E Journal of Cardiovascular Medicine
EJCM 2021;9(1):70-75
DOI: 10.32596/ejcm.galenos.2020-11-062

Being on the Edge of Amputation: Vessel-sparing Approach and Biological Bone Reconstruction in a Toddler with Extremity Rhabdomyosarcoma

Atilla Orhan[1], Bahattin Kerem Aydın[2]

[1]Selçuk University Faculty of Medicine, Department of Cardiovascular Surgery, Konya, Turkey
[2]Selçuk University Faculty of Medicine, Department of Orthopedic and Traumatology, Konya, Turkey

Abstract

Vascular reconstruction leads to surgical difficulties such as acute occlusion and critical leg ischemia in childhood rhabdomyosarcoma (RMS) pressuring on the limb's vascular structures. Finally, amputation is inevitable. Also, use of prostheses to repair bone defects in these cases results in long-term failure or complications such as infection. To avoid a catastrophic problem such as amputation, a vessel-sparing approach and biological bone reconstruction may be a solution in these cases. Here, we present a case of total surgical tumor excision that keeps the native vascular and biologic bone reconstruction in a RMS case involving the left thigh in a two-year-old toddler.

Keywords: Rhabdomyosarcoma, limb-salvage, vessel-sparing surgery, soft tissue sarcoma

Introduction

Rhabdomyosarcoma (RMS), a soft tissue sarcoma, is very common in children and accounts for approximately 10% to 15% of solid malignant tumors[1-3]. The RMS is heterogeneous and has a rapid, invasive, and infiltrative growth pattern[4]. The outcome depends on primary involvement site, tumor size, histological structure, and the patient's age[5]. The limbs are invaded in 20%

Address for Correspondence: Atilla Orhan, Selçuk University Faculty of Medicine, Department of Cardiovascular Surgery, Konya, Turkey
e-mail: atillaorhan@gmail.com **ORCID:** orcid.org/0000-0003-0110-3290
Received: 05.11.2020 **Accepted:** 22.01.2021
Cite this article as: Orhan A, Aydın BK. Being on the Edge of Amputation: Vessel-sparing Approach and Biological Bone Reconstruction in a Toddler with Extremity Rhabdomyosarcoma. EJCM 2021;9(1):70-75.
DOI: 10.32596/ejcm.galenos.2020-11-062

of the cases[6]. The cure chance of RMS increases with a multidisciplinary treatment approach that includes chemotherapy, radiotherapy, and wide excision surgery[2, 7]. Today, limb salvage surgery is applied as standard treatment with the development of surgical techniques, imaging methods, and the presence of vascular grafts[8,9]. Autologous grafts are preferred in these cases compared to prosthetic grafts due to their superior patency[8,10,11]. However, the choice of vascular grafts in children is difficult due to the size limitations of autologous grafts, unusable small-diameter prosthetic grafts, and insufficient surgical experience. The difficulty is increased in toddlers, and limb salvage with vascular repair may be impossible[12].

Although synthetic bone grafts used to repair the extremity defect provide recovery in the short-term, they are not preferred due to infection and durability problems in the long-term. Biological reconstruction grafts obtained by processing tumor-containing bone tissues with liquid nitrogen may be a more natural and permanent solution[13,14].

A vascular-sparing surgical approach and the biologic bone autograft technique in childhood RMS will save the limb from amputation because of these difficulties.

Here, we present a case of total surgical tumor excision that keeps the native vascular and biologic bone reconstruction in the RMS case involving the left thigh in a two-year-old toddler.

