

**FOREQUARTER AMPUTATION FOR PRIMARY MALIGNANT TUMOURS OF PROXIMAL UPPER LIMB:
A REPORT OF THREE CASES.**

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Abstract

Two men and one boy from Iraq with advanced primary malignant tumors of the proximal upper limbs (fibrosarcoma, ulcerating basal-cell carcinoma and osteosarcoma) who were cured by forequarter amputation (FQA) are reported herein. The presenting signs and symptoms were huge masses in the axilla, supraclavicular region and shoulder, with severe pain and recurrent bleeding. Due to the advanced stage of these tumors with bone and vascular involvement (in 2 cases) and neurovascular involvement (in one case), limb salvage was not possible. Forequarter amputation was successfully performed by vascular and orthopedic surgeons with direct skin closure and uneventful postoperative course apart from simple wound infection in one case. The operations deemed to be curative as there were no demonstrated metastases at the time of surgery. Two patients were lost for follow up while the third survived for at least 5 months. Surgical removal of the bones and soft tissue of the upper limb including the scapula and clavicle (FQA) is an old procedure that was done for the first time more than 2 centuries ago for major trauma and then became the preferred approach for proximal upper limb malignant growths. As limb-sparing procedures evolved, FQA became less popular. Despite the significant disfigurement imposed by the procedure and the frequent occurrence of phantom limb pain, FQA still has a vital role in management of selected advanced proximal upper limb malignant tumors for which limb salvage is not possible. Hence, surgeons need to be familiar with this uncommon yet important operation.

Key Words Forequarter, shoulder, interscapulothoracic, amputation, upper limb, malignant tumors.

Background

Forequarter amputation (FQA) also known as shoulder girdle or interscapulothoracic amputation refers to surgical removal of the bones and soft tissue of the upper extremities including the scapula and clavicle ^{[1]-[5]}. Ralph Cuming performed this operation for the first time in 1808 ^[1] while Dixie Crosby was the first to use this procedure for a case of osteosarcoma in 1836 ^[1]. Severe traumatic injuries of the upper limbs were the original indications. Currently, malignant tumours of the arm, axilla, shoulder and scapula represent the most frequent indication ^{[1][2]}. It can be done as a curative or palliative procedure ^{[3]-[5]}. As limb sparing surgery has evolved, FQA became less popular ^{[3] [4]}. However, this procedure still has a definite role in the management of selected patients with malignant tumours of upper extremities particularly the advanced cases ^[2] or recurrent tumours after conservative treatment ^[1]. The disfiguring result of this operation is significant and there are many complications especially phantom limb pain ^{[1][3]}. This explains the reluctance of most surgeons and patients to use this procedure. In certain patients presenting with unresectable tumors of the upper limb that have intractable pain, bleeding, sepsis or useless limbs, FQA remains a valuable option and surgeons need to be familiar with this uncommon surgery ^{[1] [2]}. Herein, three cases of unresectable primary malignant tumours of upper limbs cured by FQA are presented with review of relevant literature.

Case 1: Fibrosarcoma of Right Shoulder Girdle

A man of 55 was admitted to Basra Teaching Hospital (BTH) on September 16, 2001 because of a big right supra-clavicular mass of 3-year duration associated with severe shoulder pain radiating down the arm. The mass was diagnosed as a non-epithelial tumour by fine needle aspiration (FNA) done 3 months earlier. Physical examination revealed a solid mass bulging above the right clavicle and fixed to deeper structures, with marked varicose superficial veins on anterior chest and right upper arm, palpable right radial pulse and a normal chest examination (**Figure 1**). Plain chest radiogram (**Figure 2**) as well as complete blood picture (CBP) were normal with an erythrocyte sedimentation rate (ESR) of 40 mm/hr. MRI scan showed a huge well-defined encapsulated mass extending up to the neck and down to the axilla without intra-thoracic extension (**Figure 3a and 3b**). The high vascularity of the tumour and its relation to the subclavian artery were evident on MR Arteriography (**Figure 4**). The patient underwent 2 operations (**Figure 5**); unfortunately, the first one was not well planned. A direct incision over the mass was followed by severe bleeding from highly friable vascular tissue. The wound was therefore, closed over packs and the biopsy result was undifferentiated sarcoma. The second surgical intervention was carried out 1 week later after a discussion with an orthopedic surgeon and informing the patient about the contemplated radical resection via a FQA. Proximal control of the innominate vessels was achieved by median sternotomy with supraclavicular extension and clavicular resection. The major veins were greatly engorged by mass compression while the subclavian artery was coursing through the tumour. After tedious dissection, subclavian vessels were isolated and ligated proximal to the mass. Forequarter amputation was then completed in a lateral position. The mass was 23 x 9 x 8 cm in size involving the head of the humerus. Good hemostasis was followed by placing mediastinal, right pleural and submuscular drains and primary wound closure. The postoperative course was uneventful. Biopsy result was fibrosarcoma. The patient received postoperative radiotherapy and was well for 5 months following the FQA (**Figure 6**).

