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Scapulectomy for Fibromatosis – Ocological and Functional Outcomes

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ABSTRACT

Aggressive fibromatosis is a locally invasive tumor that does not metastasize. Only few cases of scapulectomy for fibromatosis of scapular muscles are reported in the world literature. We present two consecutive cases of scapulectomy done for fibromatosis of periscapular muscles at the Department of Surgical Oncology in our centre between 2014 and 2019. One patient underwent subtotal scapulectomy by posterior approach and the other patient underwent scapulectomy below spine of scapula by combined anterior and posterior approach. The follow-up period of both the patients was 5 years. One patient developed recurrence after 9 months of surgery dealt by re-resection followed by adjuvant radiation. The modified Musculoskeletal Tumor Society (MSTS) score for scapular surgery was 20 in the first patient and 22 in the second patient. Both of them are able to carry out normal regular activities with the compensation from the opposite upper limb. Scapulectomy for symptomatic fibromatosis of the scapular muscles gives acceptable functional results and good disease-free interval. Scapulectomy should be considered as part of resection for better tumor clearance in fibromatosis involving muscles around the scapula. Further studies are needed to evaluate and validate the need for radical surgery in this setting.

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INTRODUCTION

Fibromatosis is caused by the proliferation of differentiated fibroblasts, and aggressive fibromatosis (AF) is a subtype of fibromatosis known as desmoid tumor or desmoid-type fibromatosis (DF) which is a monoclonal fibroblastic proliferative disease (Foà *et al.*, 2014). Aggressive fibromatosis is a locally invasive tumor that does not metastasize (Goldblum and Fletcher, 2013). Scapulectomy is a good option for treating patients with tumors of the scapula (Veth *et al.*, 2003). Scapulectomy achieves wide margins of tumor resection with-

out the need for amputation. Scapulectomy results in excellent local tumor control.Only few cases of scapulectomy for fibromatosis of scapular muscles are reported in the world literature (Gibbons *et al.*, 1998; Bhaskar *et al.*, 1994). Here we describe our cases of fibromatosis of scapular muscles who underwent scapulectomy and are surviving without disease beyond 5 years.

PATIENTS AND METHODS

We present two consecutive cases of scapulectomy done for fibromatosis of periscapular muscles at the Department of Surgical Oncology in our centre between 2014 and 2019. The procedures were done after getting informed written consent and in accordance to the institutional ethics policy.

Case 1

A 41-year-old gentleman, known case of congenital scoliosis, presented with swelling in the right scapular region of four years duration with recent onset of pain and limitation of shoulder movements. Physical examination revealed a 15*10 cm swelling over the right scapular region fixed to the scapula. All ranges of shoulder movements were restricted. Chest X-ray was normal. CT Chest with contrast revealed a 4.2*9.2*16 cm lesion involving the right Teres major, posterior intercostal muscles and subscapularis. A small 2.0*2.7 cm lytic lesion was present in the proximal body of the scapula. Trucut biopsy was suggestive of fibromatosis. Immunohistochemistry was positive for Beta-Catenin, Vimentin and negative for S-100 and Smooth Muscle Actin (SMA). In view of the pain and limitation of shoulder movements with CT chest showing a lytic lesion in proximal body of scapula, the patient underwent subtotal scapulectomy. Patient in semilateral position, arm was draped freely. A curvilinear posterior incision was made along the medial border of the scapula with lateral extension at the suprascapular region towards the shoulder joint and flaps raised. Trapezius muscle attachments were divided. Lattismus dorsi was divided and the tip of scapula was elevated. Medially the rhomboideus muscles attachment was released. Laterally the teres major was divided with adequate tumor free margins. Levator scapulae muscle was divided. Subscapularis muscle was divided with adequate margins. The axillary vessels and nerves were identified from posterior aspect and safeguarded. The scapula with tumor was defined all around and osteotomy done proximal to the neck of the scapula preserving the glenohumeral joint [Figure 1]. Subtotal scapulectomy was done. The divided edges of the muscles were tenodesed around the shoulder joint. Blood loss was 600 ml and one unit Packed red cell was transfused intraoperatively. Duration of surgery was 4 hours. Wound closed with suction drains. There was no intraoperative or postoperative complications. Postoperative histopathology confirmed the lesion to be fibromatosis. Postoperatively the patient was maintained in sling for 4-6 weeks and shoulder movements were gradually started by the rehabilitation team. Patient received adjuvant Tamoxifen for 5 years.

