“Whoops” fixation of proximal humerus pathological fracture ended with forequarter amputation – Case report

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Abstract

INTRODUCTION: Even with the advancement of limb salvage surgery techniques, forequarter amputation (FQA) is still used in orthopedic oncology. Even though it might pose catastrophic sequelae on the patient’s lifestyle, debilitating one’s ability to perform regular tasks, FQA is still considered as a treatment of last resort for huge fungating tumors of the upper extremity.

CASE PRESENTATION: We present a case of an 18-year-old male patient, who was diagnosed in Libya with left proximal humerus fracture after a trivial trauma and underwent open reduction and internal fixation using k-wires as it was thought to be a simple fracture. Soon after, pain and swelling progressed severely and an open biopsy confirmed a diagnosis of osteosarcoma and imaging suggested metastatic disease to the lungs for which he was started on chemoradiotherapy. He was referred to our cancer center to continue his management and due to the aggressive nature of the tumor, the patient underwent palliative forequarter amputation followed by multiple lines of chemotherapy and radiotherapy, all of which failed to halt the progression of the disease. The patient was lost to follow up due to his decision to go back to Libya.

CONCLUSION: “Whoops” surgeries are fixated upon repairing fractures without looking for the alarming signs on radiographs to exclude pathological entity. As in our case, the procedure done escalated the osteosarcoma into such a massive fungating tumor due to the violation of the osteosarcoma pseudo capsule, in which the only available option is to do a palliative forequarter amputation.

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1. Introduction

Forequarter amputation (interscapulothoracic amputation) entails the surgical removal of the entire upper extremity and shoulder girdle, including the scapula and a portion of the clavicle [1, 2]. Forequarter amputation (FQA) was first introduced in 1808 by Ralph Cuming and it was then performed after thirty years by Gaetani in Cairo, both procedures being performed in war trauma patients [1]. FQA was used for the first time in malignant disease by Dixie Crosby in 1836 followed by Paul Berger who amputated the whole upper limb of a patient with an enchondroma of the humerus [3].

The use of FQA for upper extremity malignant disease has decreased substantially due to the advancement of radio-chemotherapy and limb salvage surgical techniques and it is now considered as the treatment of last resort in cases with fungating tumor growth, ulceration, impending vascular disruption, paralysis, sensory disorders and lymphedema [4].

We present a case with a huge fungating left proximal humerus osteosarcoma that was provoked by a violation of the sarcoma entity via unplanned surgery which necessitated a palliative forequarter amputation, patient had disease progression and was lost to follow-up. Our manuscript has been reported in line with the SCARE criteria [5].
2. Case presentation

An 18-year-old male Libyan patient, who is medically free and with a history of smoking 5-pack-years, started to complain of left arm pain in March of 2017 and was not able to seek medical care due to the political instability in his country. In July 2017, the patient was admitted to an emergency unit with a fractured humerus after sustaining a minor trauma and underwent open reduction and internal fixation (Fig. 1). Shortly after, post-operative pain and swelling were noted and he had further imaging that showed pulmonary nodules and left supraclavicular lymph nodes enlargement. Tissue biopsies were done and confirmed a diagnosis of metastatic left proximal humeral osteosarcoma after which treatment was initiated thereby one cycle of cisplatin and doxorubicin and two sessions of radiotherapy (Fig. 2).

He presented to our emergency department at King Hussein Cancer Center (KHCC), in October 2017, with a massive fungating left shoulder mass measuring roughly (20 cm × 20 cm × 20 cm) complaining of an excruciating left shoulder pain extending to his forearm with limitation in hand extension. The previous surgical wound had evidence of active bleeding but it was infected and profuse foully smelling discharge was noted from the extensive fungation of the tumor that mandated frequent dressing changes daily (Fig. 3). His laboratory workup upon presentation showed hemoglobin of 11 g/dl and white blood cells count of 20.60 10⁹/µL and platelets of 434 10⁶/µL with a normal kidney and liver functions. He had a fever of 38.4 °C and was admitted for intravenous antibiotics.

