Limb Salvage Surgery using Autogenous Irradiated Bone Graft for Low-grade Central Osteosarcoma of the Distal Tibia: A Case Report

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Abstract

Background: Low-grade central osteosarcoma (LGCOS) is an extremely rare variant of osteosarcoma, and its occurrence in the distal tibia is uncommon. While limb salvage surgery for a primary malignant bone tumor around a knee joint is now accepted worldwide as a standard procedure, challenges to improve the outcome for surgical treatment of lesions of the distal tibia still remain.

Case description: A 26-year-old woman presented with swelling and persistent pain in her left lower leg for 6 months. Incisional biopsy demonstrated LGCOS in the distal tibia. Wide resection and reconstruction using intraoperative extracorporeal autogenous irradiated bone graft (IORBT) were performed. A cross-leg skin flap procedure was added to treat the wound complication of IORBT. There has been no evidence of disease in the last 15 years after surgery.

Conclusion: The key to successful functional and oncological outcomes for the long-term in the rare case of LGCOS in the distal tibia is careful investigation before diagnosis, curative tumor resection, reconstruction using the IORBT procedure, and postoperative care, including the delay of weight-bearing and attaching of a patellar tendon-bearing (PTB) brace.

Clinical significance: IORBT for the patient with LGCOS in the distal tibia is low-cost, safe, and effective.

Keywords: Ankle joint, Distal tibia, Irradiated bone graft, Low-grade central, Osteosarcoma.

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Background

Low-grade central osteosarcoma is an extremely rare variant of osteosarcoma and accounts for approximately 1% of all osteosarcomas.[1] It typically occurs in the 3rd decade of life, and the most common locations are the distal femur and proximal tibia.[1,2] An important differential diagnosis of histology is fibrous dysplasia or desmoplastic fibroma.[1,3–5] Currently, no evidence exists for the effectiveness of chemotherapy for LGCOS. Therefore, it is essential to achieve curative surgical resection for local control and prevention of metastasis in the treatment of LGCOS.

Since the 1980s, limb salvage surgery for a primary malignant bone tumor around a knee joint, following neoadjuvant chemotherapy, has been reported resulting in many studies and is now accepted worldwide as a standard procedure.[6,7] However, changes to improve the outcome of surgical treatment of lesions of the distal tibia still remain.[8] The anatomical features of the distal tibia are as follows: the muscle coverage of the distal tibia is less than that of the distal femur or proximal tibia, and the distal tibia is close to the neurovascular bundle and the ankle joint.

Owing to these reasons, it is difficult to choose a suitable surgical procedure for the patient with LGCOS in the distal tibia. To our knowledge, there has been no report of limb salvage surgery for LGCOS of the distal tibia. Here we report a young female with LGCOS in the distal tibia treated with limb salvage surgery using reconstruction of an IORBT over follow-up periods of 15 years.

Case Description

A 26-year-old woman presented with swelling and persistent pain in her left lower leg for 6 months. She had no history of major trauma.