Case Report

A two-year-old girl was admitted with a huge left thigh mass adjacent to the femoral bone's medial side. The patient had pain in her left thigh with stiffness and swelling for about five months. The patient did not have a family history of malignant disease or recent trauma. There was a mass of 6 cm x 12 cm in the left thigh on physical examination (Figure 1). No neurologic deficits were detected. On magnetic resonance imaging (MRI), a large mass of 6 cm x 12 cm was detected in the middle of the left thigh, compressing the neurovascular bundle (Figure 2a). The MRI showed that the femoral artery, femoral vein, and femoral nerve were compressed laterally by the tumor (Figure 2b). On positron emission tomographic imaging, the patient had a few small metastatic lesions in her lung. A tissue sample was taken from the patient with a trucut needle biopsy under ultrasound guidance with an anteromedial approach. Histopathological diagnosis was embryonal RMS. The lesion in the femoral bone segment persisted after preoperative chemotherapy.

Surgical Procedure

Two weeks after the diagnosis, the surgical procedure was performed with an anteromedial approach to protect the natural vascular structure and save the limb (Figure 3). Skin and subcutaneous incisions were made from the groin to the distal of the limb in a longitudinal fashion. The neurovascular bundle was exposed without touching the

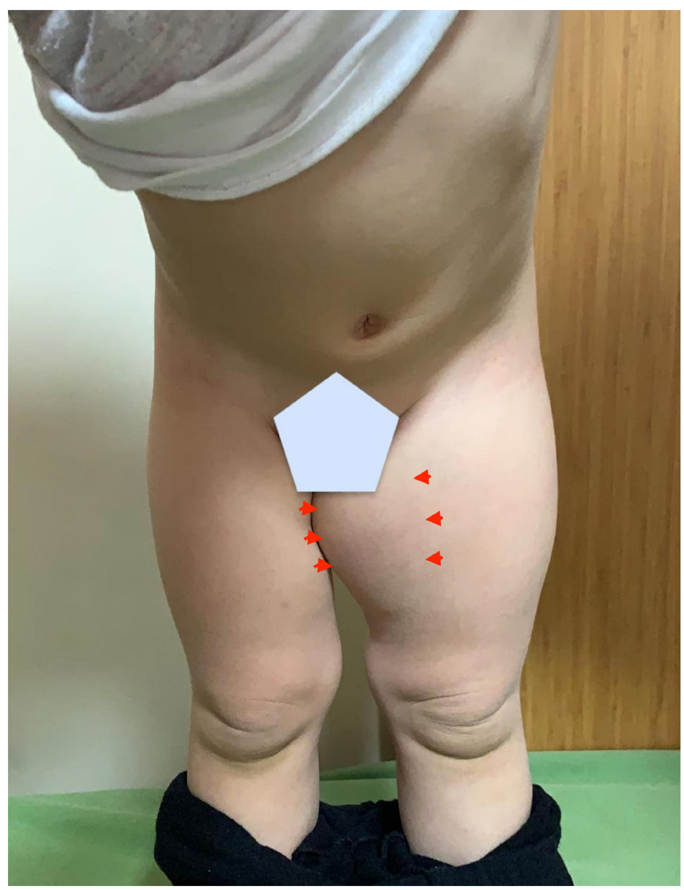

Figure 1. Preoperative view. The arrows show the soft tissue mass borders

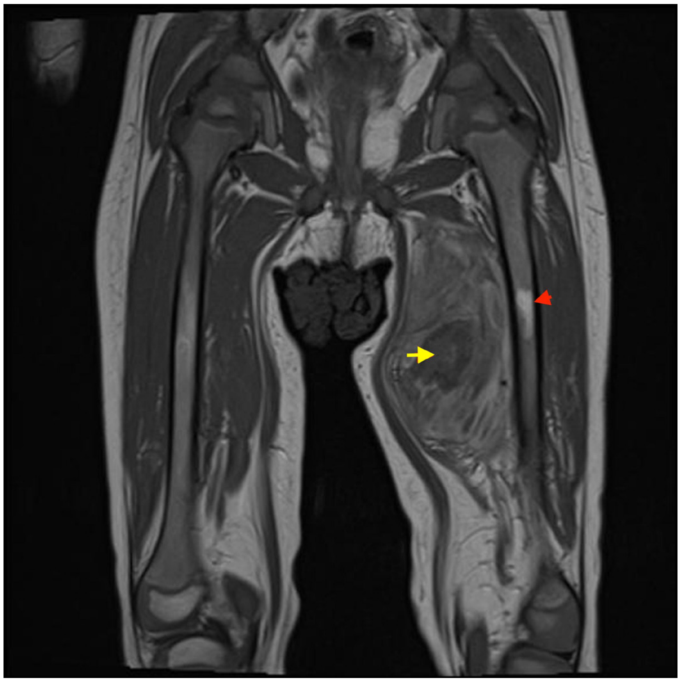

Figure 2a. On MRI, the yellow arrow shows the soft tissue mass in the left thigh's medial border, and the red arrow shows the bone involvement of the soft tissue mass

