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Case Report

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Extraosseus Ewing´S Sarcoma. Reconstruction after Irradiation Pathological Fracture

Manrique Gamo Elena[1]*; Buendía Pérez Javier[2]; García Maroto Roberto[1]; Cebrián Parra, Juan Luis[1]

[1]Orthopaedic Surgery and Traumatology, Musculoesqueletal Sarcomas Department, Hospital Clínico San Carlos, Madrid, SPAIN.

[2]Plastic Surgery, Musculoesqueletal Sarcomas Department, Hospital Clínico San Carlos, Madrid, SPAIN.

Abstract

Introduction: Ewing's sarcoma (ES) is a highly malignant tumor. Extraesqueletal location is less frequent (25% of all Ewing's sarcomas). The management is limb-preserving surgery if it is possible, but it also means adyuvancy treatment which includes radiotherapy, with the risk of pathological fractures which mean a great challenge for successful management.

Material and methods: We present a case of a 53 years old female patient with a tibial pseudoarthrosis after a pathological fracture in the context of extraosseus ES which received neoadyuvant radiotherapy many years ago. At the beginning she underwent endomedular nailing, with failure of the treatment. The clinic symptoms were pain and progressive deformity.

After subsequent studies she was diagnosed of aseptic pseudoarthrosis.

The solution for this case was a carefully surgery planification with the performing of an osteocutaneous fibula flap from healthy leg after previous osteosinthesis removal.

Discussion: She had a good outcome. At this moment, fourteen months after surgery, plain X-Rays show bony fusion and no implant failures. but she uses one crutch in her right hand.

Conclusion: Other options exist for the reconstruction, but in our opinion, vascularized fibular grafts are useful in the reconstruction of large skeletal defects, especially in cases of scarred and avascular recipient. Even though, this graft provides mechanical strength, predictable vascular pedicle, and hypertrophy potential.

Keywords: Extraosseus ewing sarcoma; Pathological fracture; Postradiotherapy; Osteocutaneous fibular flap; Lower limb.

Abbreviations: ES: Ewing's sarcoma; PNET: Primitive Neuroectodermal Tumor.

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Correspondance: *Elena Manrique Gamo, Orthopaedic Surgery and Traumatology, Musculoesqueletal Sarcomas Department, Hospital Clínico San Carlos, Madrid, SPAIN. Tel: +34670040423; Email: manrique.elena@gmail.com*

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Introduction

Ewing's Sarcoma (ES) is a highly malignant tumor which affects overall children and teenagers. It's a representative of the so-called round cell tumors. It's origin is not clear, but it seems to be originated from bone marrow cells, or a sarcoma neutrally derived small round cell malignance, very similar to the primitive neuroectodermal tumor (PNET). Most of the cases occur before 25 years old (90%), and it is extremely rare in black persons.

It typically arises from the medullary cavity with invasion of the Haversian system and has been described in every bone in the body, most of all, the diafisis of long bones.

Extraskeletal locations of Ewing sarcoma are less frequent (25%), and include the lower extremities (32%), the paravertebral region, (15%), the chest wall (11%), and the retroperitoneum (11%). The imaging features of extraskeletal ES have not been extensively described, and it does not always show the characteristic appearances of other types of ES.

Primary malignant bone tumors account for less than 1% of all cancers.

The evolution in management of sarcomas in the lower extremities has been from amputations to limb-preserving surgeries with evidence to support that they have equal overall survival, but with better functional outcome. The challenge of reconstruction lies in providing a durable, functional, and a esthetically limb [1].

Coordinated multimodal local therapy is critical in ensuring recurrence-free limb sparing surgery. Radiotherapy has been shown to decrease local recurrence rates in limb-salvage procedures [2], but high dose radiation is injurious to bone and is a known risk factor for late fractures, which should be managed with intramedullary nailing.

The incidence of postradioterapy fractures ranges from from 1.2% to 25% with a consolidation rate of 33% to 75%, being more frequent in the ribs, pelvis, and femur. The time between irradiation and fracture, occurs years after radiotherapy. Risk factors include age above 50 years, female gender, extensive periosteal detachment, circumferential irradiation, tumor size, and anterior thigh location. The etiology is still uncertain, but cellular disappearance, reduction of bone turnover and activity were observed hematopoietic as possible causes of failure of consolidation [3].

Pathologic fractures after the bone is exposed to ionizing radiation, although relatively infrequent, have challenging treatment due to the high rate of complications, especially the failure in consolidation and infection, leading to disastrous functional results [4].