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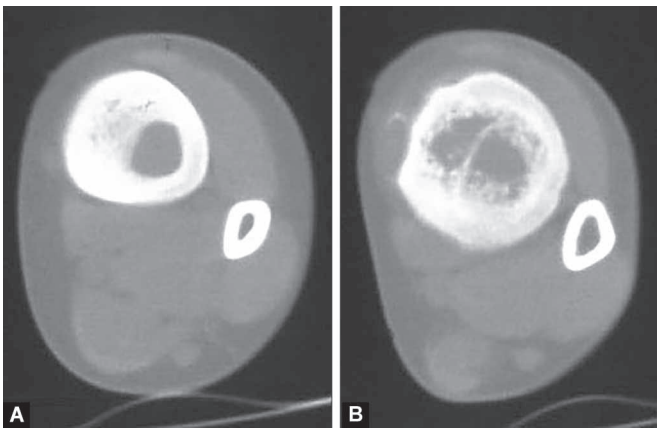
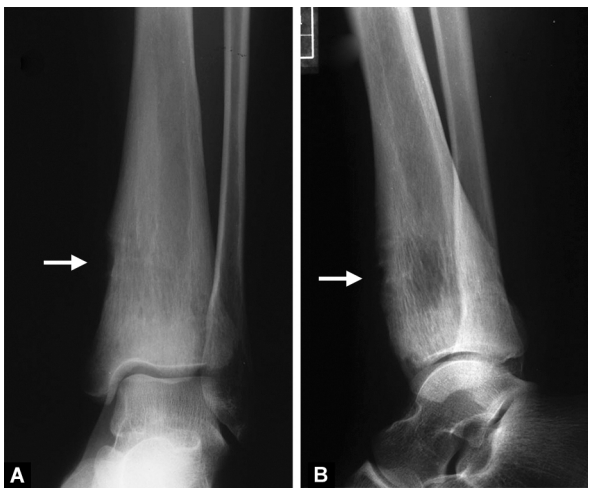
Physical examination revealed swelling on the left lower leg and mild limitation in the range of dorsiflexion motion of the left ankle joint. A plain X-ray of the ankle revealed irregularity of the cortex and osteolysis of the medulla of the left distal tibia (Fig. 1). Computed tomography (CT) revealed a partial thickening of the cortex and multiple patches of osteosclerosis in the endosteal region (Fig. 2). Magnetic resonance imaging (MRI) revealed a lesion of the distal tibia, as well as homogeneous low signal intensity on T1-weighted and heterogeneous low and high signal intensities on T2-weighted images (Figs 3A and B). Radiological differential diagnoses included fibrous dysplasia, osteosarcoma, lymphoma, and osteomyelitis. Incisional biopsy was performed under general anesthesia using an air tourniquet. The tumor tissue extended from the subperiosteal issue to the medullary canal but not to the subcutis and crural fascia.

Microscopically, the tumor was abundant in collagen fibers and composed of spindle cells with mild nuclear atypia. The tumor cells grew into the intertrabecular region resulting in a small focal bone formation. From these microscopic findings, the histological diagnosis of LGCOS was made (Figs 4A and B). Thereafter, two other pathologists with expertise in bone tumors confirmed the same diagnosis. Neither pulmonary metastases on the chest CT nor bone metastases on bone scintigraphy were observed. The surgical staging was stage IB (T2N0M0). Wide resection and reconstruction using IORBT without neoadjuvant chemotherapy were planned. The patient provided informed consent for surgical treatment according to the local institutional review board guidelines.

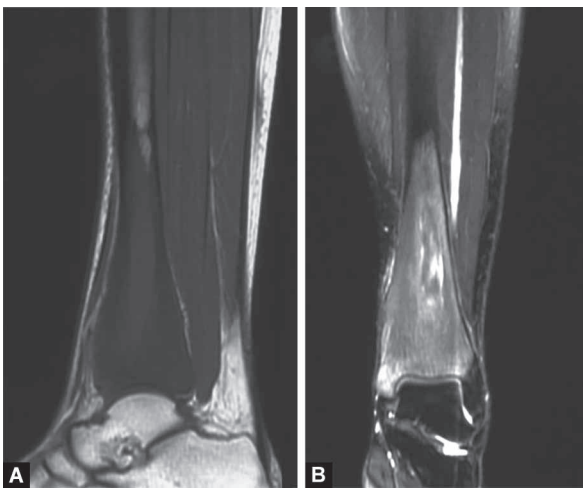
The patient underwent en-blocked resection of the tumor after cutting at the proximal end of the tibial shaft, 15.5 cm away from the ankle joint. The soft tissue surrounding the tumor was trimmed on a separate table, and the tumor was placed in the center to the embedding gauze in a sterilized plastic case, followed by intraoperative irradiation at a dose of 50 Gy according to our