Case 2

A 39-year-old gentleman, a known Rheumatic heart disease patient with past history of recovered cerebrovascular event, presented with pain and swelling in the left axilla of 6 months duration. Open biopsy of the lesion done outside was suggestive of fibromatosis. MRI shoulder showed a heterointense lesion in the axilla abutting the axillary vessels and extending posteriorly onto the subscapularis muscle. Patient underwent enbloc resection the tumor with partial scapulectomy by combined anterior and posterior approach preserving the axillary vessels and nerves with a Utilitarian incision. Patient in semilateral position with arms freely draped, anterior approach was done first with extension of the utilitarian incision along the medial border of the arm. Axillary vessels and nerves abutting the tumor were dissected and safeguarded [Figure 2]. Then posteriorly the incision was extended along the lateral border of scapula. Flaps were raised. Muscular attachments were divided similar to the previous patient. Osteotomy was done just below the spine of the scapula in this patient [Figure 3]. Enbloc excision of the axillary and scapular component of the mass along with the infraspinous portion of scapula including the biopsy scar was done. Scapula above the spine and Glenohumeral joint was preserved. Blood loss was 400 ml and no transfusion was given intraoperatively. The duration of the surgery was 4 hours 30 min. Wound closed in layers with suction drains. There was no intraoperative complications or postoperative morbidity. Patient developed a recurrence in the axilla over the axillary vessels without infiltration at a later date. Patient underwent re-excision of the lesion by anterior approach. Postoperative histopathology of both the procedures were suggestive of fibromatosis with close margins on the vascular side of the lesion. Clips were placed at the margins as markers facilitating post-operative radiotherapy. Patient received adjuvant radiotherapy in view of the recurrence. T.Tamoxifen was started and continued for 5 years.

Patients were followed up every 3 months for first

three years and every 6 months subsequently.

RESULTS

The follow-up period of both the patients was 5 years. One patient developed recurrence after 9 months of surgery dealt by re-resection followed by adjuvant radiation. Shoulder abduction was up to 15 to 20 degrees in first patient [Figure 4] and up to 60 degrees in the next patient [Figure 5]. Shoulder flexion was 60 degrees in both patients and extension was 30 degrees. The modified MSTS score for scapular surgery was 20 in the first patient and 22 in the second patient. Both of them were able to carry out normal regular activities with the compensation from the opposite upper limb. Both of them had good emotional acceptance postoperatively. The first patient was able to ride his vehicle for delivery of goods and second patient carried on with his printing press activities. The professional activities were not interrupted in both the patients. Both the patients are disease free and alive for the past 5 years.

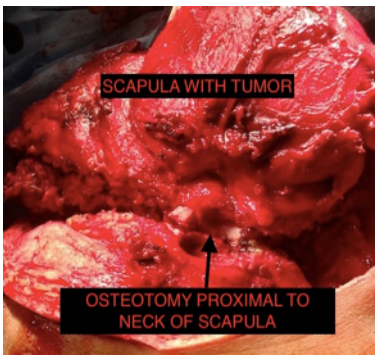

Figure 1: The scapula with tumor defined all around and osteotomy done proximal to the neck of the scapula

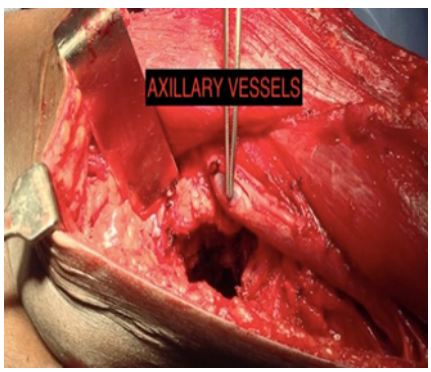

Figure 2: Post resection surgical bed showing the axillary vessels and nerves anteriorly