The options for the reconstruction of pseudoarthrosis are nonvascularized cancellous and cortical autografts, vascularized bone (auto)graft (VBG), cadaveric bone allografts or a combination thereof, induced-membrane, and bone transport techniques (isolated shortening, compression followed by distraction and segmental bone transport),biological reconstruction with a combination of techniques including autoclaved autograft bone and endoprosthetic reconstruction [5].

VBGs can be useful in large bony defect cases and especially when infection, scarring, and poor vascularity. They are also superior in strength, remodeling, and hypertrophy. Typically, microvascular free fibular VBG is the most suitable in lower limb reconstruction due to its shape, vascular pedicle predictability, and potential for hypertrophy [6]. Bibliography supports that the microvascular free fibular flap has a lower infectious complication rate and higher rate of bone union when compared with the traditional allograft.

Material and methods (Case presentation)

Clinical data

53 years old woman at the present moment. Diagnosis of extraosseus ES (QTNA+) on her right leg in 2002. As surgical therapy she underwent distal fibula and surrounding muscles surgical resection (Figure 1a/1b). After surgery the patient underwent adyuvant quimiotherapy and radiotherapy, with positive outcome. Preoperative and postoperative anatomopathological studies confirmed extraosseus Ewing sarcoma.

During the follow-up, she was free of local and distal recurrence of the local disease.

In August 2018, she suffered a pathological fracture of her right tibia. The treatment was reduction and endomedular nailing. During the follow-up, we found out tibial pseudoarthrosis (Figure 2a/2b). She felt pain, insestability and progressive deformity of her right leg. She had not systemic symptoms. Between the different options for the treatment, we decided that the optimal one was the performance of a vascularized osteocutaneous fibula flap. We did presurgical planning first for this procedure.

Physical examination was routine except for limp and significant pain in her right leg. The range of motion was also between normal limits. Neurologic examination revealed no specific findings. Laboratory analysis revealed parameters in normal ranges, without infections symptoms and signs.

Imaging examinations

Lower limb arteriography (Figure 3a/3b):

- Right lower limb: preserved iliac, femoral, and popliteal sector. Distal outflow through two vessels (anterior and posterior popliteal vessels). The peroneal artery had an occlusion in its proximal third. Pedal artery and plantar arch preserved.
- Left lower limb: The iliac and femoropopliteal sector conserved. Distal outflow through three vessels. Pedal artery and plantar arch preserved.

Plain X-rays of the right tibia showed tibial pseudoarthrosis.

Operative procedures

After preoperative studies which not contraindicate surgery, she underwent surgical intervention under general anesthesia and antibiotic prophylaxis. The surgery was carried out by two surgical teams together, (orthopedic and plastic surgeon), performing:

Through previous incisions, first of all, we removed the endomedular nail from the tibia nail and screws. By right ankle medial approach, we confirmed the absence of consolidation with pathological mobility at the tibial fracture site. We removed fibrotic tissue and reduced the fracture reduced with the implant of LCP 3,5 mm fourteen holes plate.

An osteocutaneous fibula flap rose on the healthy leg (Figure 4a/b). The skin paddle was designed after the location of perforators by handheld doppler. The previous arteriography excluded the presence of a peroneal magna artery.

We performed the standard flap dissection technique under tourniquet. After the flap was utterly elevated, we deflated tourniquet, and we carried out hemostasia.

On the other leg, we carved an intramedullary socket for the inset of the fibula after fixing the plate (Figure 5). Then the flap is ligated and placed in the socket. Fine-tuning of the margins of the bone flap was done to press-fit the fibula in the socket. We fixed the fibula with two screws taking care to avoid any damage to the vascular pedicle. We prepared receptor vessels. We chose the proximal stump of the posterior tibial artery after checking adequate pulsatility of the artery way far the radiated area. We did not choose the anterior tibial artery to preserve foot irrigation. 13 cm interposition vein grafts were needed to reach the receptor vessels and were harvested from the great saphenous vein from the healthy leg. We performed vascular anastomosis were performed in a termino-terminal fashion, and we obtained excellent patency. After the assessment of good perfusion of the flap, we closed the skin directly (Figure 6). We closed the donor leg with a skin graft.

The postoperative period was uneventful, and the patient was discharged (Figure7a/7b).

Postoperative outcome

Microbiologic examination showed negative culture results for aerobial and anaerobial microorganisms. The patient was not allowed weight-bearing during the first month after surgery. After that time, she was allowed for progressive partial weight-bearing for three months.