MRI: Magnetic resonance imaging

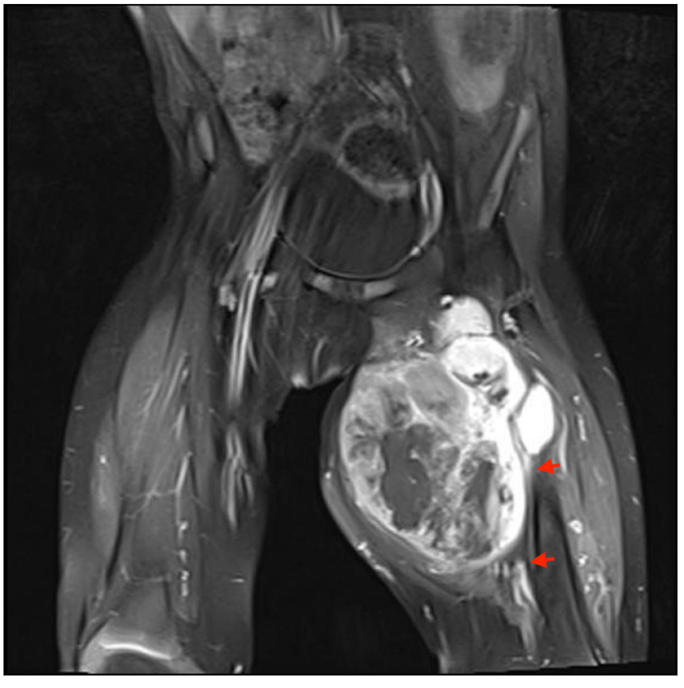

Figure 2b. Compressed neurovascular bundle on the MRI. The arrows show the compressed neurovascular bundle

MRI: Magnetic resonance imaging

tumor from the lateral of the tumor. The superficial femoral artery and vein and the femoral nerve were exposed and protected by making a gentle dissection. Since the most important factor for successful sarcoma surgery is wide resection, the deep femoral artery was ligated just below the femoral bifurcation and the deep femoral vein at the level of the saphenofemoral junction. Distal pulses were checked, and no ischemia was observed. Both arterial and venous flows were sufficient.

According to the concept of surgical margins, wide resection was made with soft tissue mass and 5-centimeter femoral bone resection (Figure 4). The tumoral mass was stripped from the bone segment, including the periosteum. Aggressive curettage was applied to the intramedullary cavity. Then, the bone segment was frozen with liquid nitrogen for 20 minutes. The bone segment was thawed at room temperature for 20 minutes and in distilled water for 15 minutes. Ultimately, a bone graft was obtained, which provided the best morphological matching from the bone segment. (Figure 5). Subsequently, the bone graft was fixed with a 3.5 mm reconstruction plate (Figure 6). The wound was irrigated and sutured in a standard fashion. The operation was completed successfully.

The patient's pedal pulses were checked regularly after surgery. The patient was discharged on the seventh postoperative day without any signs of ischemia.

