

Received: 2019.12.19

Accepted: 2020.03.19

Available online: 2020.04.22

Published: 2020.05.22

Emergency Surgery in High Volume Osteosarcoma of Left Proximal Humerus Due to Vascular Compromise: A Case Report

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Conflict of interest: None declared

Patient: Male, 14-year-old
Final Diagnosis: Dead limb • osteosarcoma
Symptoms: Lump on left arm • numbness of the fingers on the left hand • pain
Medication: —
Clinical Procedure: Forequarter amputation
Specialty: Oncology • Orthopedics and Traumatology

Objective: Unusual clinical course

Background: Osteosarcoma is the most common type of malignant bone tumor arising from mesenchymal stem cell. When occurring on the proximal humerus, it is associated with poor outcomes; there are numerous neurovascular structures around proximal humerus. Unfortunately, the degree of vascular involvement in osteosarcoma is rarely evaluated and reported. Thus, we would like to highlight our case.

Case Report: We reported a case of left proximal humerus osteosarcoma causing dead limb in a 14-year-old boy. The dead limb progressed in the span of 3 weeks. An emergency forequarter amputation (FQA) was conducted to prevent further complications such as septicemia and mortality. Two months after the surgery, the patient had improved quality of life. One year after, the patient had no local recurrence. However, there was a lung metastasis detected 9 months after the surgery. The patient died 13 months after the surgery.

Conclusions: Osteosarcoma of the proximal humerus can potentially compromise vascular structures. Early diagnosis and treatment are mandatory to prevent such complications.

MeSH Keywords: Amputation • Bone Neoplasms • Humerus • Ischemia • Osteosarcoma

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/922257>



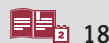
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Background

Osteosarcoma is the most common type of bone tumor arising from primitive mesenchymal stem cell. In our hospital, osteosarcoma is also the most prevalent type of bone tumor. This tumor has a 2-peak incidence: the first is among children age 10–14 years old, and the second is among older adults age >65 years old. Although the mortality from osteosarcoma has declined steadily each year, the rate is still as high as nearly 70% among the children affected [1].

The goal of osteosarcoma treatment is a complete resection of the tumor to prevent recurrence and improve patient survival. The treatment usually consists of preoperative chemotherapy regimen, followed by limb salvage surgery or amputation. Several studies have shown that limb salvage surgery is more favorable, because the procedure preserves the limb with a comparable recurrence rate and better 5-years survival rate compared to amputation [2]. Unfortunately, advanced cases still require surgical amputation to control the disease [3].

Hu et al. found that prolonged symptoms before initiation of treatment was associated with worse overall survival [4]. Whereas one cohort study found that delayed surgical control of more than 14 weeks was associated with worse prognosis in cases of osteosarcoma. The event-free survival (EFS) in patients with timely local control was found to be significantly higher than those with delayed control, 80% versus 58% [5]. However, a shorter delay also possibly leads to a less favorable outcome such as in the case reported here. Other than treatment delay, tumor volume and its progression are predictors of worse survival rate in osteosarcoma of humerus [6]. We present a case of delayed high-grade progressive

osteosarcoma prompting emergency forequarter amputation (FQA) due to dead limb.

Case Report

A 14-year-old boy presented to our hospital with a chief complain of a lump on his left arm for 3 months. The initial size was not significantly large but expanded rapidly to the present size. The patient also complained of dull, intermittent pain which was aggravated with arm movement. Since the previous month, the patient felt numbness on the fingers of his left hand and had difficulty moving them. He had no history of prior trauma. His past medical history and familial history were not remarkable.

During the first physical examination, the mass on the proximal humerus was 64 cm in diameter; the overlying skin was shiny with bullae. In the course of 3 weeks, the diameter increased to 84 cm, and the overlying skin became red with pronounced heat on palpation; his left-hand fingers and distal forearm became necrotic, and the patient could not feel any sensation nor execute any active movements of his fingers. The initial clinical photograph is depicted in Figure 1A, and the clinical photograph 3 weeks after is depicted in Figure 1B.