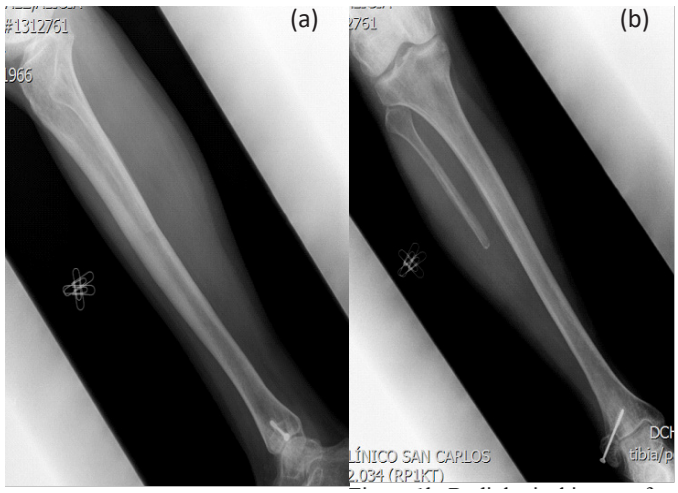

Figure 1 a&b: Radiological image after surgical resection of extraosseus ES.

She did not need the support of professional rehabilitation.

At this moment, fourteen months after surgery, plain X-Rays show bony fusion and no implant failures.

At this moment, she walks with full weight-bearing, but she uses one crutch in her right hand.

No adverse effects registered at any moment of the treatment or outcome.

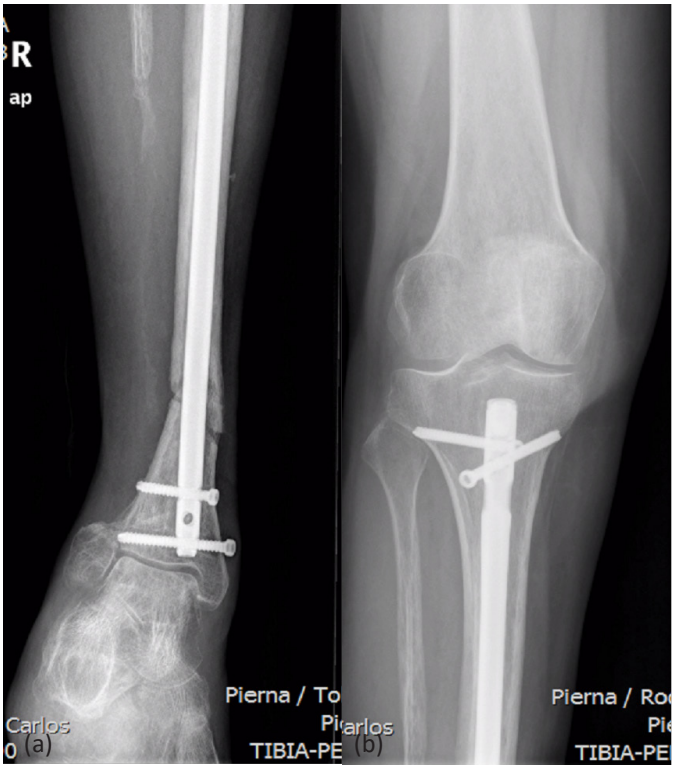

Figure 2 a&b: Tibial pseudoarthrosis of the pathologic fracture after endomedular nailing.

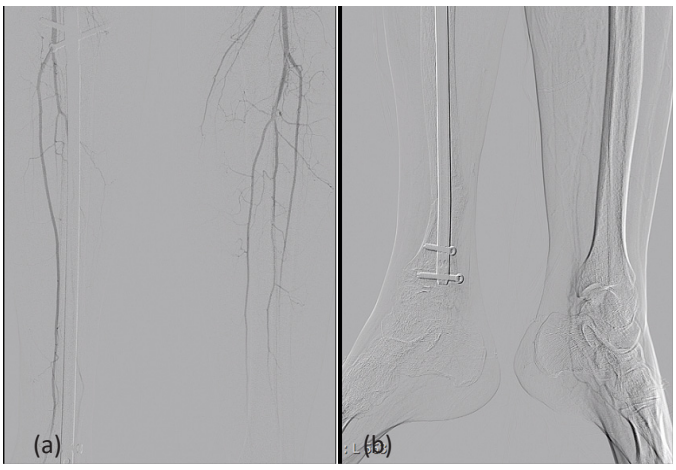

Figure 3 a&b: Lower limb arteriography (Explication in text).