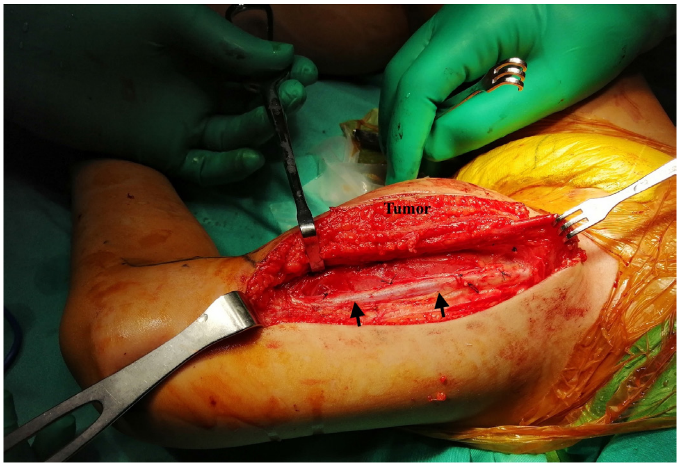

Figure 3. Intraoperative view. The arrows show spared-vascular structures during the surgery

The pediatric oncology clinic planned postoperative radiotherapy for lung metastasis and neo-adjuvant chemotherapy. Pathological samples confirmed the diagnosis and showed that all sides of the resected tumor mass were tumor-free.

Results

There was no loss of function in the left lower limb (Figure 7). In the postoperative second month's control examination, the patient was walking comfortably, and no neurological or orthopedic problems were detected. The patient continued the chemotherapy program.

Currently, the patient has had her five-month follow-up. The patient had no limb length discrepancy at her last follow-up. There was no problem with her bone growth and limb circulation. Finally, no tumor recurrence was detected in the left lower limb of the patient during the five-month follow-up.

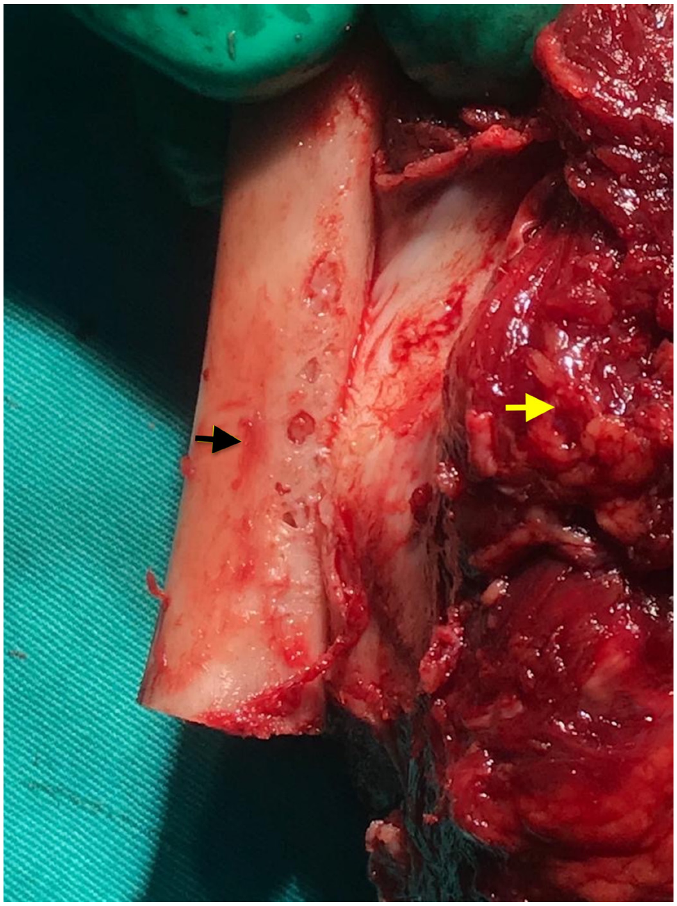

Figure 4. Perioperative view of the resected tumor involving soft tissue (yellow arrow) and the bone part (black arrow)

Discussion

Soft tissue sarcomas such as RMS is a rare condition in children[8]. Limb loss may occur because of vascular involvement by the tumor[15]. Vascular reconstructive surgery may be required in such cases[2]. However, in

