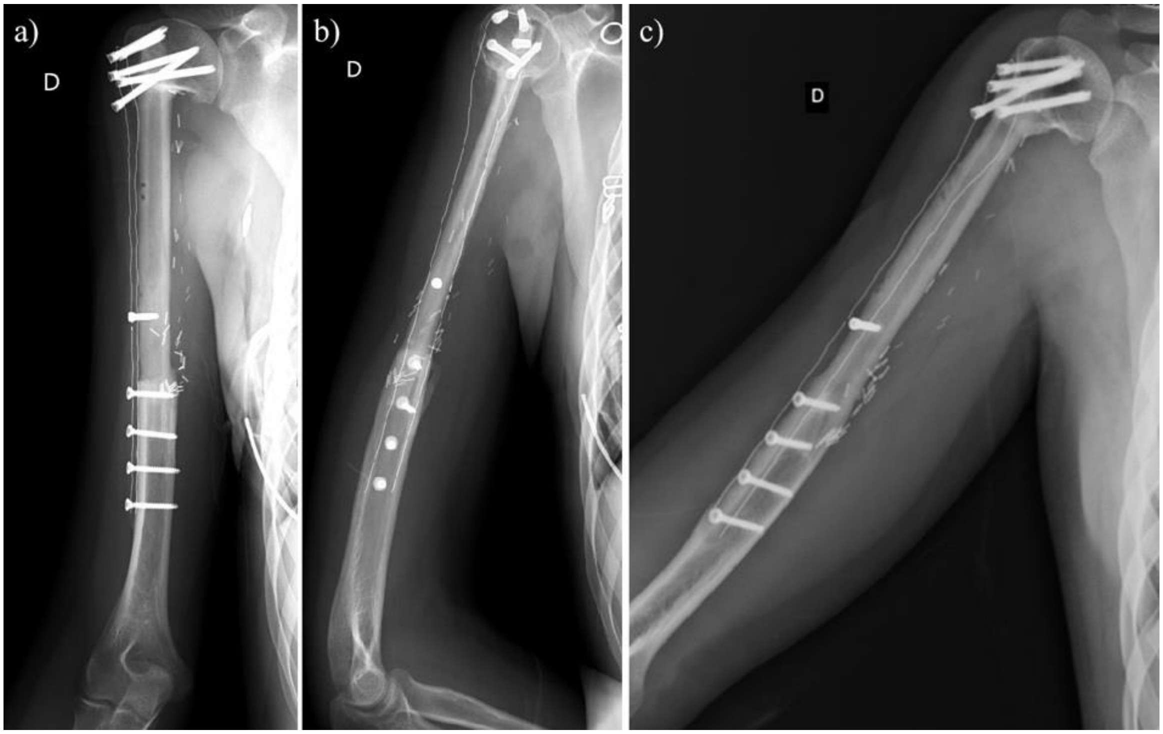

Figure 12: Right arm plain radiographs during follow-up. Legend: a) anteroposterior view 5 months postoperative; b) side view 5 months postoperative; c) 3 years postoperative.

Discussion

Ewing's Sarcoma is the second most common primary malignant bone tumor in pediatric population [1,2]. The majority of patients have nonspecific clinical features, like local pain (often mild) and swelling. So, as seen in this case, symptoms may be mistaken for injuries resulting from sports or other activities and diagnosis, delaying the diagnosis for 3-9 months [7]. Fortunately, prognosis does not seem to be associated with time to diagnosis [8]. It is mostly dependent on the presence of metastases at the time of presentation, tumor size and location, and histological response to chemotherapy [3]. Considering these prognostic factors, our patient had localized disease, in an non-axial location and a complete histological response after neoadjuvant therapy, which are favorable factors. However, the tumor was more than 10cm long, which is associated with poorer survival [3].

Neoadjuvant chemotherapy has been shown to reduce tumor dimensions, increasing the success of the surgery. After surgery, as adjuvant therapy, is used to eliminate residual cells. This reduces the rate of recurrence and metastasis, which is the most important prognosis factor [3,9]. In this case, the patient received six cycles of chemotherapy prior surgery, with complete histological response, and eight cycles after surgery.

There have been advances in the treatment of extremities sarcomas, allowing limb salvage surgery to replace amputation, without negatively impacting survival [10,11]. After surgical resection, there are multiple reconstructive options: prosthetics, allografts, and autografts. Prosthetics devices allow for earlier rehabilitation and without donor site morbidity, however, they have higher risk of infection and mechanical complications [12,13]. Also, in young patients, it is imperative to discuss the possibility of revision surgery. Allografts permit reconstruction of segmental defects without donor site morbidity but have a higher risk of fracture and infection than autografts, specifically in large bone defects. Vascularized bone grafts is an attractive alternative given their higher rate of union and ability to remodel, resulting in hypertrophy under mechanical stress [6,14]. Free vascularized grafts, although technically challenging, have more versatility [15-17]. The fibula in one of the vascular autografts most frequently used in long bone defects, mostly because of the length of its vascular pedicle and the direct approach to harvest. An additional advantage, which was also considered in our surgical planning, is the similar shape and size comparing with a pediatric humerus [14,15].

The most common complications following free vascularized fibula autograft for upper extremity reconstruction are fracture [18,19] and infection [14,19], none of which was diagnosed in our patient. Long-spanning plate fixation may reduce the risk of fracture rate [17]. In this case, we used a long plate of carbon-fiber material due to its major advantage: radiolucency, which facilitates postoperative surveillance of tumor recurrence [20].

In this case, bony union between the graft and the host bone was achieved after 5 months, similar to the average time described in previous studies [18,19,21,22]. After confirming bone union, the patient had regular imaging controls to assess hypertrophic changes of the graft so we could guide the pace of gradual resumption of sports activities.

Ewing's Sarcoma is a rare disease, and the use of free vascularized fibula grafts is a procedure seldomly performed so, there is lack of information about the long-term effect of this reconstruction technique after tumor resection. This article has a long follow-up period and reports good clinic and radiographic outcome of free vascularized fibula grafts used for reconstruction after humeral sarcoma resection. It shows that it is possible to achieve a good oncologic and functional result, with complete return-to-practice, even for athletes of high demand such as volleyball.

Conclusion

Ewing sarcoma is associated with significant morbidity and mortality in the pediatric population, and wide surgical resection is essential for local control of the disease. This case report highlights the advantages of using a free vascularized fibular autograft in a limb sparing approach of the upper extremity of a volleyball athlete. The oncologic and functional outcomes support the use of this surgical technique to reconstruct a humeral segmental defect of at least 14 cm. In a 5-year follow-up period, there was no tumor recurrence, and she achieved complete return-to-practice.

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