Case 2: Basal Cell Carcinoma of Axilla

A man of 50, a known case of malignant ulcer of left axilla for 5 years presented with recurrent bleeding and loss of sensation over the forearm. He was referred to Sulaimaniyah Teaching Hospital (STH) in August 2004. Left FQA was performed in collaboration with an orthopedic surgeon. The biopsy result revealed features of basal cell carcinoma (solid and adenoid types) with invasion of the wall of the axillary artery as well as perineural invasion. The patient had a smooth postoperative course (**Figure 7**).

Case 3: Osteosarcoma of Left Shoulder

A boy of 9 presented with a huge painful swelling of left shoulder of 2 years duration diagnosed by a previous open biopsy as osteosarcoma (**Figure 8**). The old scar, as well as the dilated superficial veins, was visible and the mass was hot and tender to feel. Plain chest radiogram showed a big soft tissue swelling around the left shoulder with involvement of the humerus (**Figure 9**). The patient was treated by left FQA (**Figure 10**) with a smooth postoperative course.

Discussion

The shoulder girdle is the third most common site for malignant bone and soft-tissue tumors ^[4]. In a review of more than 46,000 cases by the National Cancer Centre, tumors around the shoulder girdle involving scapula, distal clavicle and proximal humerus constituted 12% of all primary malignant bone tumors treated in Japan since 1972 ^[4].

When faced with a malignant tumor of the proximal upper limb, the goal of surgery should be the complete removal of the tumor and preserving the limb. Unfortunately, this goal is not always achievable. Limb-salvage procedures may not be feasible in advanced tumors involving vessels and/or nerves. In such circumstances, removal of the tumor necessitates limb sacrifice (major amputation). Fortunately, this option has become infrequently required with the advent of new surgical techniques that permit limb salvage. Daigeler et al confirmed this fact by studying 1597 cases of limb sarcoma in which proximal major limb amputation was necessary in just 37 cases (2.3%) ^[3].

Previous Indications

Initially, FQA was used to treat traumatic injuries of the upper limb ^[1]. Later it became the standard of care for treating and curing patients with bone and soft tissue sarcomas of the limbs ^[1]. However, this operation is a major undertaking resulting in the loss of a limb, which is an important part of the body schema. Most patients and surgeons dislike this operation due to its significant disfigurement and the frequent occurrence of phantom limb pain (reported in 87% of patients) ^[3]. This pain, beside other factors, puts a strain on the patients; some of them complain of sadness and may even attempt suicide ^[3].

Is There a Way Out?

It is logical that surgeons began to search for new surgical techniques that permit removal of the tumor and preserve the upper limb at the same time. This led to the concept of limb-salvage surgery.

Limb salvage for tumors of the shoulder girdle was first described by Linberg in 1927 [4]. Taira et al treated 15 patients with malignant bone and soft-tissue tumors of the shoulder girdle using limb salvage operations involving resection of the proximal humerus, scapula and clavicle [4]. Oncological evaluation of these patients revealed that in 11 patients with adequate surgical margins, six were continuously disease free; two had no evidence of disease; and three patients with marginal surgical margins died. The authors suggest that only where adequate surgical margins can be obtained should limb-salvage be performed and recommend FQA for patients with complicated neurovascular involvement [4]. An inadequate resection makes the patient prone to the risk of local recurrence and subsequent fatal distant metastasis [4].

What to do when limb salvage is not possible?