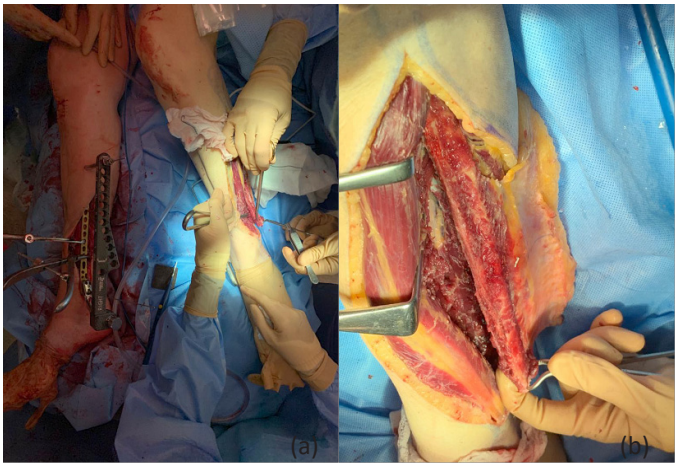

Figure 4 a&b: Osteocutaneous fibula flap from healthy leg (left).

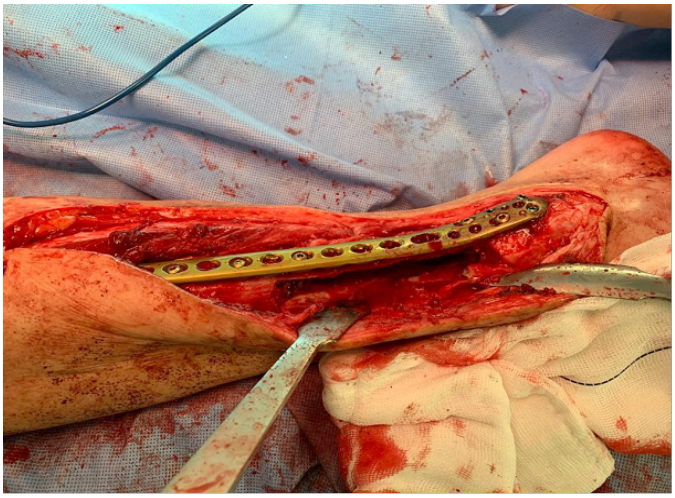

Figure 5: Intramedullary socket for the inset of the fibula.

Figure 6: Detail of skin paddle two days after surgery.

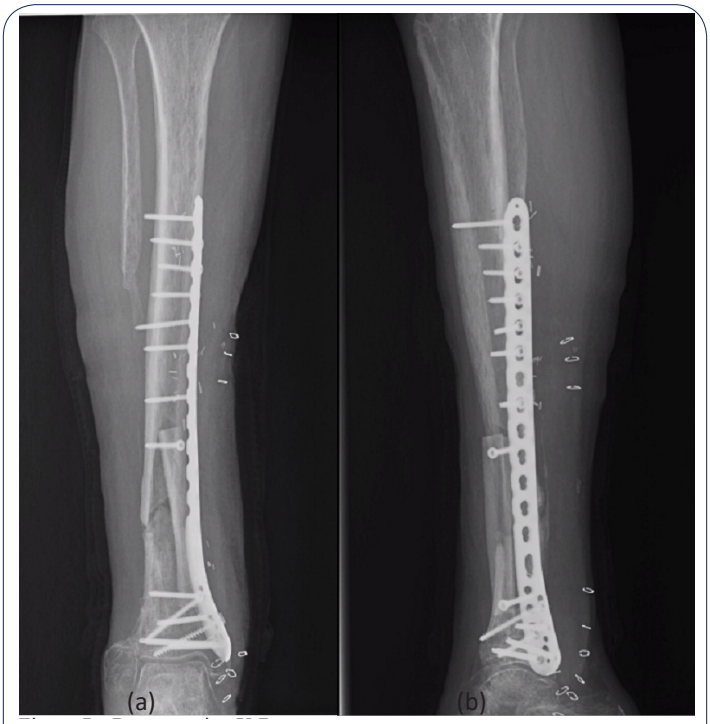

Figure 7 a&b: Postoperative X-Ray control.