The plain radiography showed a lytic lesion on the proximal humerus, extending through the diaphysis, metaphysis, and epiphysis (Figure 2). The lesion was in a central position with periosteal reaction (Codman's triangle and sunburst appearance). There was a wide transition zone with soft tissue involvement. There was no pathological fracture. We ordered a chest radiograph to rule out metastases; there was no evidence of metastasis on the chest radiograph. We did not conduct



Figure 1. (A) Swelling on left proximal humerus, 64 cm in diameter during the initial physical examination. There was no necrosis found on distal part, (B) The patient's left arm 3 weeks after the initial examination. The mass increased in size into a diameter of 84 cm, and there was marked necrosis of the forearm and hand.

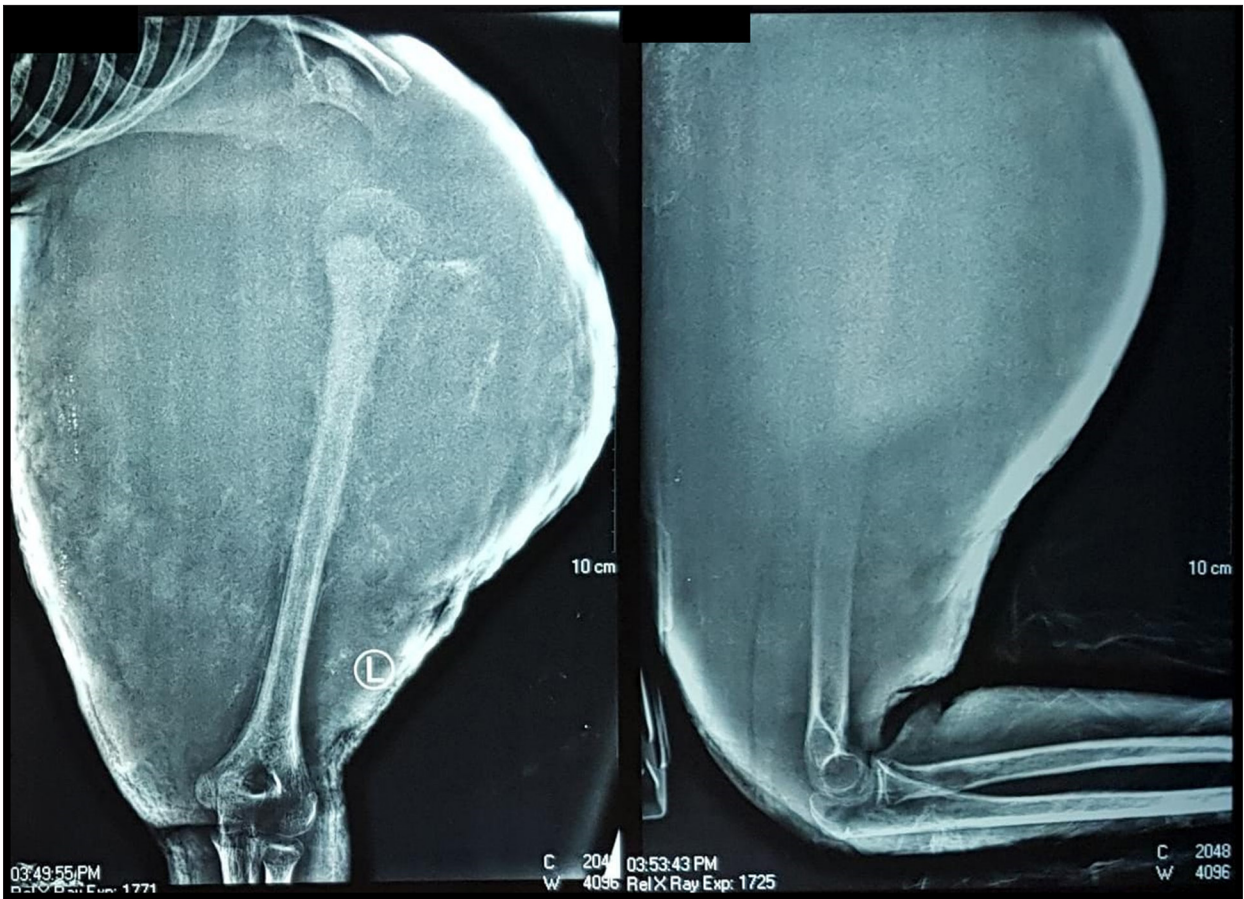


Figure 2. The plain radiograph of the patient's left humerus, anteroposterior (AP) and lateral (L) view.

a computed tomography (CT) or magnetic resonance imaging (MRI) examination because the clinicopathological conference (CPC) in our hospital concluded that additional CT or MRI would not change the preoperative planning. CT or MRI would only delay local control of the disease.

Laboratory examination showed elevation of alkaline phosphatase (ALP) of 378 U/L and lactate dehydrogenase (LDH) of 1605 U/L. There was leukocytosis of 25 510/ μ L, elevated C-reactive protein (CRP) of 0.53 mg/dL, and elevated erythrocyte sedimentation rate (ESR) of 72 mm/hour. Histopathologic examination of fine needle aspiration biopsy (FNAB) specimen showed pleomorphic small round cells with osteoid matrix.