The answer is straightforward! FQA would remove the entire limb including the tumor providing mostly a palliative solution for those with metastatic disease while it gives a chance of cure in the absence of metastases. An example from the literature is the following case “Marinescu et al described the case of a 33 year old lady with giant synovial sarcoma involving left axilla, arm and scapula with vascular and neural involvement treated by FQA as a life saving procedure” [2]. In their opinion, FQA represents a mutilating surgical intervention, but can give a chance for desperate cases with extensive cancers [2].

To clarify, we reviewed the literature to define the current indications for the procedure. “Advanced tumors, adjacent to crucial structures and close to the trunk, currently justify a sacrifice to an extremity. In those cases with excessive fungating tumor growth, ulceration, impending vascular disruption, intractable pain, paralysis, sensory disorders, lymphatic edema and a largely useless extremity, a proximal major amputation as a last resort may improve quality of life in an often palliative situation” [3] and “When the brachial plexus and sub-clavian vessels are involved by the tumor, FQA is also indicated” [4]. Moreover, FQA has a role in the treatment of recurrent breast cancer [5]; to date 23 such cases have been described in the literature [5].

Technique

The technical aspects of this major surgical intervention deserve special attention, particularly related to the team approach and wound closure. The patients reported herein were managed by a team of an oncologist, vascular and orthopedic surgeons. Proximal control of subclavian vessels is a safe precaution to minimize blood loss. Tsai et al recommend a dedicated multidisciplinary team to reduce complications and achieve a successful rehabilitation [5]. A plastic-reconstructive surgeon may be required if skin graft or myo-cutaneous flaps are needed to achieve wound closure after shoulder amputation [5]. Primary direct skin closure could be achieved in our cases although the first patient was in need of a small primary skin graft. This is due to the fact that skin is usually sufficient when the shoulder is amputated unless the primary tumor has already involved a big area of skin. Soft tissue reconstruction is required for large defects that cannot achieve primary closure [5]. Many methods of soft tissue reconstruction are reported in the literature such as split skin graft, fasciocutaneous deltoid flaps and others [5]. Daigeler et al and others [Pant et al, Sugarbaker & Chretien cited in 3] could achieve sufficient closure with local flaps [3]. Gibbons et al showed the usefulness of the latissimus dorsi musculocutaneous flap to cover preserved tissue [Gibbons et al cited in

4]. Taira et al found soft-tissue reconstruction of great value to avoid skin necrosis induced by tensile skin closure as well as hematoma formation within the large dead space ^[4].

Quality of Life

In regard to quality of life following the procedure, there are different opinions. Wittig et al [cited in 3] reported pain relief and improvement of quality of life after proximal major amputations. Likewise, Daigeler et al reported that most of their patients have accepted the aesthetic outcome of the operation and felt a significant improvement in their quality of life ^[3]. The greatest benefit was observed among those with low preoperative life quality and high pain levels ^[3]. In contrast, Taira et al believed that the functional result after forequarter amputation was extremely poor, even if better results were achieved oncologically ^[4]. Some patients (15.6%) considered their quality of life worse than before the operation ^[3]. Thirty two percent of the amputees harbor suicidal ideas and 65% suffer from sadness ^[3] probably related to phantom limb pain beside other factors [Shukla et al and Wright et al cited in 3].

Survival

A prospective randomized trial from the National Cancer Institute concluded that the rate of disease-free survival and overall survival of limb salvage versus amputation were comparable ^[4]. Most patients treated with palliative forequarter amputation survive less than two years after surgery ^[5]. In a series of palliative forequarter amputations, the patients' post-operative survival ranged from 3 to 12 months [Wittig et al cited in 3]. However, when this operation is performed for patients with solitary lesions without associated co-morbidities, then it can provide the patient symptomatic relief and even a chance of cure ^[5]. In such circumstances, longer survival (for at least 3 years) has been reported ^[5]. Our first patient remained well for 5 months after which he was lost for follow up while the fate of the other 2 patients is unknown. Therefore, it is not possible to comment on potential complications such as phantom limb pain, local tumor recurrence or distant metastases.

Conclusion

Although the number of patients in this article is too small to draw meaningful conclusions, we do agree with authors who consider the operation safe and reliable for certain patients with upper limb malignant tumors in whom limb salvage is not possible.

Conflict of Interest

None declared.