As an inpatient, he had magnetic resonance imaging (MRI) of the left shoulder that showed evidence of left humeral fixation with a metallic artifact, and a massive destructive proximal left humeral lesion with an associated huge soft tissue component measuring 23 cm × 20 cm in axial dimension and 24 cm in craniocaudal dimension (Fig. 4). The soft tissue component extended medially laying on the chest wall causing secondary deformity and into the left shoulder joint with evidence of inferior subluxation, surrounded the left scapula with surrounding bony erosion, fungated into the subcutaneous layers and invaded the roots of the brachial plexus and the major axillary and brachial vessels. Whole-body positron emission tomography/computed tomography (PET/CT) scan showed multiple enlarged metastatic left axillary and left supraclavicular lymph nodes, multiple bilateral metastatic pulmonary nodules, the largest was 2.5 cm in diameter and a small right-sided pneumothorax. Evidence of thrombosis involving the left brachiocephalic and left subclavian veins with evidence of non-occlusive thrombus noted within the left internal jugular vein.

Our patient and his family were consulted at a multidisciplinary clinic (MDC) in which a review of pertinent images, explanations of the development and natural history of all findings, and the surgical option which is left forequarter amputation followed by adjuvant

Anterior-posterior simple X-ray of the left shoulder post open reduction and internal fixation with K-wires.

Fig. 1. Shoulder X-ray post open reduction and internal fixation.
chemoradiotherapy. All details regarding the complicated nature of the surgery and all possible outcomes including recurrence were explained and the patient opted to proceed with the operation which was scheduled in November of the same year.

With the patient in a semi lateral position, covering the fungating wound with abdominal pads secured by 3M Ioban dressing, the anterior limb of the utilitarian incision is used, and the pectoralis major muscle is detached from the clavicle and an extended paraclavicular incision was made, through which a clavicular osteotomy was performed through a Gigli saw to allow control of the subclavian vessels, as those were exposed and encircled with Silastic loops. Proximal subclavian artery control was obtained, which served to minimize bleeding during the majority of the procedure which was performed anteriorly. A posterior incision is then made to detach the scapula from the rhomboids, trapezius, levator scapulae, and latissimus dorsi muscles. Division of contents of the supraclavicular triangle and serratus anterior muscle. The elevation of the scapula allowed the structures of the supraclavicular triangle to be divided between hemostats. The serratus anterior muscle is divided at its origin on the chest wall. The scapula is then resected from the chest wall, and an axillary incision is made to connect the anterior and posterior incisions. The subclavian vessels and brachial plexus are ligated, and the forequarter is removed. The posterior skin flap was maintained as thick as possible to allow primary closure. After copious irrigation, insured hemostasis, closure in layers was done under negative suction (Fig. 5). The pathology report showed high-grade osteosarcoma.

The patient was then transferred to the intensive management unit for observation and his pain was controlled by left brachial plexus nerve block by continuous Bupivacaine infusion where a small catheter was inserted under the epineurium of the brachial plexus stump at the time of the surgery to decrease the nerve pain and the phantom pain, epidural catheter anesthesia, patient-controlled analgesia (PCA) morphine, and oral analgesics. He was transferred back to the surgical ward on postoperative day one then was discharged on day eleven on analgesics and anticoagulants after a smooth post-operative course (Fig. 6) and was followed up by occupational physiotherapy and pain clinic.

Three weeks after surgery, after his incisions fully healed, the patient was started on Cisplatin and Doxorubicin and underwent chest, abdomen, and pelvis CT scan after the third cycle which showed partial visualization of a soft tissue lesion posterior and inferior to the remaining part of the left clavicle and extending into the proximal part of the left internal jugular vein, highly sugge-
An image taken in the emergency department for the first presentation of the patient showing a huge fungating mass at the left shoulder with minimal amount of blood oozing from the tumor. The infected surgery site is not seen in the image.

Fig. 3. First presentation image showing the fungating mass.
With an incidence of 4/1,000,000 [20], OS poses a challenge in proper diagnosis and treatment which might result in fallacious surgical management and failure to establish neoadjuvant therapy. Based on the NCCN guidelines, limb salvaging surgery compared to amputation has the same survival and local recurrence rates, and addition of neoadjuvant and adjuvant chemotherapy, using at least 3 of the following chemotherapeutic agents: doxorubicin, cisplatin, cyclophosphamide or ifosfamide, dactinomycin, and high dose methotrexate, showed improved outcomes in patients with localized disease. The addition of muramyl tripeptide phosphatidylethanolamine (MTP-PE) to chemotherapy showed improvement in overall survival in non-metastatic disease [21].