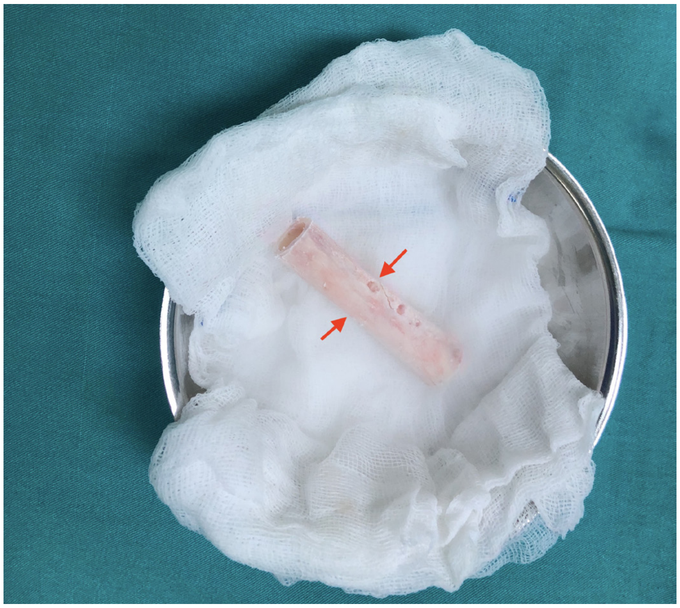

Figure 5. The biological bone graft prepared with the liquid-nitrogen application. The red arrows show the tumor-free bone graft

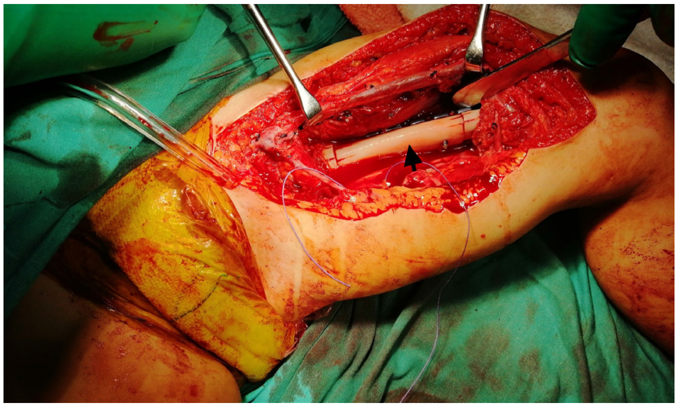

Figure 6. Perioperative view of the re-implanted biologic bone graft. The arrow shows the re-implanted bone segment

infants and toddlers, reconstruction is difficult due to small diameter vein grafts, or prosthetic grafts, which trend to thrombosis[12]. Vascular reconstruction surgery may be necessary to avoid catastrophic consequences such as amputation due to distal limb ischemia[15]. However, in toddlers, arteries or veins are of relatively small diameter[12]. Therefore, the vascular reconstruction procedure is often complicated due to small-sized native grafts or small diameter prosthetic grafts. Sometimes, amputation is mandatory because artificial grafts applied to small vessels are thrombosed[12].