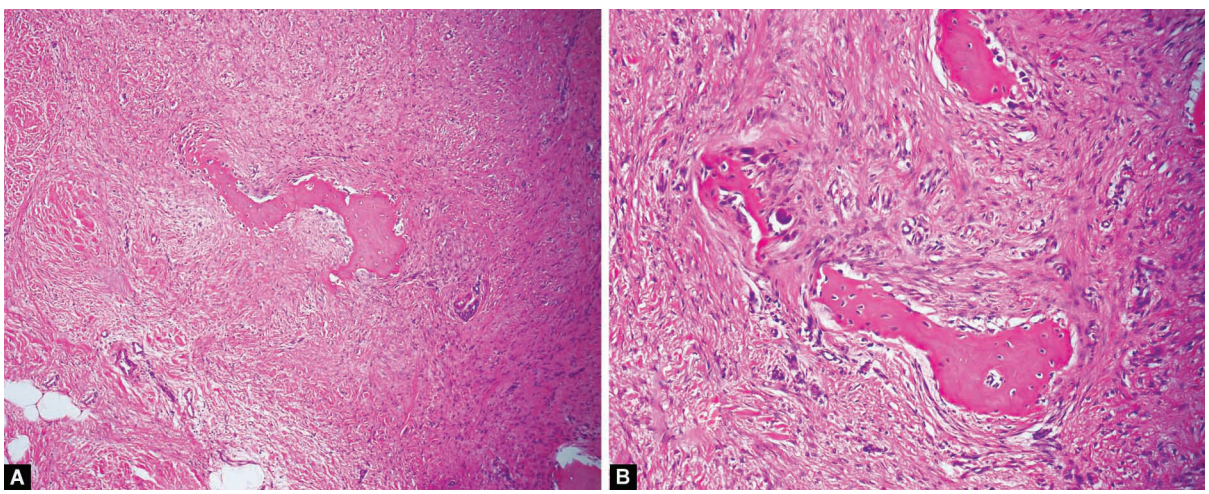
Figs 2A and B: An axial CT demonstrates endosteal thickening of the cortex and the thick and coarse trabeculae in the medullary canal of the tibia. (A) Distal diaphysis; (B) Distal metaphysis



Figs 1A and B: An anteroposterior (A) and lateral (B) plain X-ray examinations show trabecular sclerosis and focal osteolysis in the left distal tibia. The anteromedial aspect of the cortex is irregular



Figs 3A and B: (A) Sagittal MRI (T1-weighted image). The tumor pervades in the medullary canal in the distal tibia, showing homogeneously low signal intensity; (B) Coronal fat-saturated (T2-weighted image). The lesion shows heterogeneous low and high signal intensities



Figs 4A and B: Photomicrograph shows spindle cells with mild nuclear atypia and focal bone formation in a fibrous stroma. (A) 100×; (B) 200× hematoxylin and eosin

procedure for IORBT.[9] The irradiated osteoarticular graft was reimplanted at its original position and fixed using dual locking plates (Figs 5 and 6). The wound healing was delayed, and skin necrosis occurred on the medial malleolar region, resulting in a 25 × 10 mm sized skin defect (Fig. 7A). The scar tissue surrounding the skin defect was largely de-epithelialized (Fig. 7B). The skin defect and the surrounding de-epithelialized surface were covered with a cross-leg skin flap elevated from the medial side of the contralateral lower leg (Fig. 7C). The flap was divided 3 weeks after the surgery (Fig. 7D).

Partial weight-bearing, with the assistance of a PTB brace attached to the left lower leg, was allowed 4 months after the initial surgery. A plain X-ray examination was performed at each monthly follow-up for 1 year. The union between the recycled irradiated bone and the host bone was achieved 7 months after the surgery (Fig. 8). At the current follow-up, 15 years after surgery, neither local recurrence nor distant metastasis has occurred. She walks without support but attaches the PTB brace when going out. Regarding the range of motion of her left ankle joint, dorsiflexion was 15° and plantar flexion was 50° (Fig. 9). The musculoskeletal tumor society score was 93%.[10]

The patient was informed that data concerning the case would be submitted for publication, and she provided consent.

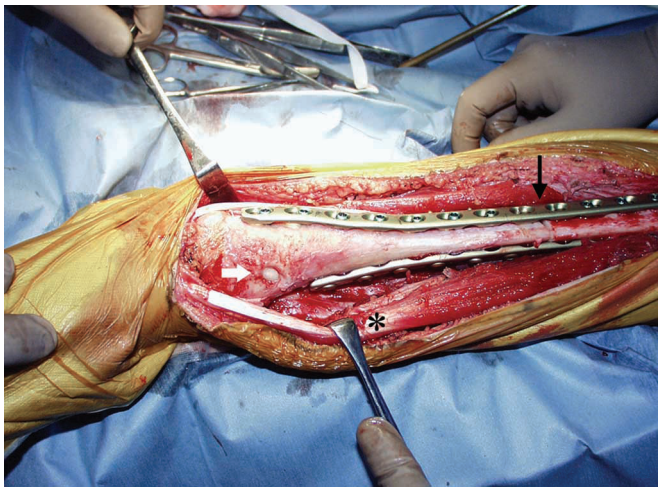


Fig. 5: Intraoperative image. Fixation of irradiated bone (distal tibia) with dual locking plates. Cut line (black arrow). Filling of the medullary canal with bone cement (white arrow). Tibialis anterior muscle (asterisk)