We decided to do FQA of the patient's left arm after improving the patient's general condition in order to prevent further complications such as septicemia and death. The blood loss was significant after the initial incision, so the subclavian artery was clamped. The surgeon then performed guillotine amputation at the shoulder level and bleeding was controlled after limb removal. The brachial plexus and axillary vessels were ligated, and the wound was closed with placement of a drain. The surgical margin was radical in our case, but the focus of our surgery

was to prevent mortality due to septic complication of the dead limb. The intraoperative photographs are depicted in Figure 3.

Postsurgical biopsy showed specimen with anaplastic osteoblast proliferation and pleomorphic small round cells. The final histopathologic findings were consistent with central high-grade osteosarcoma of osteoblastic subtype.

After the surgery, the patient was started on postoperative chemotherapy of intravenous cisplatin and doxorubicin. He received 100 mg/m² cisplatin and 25 mg/m² doxorubicin. The therapy was performed for 10 cycles, 1 time every 3 weeks. Each therapy was performed in 3 days; cisplatin was administered on the first day only, and doxorubicin was administered every day.

Two months after the surgical procedure, the patient's quality-of-life (QOL) had improved with Karnofsky score of 90 and increased body weight. On 1-year follow-up, the patient did not complain of any pain, and there was no local recurrence (Figure 4). Unfortunately, 9 months after surgery, there was a lung metastasis detected by chest radiograph. Chest CT-scan on the 13th month follow-up confirmed a metastatic process in the right lung. The patient passed away 13 months after the operation.