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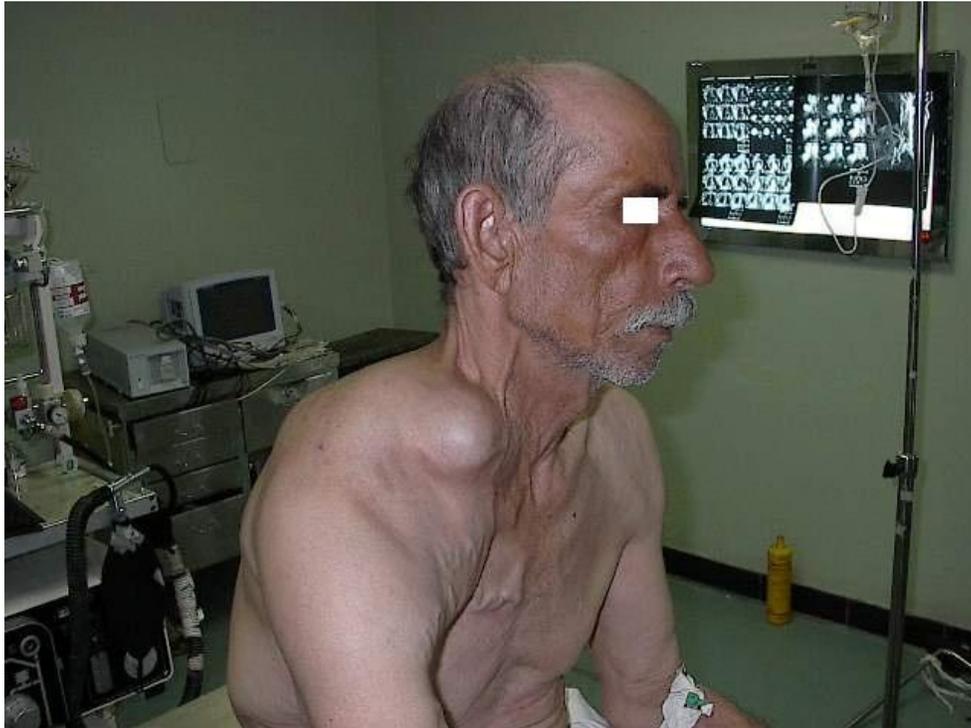