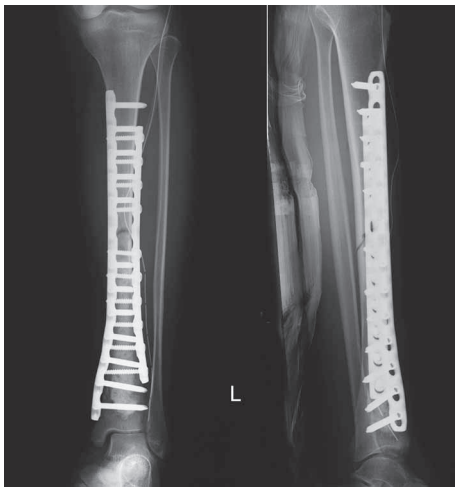
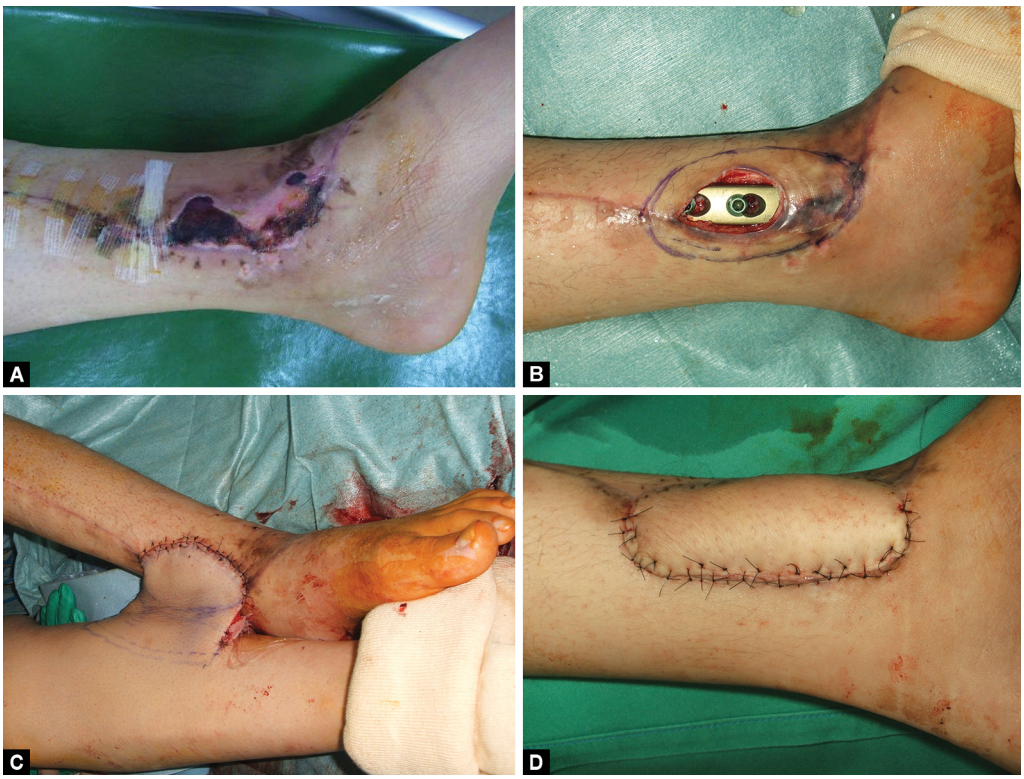


Fig. 6: Postoperative plain X-ray images



Figs 7A to D: (A) Skin necrosis around the medial malleolus; (B) Trimming of skin necrosis; (C) Cross-leg method; (D) Separation of the skin flap