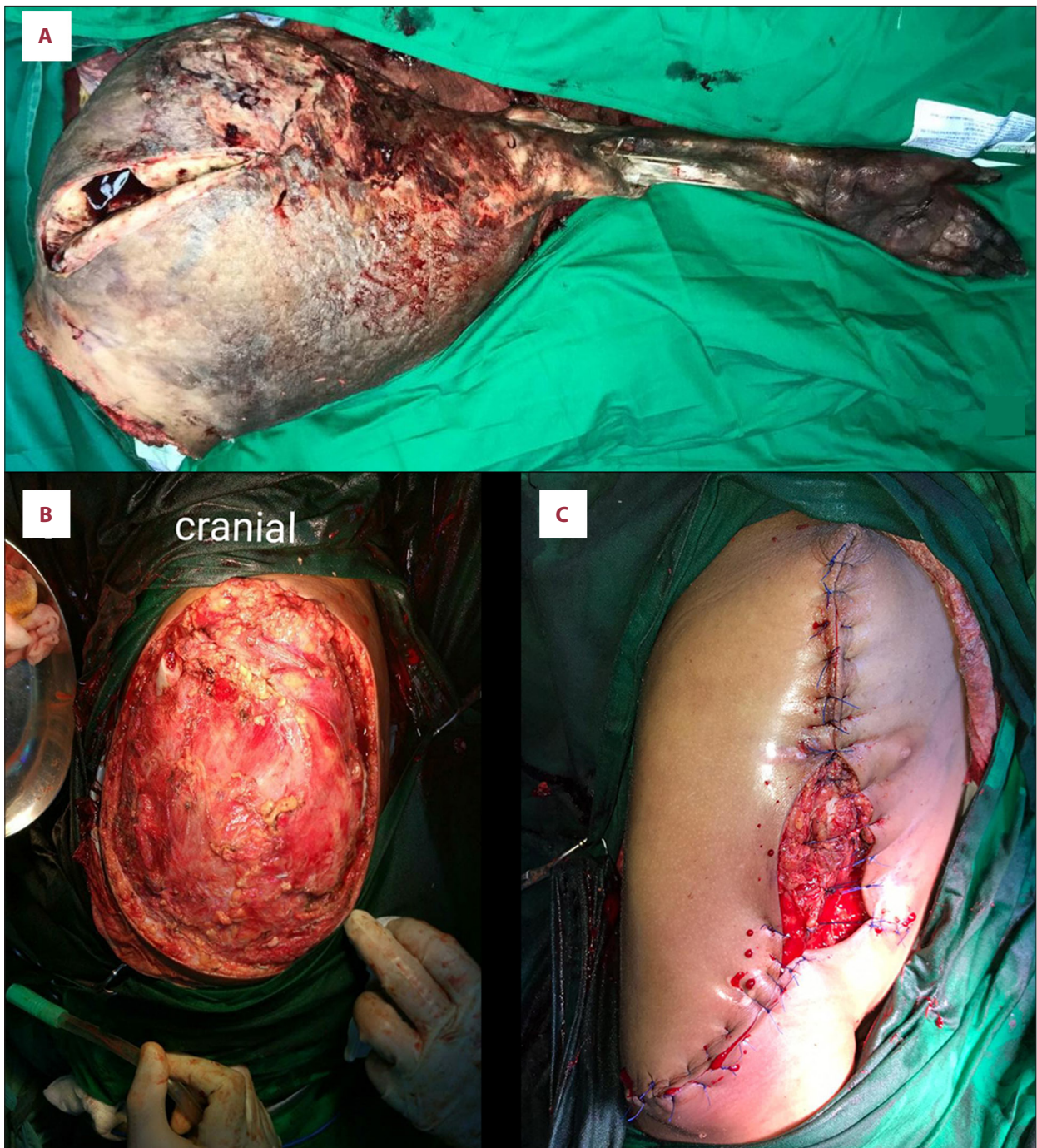


Figure 3. (A) Guillotine amputation of the non-viable left upper extremity, (B) the surgical wound, (C) the wound was closed with secondary intention.

Discussion

Humerus is one of the most common sites of osteosarcoma, comprising 10% of all cases, and 90% arising from the proximal part [1]. Unfortunately, osteosarcoma of the humerus is associated with poor prognosis [6]. In this patient, there was a significant delay of presentation to our hospital, 3 months

after the first presence of a lump. Interestingly, Goedhart et al. found that a delay of diagnosis of more than 4 months did not necessarily correlate with worse outcome [7]. However, delayed surgical control has been associated with poorer prognosis in a patient with osteosarcoma [5].



Figure 4. The patient's clinical photograph during 1-year-follow-up. The surgical wound had healed satisfactorily.