Figure 1: Huge Fibrosarcoma

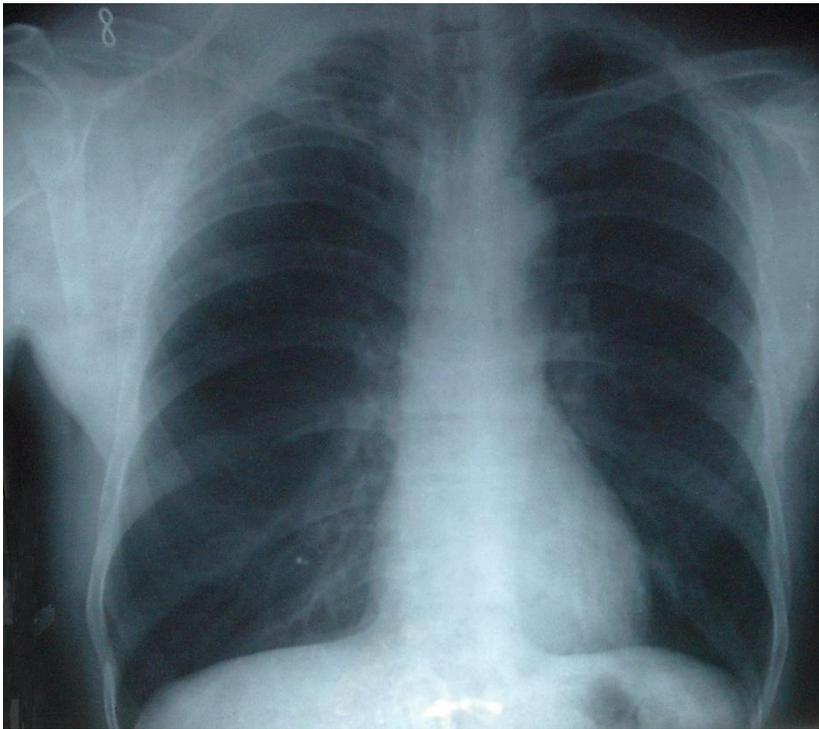


Figure 2: Chest Radiograph of First Patient

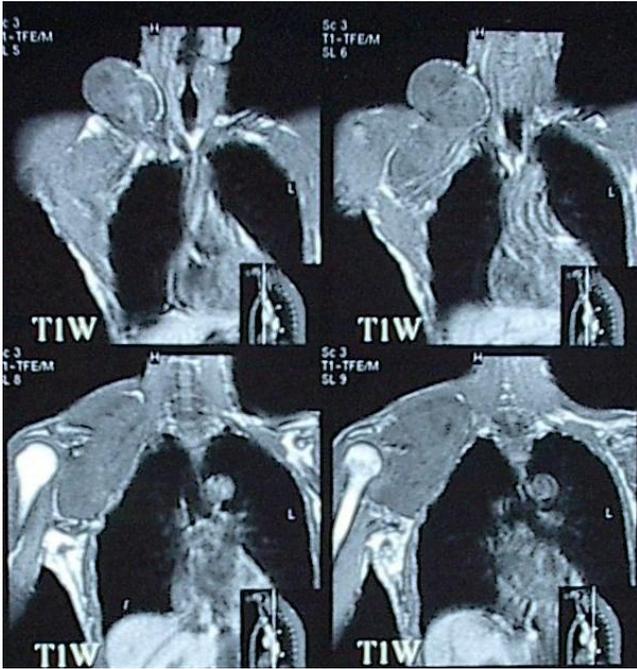


Figure 3-a: MRI of First Patient without Contrast

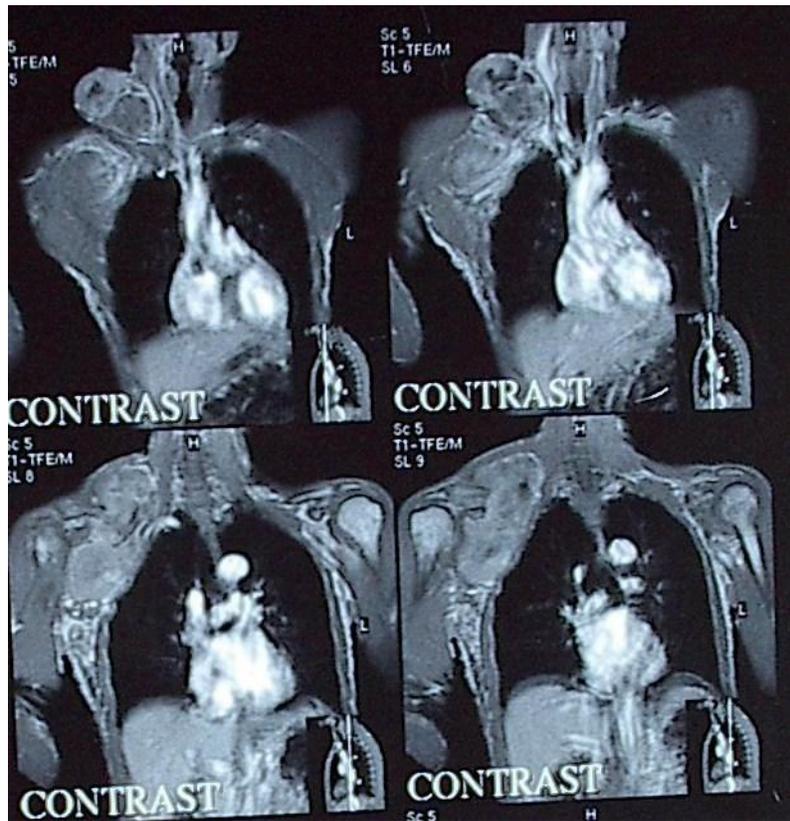


Figure 3- b: MRI of First Patient with Contrast



Figure 4: MRA of First Patient



Figure 5: First Patient just Anaesthetized



Figure 6: R Forequarter Amputation for Fibrosarcoma



Figure 7: L Forequarter Amputation for Axillary Basal-Cell Carcinoma



Figure 8: Huge Osteosarcoma of L Shoulder



Figure 9: Plain X-ray of Third Patient



Figure 10: L Forequarter Amputation for Osteosarcoma