Discussion

Low-grade central osteosarcoma is a well-differentiated fibroblastic osteosarcoma that histologically shows the appearance of benign spindle-like cells and relatively mature bone formation.[1–3,11] Therefore, the histopathological findings of fibrous dysplasia, desmoplastic fibroma, and parosteal osteosarcoma occasionally resemble those of LGCOS and should be excluded to confirm its diagnosis.[1,3,4,12] Since LGCOS is not sensitive to chemotherapy, local surgery, including wide resection or amputation, is thought to be the only credible treatment. An early study showed a high recurrence rate after curettage or marginal excision, and these cases eventually required amputation.[2,3] In addition, four of five distal tibial cases underwent amputation or disarticulation as an initial surgery.[3,11] In a study on lower limb LGCOS from the United Kingdom, 17 of 18 cases received limb salvage surgery, and their disease-free survival rate was over 90% at 5 years and 80% at 10 years.[13] However, a distal tibial case was not included in this study.

Limb salvage surgeries for conventional osteosarcoma around the knee or hip joints were established in the 1900s, and prosthetic replacement as a reconstruction procedure after tumor resection is commonly used today.[6,7,14] However, cases of distal tibial osteosarcoma are still challenging because of the presence of a small volume of soft tissue and because the distal tibia is closed to the neurovascular bundle and the ankle joint.[7,8] Therefore, below-knee amputation was a standard therapy for patients with malignant bone tumors in the distal tibia until the 1990s. Thereafter, retrospective studies comparing limb salvage surgery with amputation in the distal tibial cases indicated approximately equivalent oncological and functional outcomes.[15,16] However, most of the cases received chemotherapy, probably because the safety of the surgical procedure was achieved. Two of seven patients who did not receive chemotherapy developed local recurrence after surgery and required amputation.[15] The local recurrence rate of limb salvage in the distal tibia is variable in each report: 0,[17] 12,[16] 15,[18] 16,[19] and 33%.[20] The difference between studies may be attributed to histology, tumor size, and indications for limb salvage in each institution. Nevertheless, the risk of local recurrence after limb salvage surgery without neoadjuvant therapy seems to be higher than that after amputation. Therefore, limb salvage surgeries for these lesions are applicable only if neoadjuvant chemotherapy is effective and there is no tumor extension to the subcutis. Even if chemotherapy is administered, when the tumor extends to the subcutis during preoperative evaluation, it is not indicated for limb salvage surgery, and amputation is strongly recommended. In the present case, we confirmed no tumor extension to the subcutis through biopsy and judged that the fascia was a safe surgical margin for curative resection of the tumor. For this reason, we judged that limb salvage surgery was possible in this case.

Fig. 8: Union at the site of reimplantation of the irradiated bone graft

Various procedures of reconstruction after tumor resection in this location, including the allograft,[15,16,21] the autograft including arthrodesis[17–19,22] and biological recycled bone,[23,24] and the prosthesis[20,24–27] have been performed and have both advantages and disadvantages. The main obstacles in bone graft surgery include delayed or nonunion in the graft site and collapse or fracture after weight-bearing.[15,16] Although these complications were relatively few in the prosthetic replacement, infection and loosening remain problems to resolve. Moreover, these prostheses are not applicable to clinical practice because most of all implants are custom-made.[20,24–27] Historically, we had been performing autogenous irradiated bone grafting for the distal tibial lesions of malignant bone tumors.[9,24] In the present case, we devised three strategies to avoid the collapse of the graft such as experienced in the previous osteosarcoma case: (1) filling bone cement to the distal epiphysis and metaphysis of the tibia at surgery, (2) delaying the time of full weight-bearing, (3) attaching a PTB brace to the affected lower leg for a long period of time. Excellent functional

Fig. 9: The active range of motion of the left ankle 15 years after surgery

results were eventually achieved after utmost care was taken to follow the devised treatment regimen.

The good long-term oncological and functional outcomes of our case justified the decision to perform this limb salvage procedure using autogenous irradiated bone graft, despite the requirement of additional skin flap surgery for treatment of wound complications. To our knowledge, the long-term outcome of limb salvage surgery for the above lesion has not yet been reported.

We believe that this procedure is low-cost, safe, and effective, and thus an appropriate treatment choice for similar cases.

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