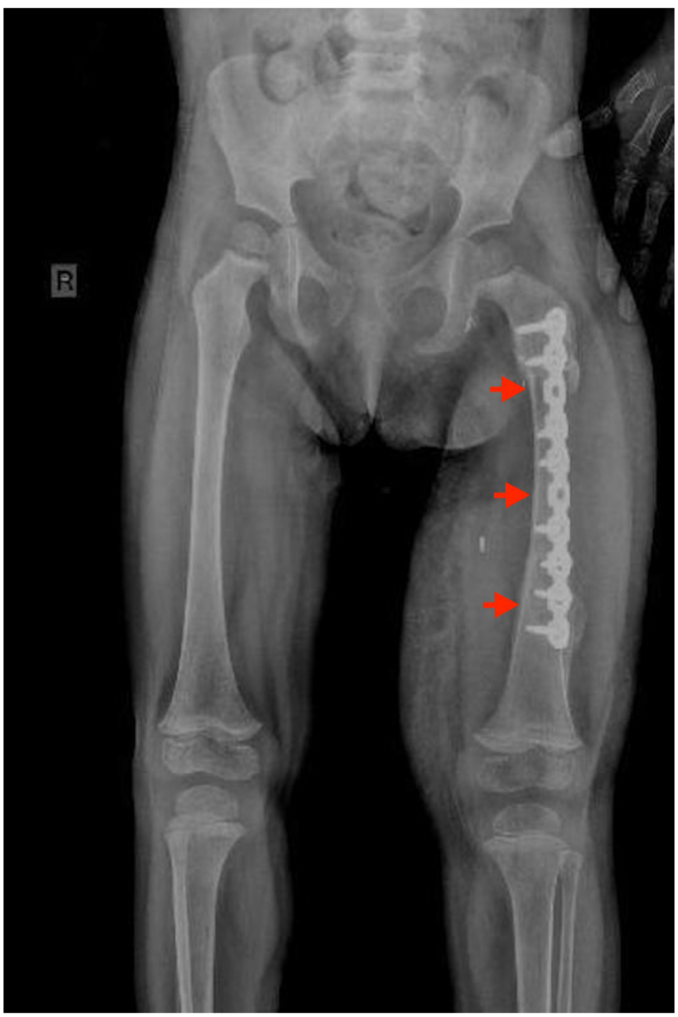

Figure 7. Postoperative second-month control X-ray image. The arrows show that the re-implanted biologic bone graft integrated with the host bone. The union of the bone graft and the host bone can be seen on the X-ray image

Furthermore, the neurovascular bundle is under pressure if the tumor invades the package[3,16]. There are various intervention scenarios to preserve the affected limb[16]. Although otherwise claimed[12], most surgeons cannot dare to ligate the main arteries in the toddlers in tumor operations with a high risk of amputation by trusting on collateral circulation. There are insufficient diameter and length of native grafts, such as a great saphenous vein in toddlers[12], and a high risk of thrombosis of prosthetic grafts complicates the surgical intervention to recover the affected limb[2,8,16]. There are two options back to recover the tumor invaded by the tumor; amputation or vessel-sparing surgery. Vessel-sparing surgery is possible only with an experienced surgical team and a rigorous tissue resection, and the results are excellent[1,4,6-9,15,16]. Today, limb salvage surgery is now accepted as a standard treatment method in limb sarcomas[8,9], and the number of surviving limbs is higher than the number of amputations[1,7]. This treatment model change was made possible by advancing surgical techniques, introducing new imaging methods, and developing vascular grafts[13].

Our team was ready before the operation for possible vascular reconstruction scenarios. In case of failure to protect the vascular structures, our first alternative was to create a new graft by opening the great saphenous vein of the patient with a diameter of 1.7 mm linearly and wrapping it around a Hegar dilator. In case this was not possible, we had planned to use a biological vein graft. Our worst scenario was transfemoral amputation.

Another goal of the treatment should also be to restore bone defects that preserve the affected limb's length and function. Arthroplasty is one of the choices, especially in adult patients. However, biological reconstruction is important, especially in children. We used the biological reconstruction technique "frozen autograft treated with liquid nitrogen" for this toddler[13,14].

Liquid nitrogen-inactivated autologous bone graft is a safe method for primary malignant extremity tumors. This method has a low complication rate and high bone healing rate, and also offers a satisfactory limb function[17].

In our case, the patient's age, clinical and radiographic findings have encouraged us to prefer a limb-sparing surgical technique that preserves native neurovascular bundle and bone structure. Our approach in toddlers with malignant limb soft tissue tumors adjacent to vascular structures can be applied successfully and it can prevent amputation. We believe that the vessel-sparing surgery can be the first choice for standard surgical technique in toddlers in terms of better cosmesis, low morbidity, and loss of function in huge sarcomas threatening the limb.