Lee et al. found that osteosarcoma development in patients during the maximum growth period (13 to 15 years old for males) had worse prognosis compared to those who were not in maximal growth phase [8]. Unfortunately, the patient in our case presented during this maximum growth period. This might explain the rapid growth, and the worsening (vascular compromise) condition which occurred during a one-week span, besides the late presentation to our hospital. The size of the tumor in this case was also not favorable. Kaste et al. concluded that both tumor depth and volume are significant predictors of overall survival and event-free survival rate among osteosarcoma cases without metastasis [9]. A nation-wide study in Japan found that tumor size larger than 16 cm was associated with worse prognosis in osteosarcoma cases [10]. Moreover, larger tumor volume was associated with increased metastasis risk [11,12].

Fortunately, there was no evidence of metastasis in our patient during initial presentation. Advanced disease with metastasis is associated with higher mortality among osteosarcoma patients [13]. However, we argue the presence of metastasis should not be solely used as a predictor because as in our case, a high-volume tumor without metastasis could cause a significant morbidity and even mortality risk.

In this patient's case, the osteosarcoma of proximal humerus caused necrosis of the hand and distal part of the arm due to

vascular compromise. Yedururi et al. reviewed the degree of cardiovascular complications among 30 osteosarcoma patients. Osteosarcoma of the humerus occurred in 15% of all cases. Vascular involvement in osteosarcoma cases has rarely been evaluated in the literatures, but careful radiologic examination is recommended to evaluate it [14]. Furthermore, perivascular tumor invasion significantly affects patient survival rate; there is, however, no standard yet on how to evaluate peritumoral vascular invasion [15]. Unfortunately, we did not perform further radiologic examination to assess the extent of vascular involvement in our patient's case. Thus, we did not evaluate the exact mechanism of this vascular compromise. We propose that the possible mechanisms of this rare vascular complication are tumor compression to the blood vessel, tumor invasion into the blood vessel, or embolization. Other than that, we suggest routine examination of vascular involvement could be done in future studies to assess its predictive value.

Several indications for FQA are high volume osteosarcoma of proximal humerus or scapula, tumor invasion of the brachial plexus, palliative therapy, failed previous limb-sparing surgery, and sarcoma-related pathological fracture [16]. Elsner et al. attempted to use FQA as a curative measure in 80% of their cases. FQA offers the advantage of wide surgical margin in hope of reducing local recurrence. FQA is also a palliative option as it may provide symptom relief and improve the patient's quality of life [17].

In our patient, a lung metastasis was detected 9 months after the surgery, and the patient died 4 months after. The overall survival rate of osteosarcoma patients treated by FQA with curative intent is 39%; and only 17% of patients treated by FQA with palliative intent survived in the span of 11 months [17]. However, in our case, we performed FQA as a life-saving procedure in order to prevent mortality from septic complications of our patient's dead limb condition. Although the patient did not survive after 13 months, the initial goal of our course of treatment was achieved. Earlier diagnosis and treatment, however, are compulsory to achieve better overall survival.

Generally, if treated early, limb salvage is still an option in cases of proximal humerus osteosarcoma. Moreover, presently there are limb salvage options such as total humeral endoprosthetic replacement which may preserve extremity function [18]. A meta-analysis concluded that limb salvage surgery and amputation have a similar rate for local recurrence; the 5-year overall survival rate, however, is higher among those treated

with limb salvage surgery [3]. Given the circumstances, limb salvage surgery was not an option in our case.

Nevertheless, to our knowledge, this is the first report of dead limb caused by proximal humerus osteosarcoma. The emergency life-saving surgery in this patient was successful in improving the patient's QOL and preventing early mortality.

Conclusions

Delayed treatment of osteosarcoma of the humerus could lead to a rare vascular complication, dead limb, such as in this case, which suggests a need to encourage early treatment planning and surgical control.

Conflict of interest

None.

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