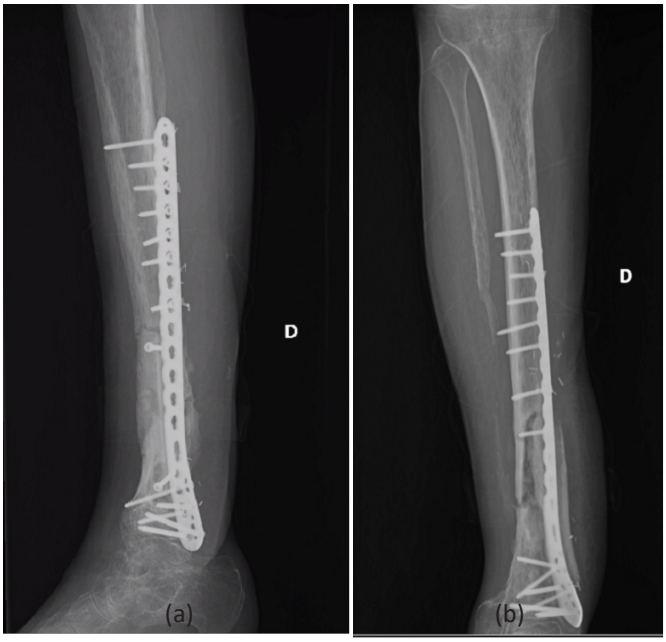

Figure 8 a&b: Outcome one year after surgery with consolidation on the autograft.

Discussion

Vascularized fibular grafts are useful in the reconstruction of large skeletal defects, especially in cases of scarred and avascular recipient sites, or in patients with combined bone and soft-tissue defects. Microvascular free fibula transfer is considered the most suitable autograft for reconstruction of the middle tibia because of its long cylindrical straight shape, mechanical strength, predictable vascular pedicle, and hypertrophy potential. The ability to fold the free fibula into two segments or to combine it with massive allografts is a useful technique for the reconstruction of

massive bone defects of the femur or proximal tibia. It can also be transferred with skin, fascia, or muscle as a composite flap [7].

Conclusion

Combining massive bone allograft and vascularized fibula in intercalary reconstructions following resection of bone tumors represents a complex reconstructive procedure that requires specialists in microvascular surgery as well as orthopedic surgery.

We need to tailor treatment to the patient in terms of size of the defect, location of the defect, the requirement for adjuvant therapy, and surgeons' familiarity with the proposed procedure. However, it is even more critical for the reconstructive surgeon to be involved in the planning of treatment as part of the multidisciplinary team from the beginning to achieve the best possible outcomes (Figure 8a/8b). It's very important the multidisciplinary teams in cases such like this. Other management options are suitable, but vascularized fibula flaps is a safe procedure and use to achieve good outcomes.

Ethical statement and conflict of interest

The submitting authors have not financial or personal conflict of interests. All procedures performed in studies involving human participants were in accordance with the ethical standards of the ethical committee in our hospital. The patient gave informed consent for the submission of this case study for publication. This study was performed in accordance with relevant guidelines and regulations.

References

1. Lim Z, Strike SA, Puhaindran ME. Sarcoma of the Lower Limb: Reconstructive Surgeon's Perspective. Indian J Plast Surg. 2019; 52: 55-61.
2. Beane JD, Yang JC, White D, Steinberg SM, Rosenberg SA, Rudloff U. Efficacy of adjuvant radiation therapy in the treatment of soft tissue sarcoma of the extremity: 20-year follow-up of a randomized prospective trial. Ann Surg Oncol. 2014; 21: 2484–2489.
3. Cannon CP, Lin PP, Lewis VO, Yasko AW. Management of radiation-associated fractures. J Am Acad Orthop Surg. 2008; 16: 541-9.
4. Van Duyn EB, Werker PM, van der Werken C. Free vascularised fibula transplantation for treatment of a fractured femur after radiation therapy. Injury. 2001; 32: 82-6.
5. Uzel A P, Lemonne F, Casoli V. Tibial segmental bone defect reconstruction by Ilizarov type bone transport in an induced membrane. Orthop Traumatol Surg Res. 2010; 96: 194-8.
6. Chen CM, Disa JJ, Lee HY. Reconstruction of extremity long bone defects after sarcoma resection with vascularized fibula flaps: a 10-year review. Plast Reconstr Surg. 2007; 119: 915-24.
7. Beris AE, Lykissas MG, Korompilias AV, Vekris MD, Mitsionis GI, et al. Vascularized fibula transfer for lower limb reconstruction. Microsurgery. 2011; 31: 205-11.