Wang et al., from China, reported 103 patients, 23 of which were misdiagnosed and received unplanned surgical intervention, “whoops surgery”, and they reported local recurrence in 37.5% of these patients. They proposed that violation of the integrity of the tumor margins might be the culprit for such recurrence [22]. Lugowska and colleagues, reported a study with 299 patients, 46 of which underwent a surgical treatment without a proper multidisciplinary decision that resulted in a 27% survival rate as compared to other groups in the study with almost double the rate [23]. Wang et al., from Taiwan, reported 150 patients with osteosarcoma, 10.7% of them were misdiagnosed and underwent improper management, and 4 of these patients had metal implant fixation post-fracture, similar to the case we are presenting. They

Staging Left Shoulder MRI (STIR and T1 fat sat post contrast sequences). Central metallic artifact secondary to fixation (White star). Huge enhancing destructive proximal left humeral lesion with associated huge soft tissue, showing areas of hypo-intense signal in all sequences in keeping with the osteoid component. Soft tissue component fungating into the subcutaneous layers (Black star). Invasion of the brachial plexus (White arrow).

Fig. 4. Staging MRI showing the extension of the tumor.
also reported that the unplanned treatment group reported a higher incidence of lung metastasis as compared to the group that received adequate planning. However, they did not report any significant difference in 5-year survival rates between the two groups [24].

The management of pathologic fractures in children with OS has been addressed by several authors. Jackson et al. proposed that any type of management should be done after a biopsy and histological diagnosis are obtained [25]. Saraph and Linhart proposed that management of pathologic fractures should take into consideration the pain and comfort of the child, local control of the lesion, stabilization and anatomical alignment of the fracture, fracture union, and functional restoration [26]. Ruggieri et al. proposed that fractures should be initially managed by cast or external fixation to avoid the microscopic spread of the tumor [27].

4. Conclusion

Pathologic fractures occur in OS patients either spontaneously or as a result of minimal trauma, along with the characteristic radiological features of sun-burst appearance, periosteal lifting with the formation of Codman’s triangle, new bone formation in the soft tissues, the permeative pattern, and wide zone of transition of the destruction of bone should alarm the physicians to immediately refer the patient to a treating specialist center for early diagnosis and treatment as this can make limb salvage possible in a large number of patients.

Despite OS Fracture management considered very challenging and it was highly variable in the literature, OS pathological fracture is shown to be not a contraindication to limb salvage in recent studies mentioned earlier, though being that said, unplanned fixation of such fracture would hinder it from being salvageable, as this would lead to violation of the tumor pseudo-capsule and possible spread of the tumor cells into a pan—compartmental disease encasing the major neurovascular bundle with possible fungation and gross infection later—on, where it mandates a major amputation at that time as in our case.

Publishing those challenging cases would enhance a more understanding and awareness in approaching similar cases in the future.
Fig. 6. Post-operative image.

A. Staging Chest CT scan with IV contrast, axial view, lung window showing mild right-sided pneumothorax and multiple bilateral metastatic pulmonary nodules the largest seen in the left lower lobe measuring about 2.5 cm.

B. Follow up Chest CT scan with IV contrast, axial view, lung window showing significantly larger metastatic pulmonary nodule.

Fig. 7. Staging vs. follow up chest CT scan showing metastatic progression.
Declaration of Competing Interest

The authors report no declarations of interest.

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Informed consent was obtained from the patient and is available upon request.

Author contribution

Mohamad K. Abou Chaar: Writing - Original Draft, Writing - Review & Editing, Visualization.
Wafa Asha: Writing - Original Draft.
Mais Zmaily: Resources.
Hussam Haddad: Resources.
Hani Al-Najjar: Data Curation.
Samer Abdel Al: Conceptualization, Writing - Review & Editing, Supervision, Visualization.

Mohamad K. Abou Chaar and Wafa Asha performed the literature search, and wrote the manuscript under guidance and direct supervision by Samer Abdel Al. Mais Zmaily reviewed the case from a radiological point of view and provided adequate imaging material, Hussam Haddad obtained and reviewed the pathology samples and provided the images. Hani Al-Najjar obtained written consent. Samer Abdel Al oversaw the manuscript’s inception, guided the literature search, counseled the patient and wrote the operative part of the surgeries he performed. All authors read and approved the final manuscript.

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