Ethics

Informed Consent: The informed consent forms were obtained for the patient.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: A.O., B.K.A., Concept: A.O., B.K.A., Design: A.O., B.K.A., Data Collection or Processing: A.O., B.K.A., Analysis or Interpretation: A.O., B.K.A., Literature Search: A.O., B.K.A., Writing: A.O., B.K.A.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Kumta SM, Cheng JC, Li CK, Griffith JF, Chow LT, Quintos AD. Scope and limitations of limb-sparing surgery in childhood sarcomas. J Pediatr Orthop 2002;22:244-8.
2. McGoldrick NP, Butler JS, Lavelle M, Sheehan S, Dudeney S, O'Toole GC. Resection and reconstruction of pelvic and extremity soft tissue sarcomas with major vascular involvement: Current concepts. World J Orthop 2016;7:293-300.
3. Ng WK. Embryonal rhabdomyosarcoma in a young boy. Mcgill J Med 2007;10:16-9.
4. Wolden SL, Alektiar KM. Sarcomas across the age spectrum. Semin Radiat Oncol 2010;20:45-51.
5. Zhu L, Sun Y, Wang X, et al. Survival stratification in childhood rhabdomyosarcoma of the extremities: a derivation and validation study. Sci Rep 2020;10:5684.
6. Hosalkar HS, Dormans JP. Limb sparing surgery for pediatric musculoskeletal tumors. Pediatr Blood Cancer 2004;42:295-310.
7. Marchetti AA, Di Giulio L, Oddi FM, Davila BO, Ciattaglia R, Ippoliti A. Strategy to preserve function in lower limbs tumors resection. Vascular competence in multidisciplinary approach. 2020;1-12.
8. Khalefa S, El Bahaey A, El-Samadoni A, Eldmarany HA. Clinical outcomes after wide resection of lower extremity soft tissue sarcomas with femoral vessel reconstruction. Egypt J Surg. 2020;39:206-14.
9. Steinau HU, Daigeler A, Langer S, et al. Limb salvage in malignant tumors. Semin Plast Surg 2010;24:18-33.
10. Nishinari K, Krutman M, Aguiar Junior S, et al. Surgical outcomes of vascular reconstruction in soft tissue sarcomas of the lower extremities. J Vasc Surg 2015;62:143-9.
11. Nishinari K, Wolosker N, Yazbek G, Zerati AE, Nishimoto IN. Venous reconstructions in lower limbs associated with resection of malignancies. J Vasc Surg 2006;44:1046-50.
12. Aponte-Tinao L, Farfalli GL, Ayerza MA, Muscolo DL. Case report: Common femoral artery ligation after sarcoma resection in an infant. Clin Orthop Relat Res 2011;469:609-12.
13. Igarashi K, Yamamoto N, Shirai T, et al. The long-term outcome following the use of frozen autograft treated with liquid nitrogen in the management of bone and soft-tissue sarcomas. Bone Joint J 2014;96:555-61.
14. Yamamoto N, Hayashi K, Tsuchiya H. Progress in biological reconstruction and enhanced bone revitalization for bone defects. J Orthop Sci 2019;24:387-92.
15. Ramamurthy R, Soundrarajan JC, Mettupalayam V, Shanmugham S, Arumugam B, Periasamy S. Limb conservation in extremity soft tissue sarcomas with vascular involvement. Indian J Orthop 2009;43:403-7.
16. Song TK, Harris EJ Jr, Raghavan S, Norton JA. Major blood vessel reconstruction during sarcoma surgery. Arch Surg 2009;144:817-22.
17. Li Y, Yang Y, Huang Z, Shan H, Xu H, Niu X. Bone defect reconstruction with autologous bone inactivated with liquid nitrogen after resection of primary limb malignant tumors: An observational study. Medicine (Baltimore) 2020;99:20442.