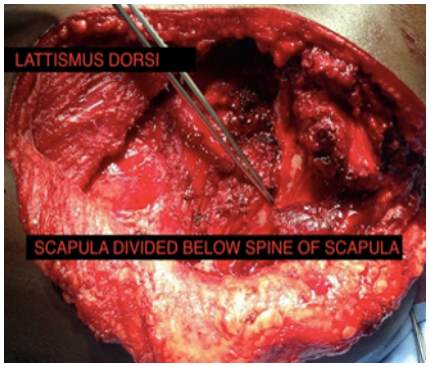

Figure 3: Post resection surgical bed showing the cut edge of the scapula just below the spine of the scapula

Figure 4: Postoperative image of the first patient with shoulder in abduction (posterior view)

Figure 5: Postoperative image of the second patient with shoulder in abduction (Anterior view)

DISCUSSION

Deep fibromatosis (DF) most commonly arises between the age groups between 15 and 60 years, with a 2 to 3-fold female predominance (Camargo *et al.*, 2010; Lev *et al.*, 2007). It accounts for 3% of all soft tissue tumors and is composed of spindle cells and collagen fibrils (Hajjar *et al.*, 2017). Most frequent sites of occurrence include the abdomen, shoulders and limbs. It lacks metastatic potential and has a high propensity for recurrence (Lee *et al.*,

2012; Trufero *et al.*, 2017).

The etiopathogenesis is not yet clear and is believed to be multifactorial. It may be sporadic or familial in nature. Some of the etiopathogenesis implicated include trauma, pregnancy and use of oral contraceptives (Trufero *et al.*, 2017; Fiore *et al.*, 2016). Pathologically, it is characterized by proliferation of uniform spindle cells resembling myofibroblasts, in the background of abundant collagenous stroma and vascular network (Muller *et al.*, 1996). On immunohistochemistry, nuclear B-catenin, vimentin, cyclooxygenase 2, tyrosine kinase PDGFRb, androgen receptor and estrogen receptor beta stains were positive in DF, but desmin, S-100, h-caldesmon, CD34 and c-KIT were negative in a study (Kotiligam *et al.*, 2008). B-Catenin and Vimentin were positive in both our patients.

The clinical presentation in DF is influenced by the tumor location and is highly variable. Most lesions present as painless mass in the abdominal and extra-abdominal locations. However, lesions adjacent to neurovascular structures and larger lesions may be associated with pain and functional impairment (Chew *et al.*, 2004). The imaging modality very useful for imaging DF is MRI, especially for lesions occurring in the extremities, head and neck, abdominal and chest wall (Walker *et al.*, 2012). Preferred approach to the management of patients with DF is surgical treatment. Radical surgery has been reported to be the most important treatment to avoid disease recurrence (Hajjar *et al.*, 2017). Both our patients were symptomatic with pain and functional impairment and hence surgery was offered. One patient with tumor abutting axillary vessels developed recurrence and underwent re-resection.

Asymptomatic DF may be managed with "wait and see" approach when newly diagnosed. Studies have shown that patients with asymptomatic DF may be initially managed with active surveillance considering the significant percentage of these tumors showing long term stability without treatment (Fiore *et al.*, 2009; Salas *et al.*, 2011). Both our patients were not candidates for 'Wait and watch policy' in view of progressive increase in size and pain with functional impairment.

Radiotherapy may be useful for unresectable or recurrent DF and in patients at high risk for surgery (Santti *et al.*, 2017). Patients with positive surgical margins in DF may be considered for adjuvant radiotherapy following surgery (Karabulut *et al.*, 2013). A recent meta-analysis showed that combination of surgery and radiotherapy had a lower local failure rate compared to surgery alone (Wood *et al.*, 2013). Our Patient with recurrent disease underwent adjuvant radiotherapy and is disease free till date.

Systemic therapy also plays an important role in the management of DF. The therapeutic agents including anti-estrogenic drugs (tamoxifen and toremifene), non-steroidal anti-inflammatory drugs (meloxicam, indomethacin, sulindac and celecoxib), cytotoxic chemotherapy (doxorubicin, methotrexate and vinblastine) and tyrosine kinase inhibitors (imatinib, sunitinib, pazopanib, sorafenib, sirolimus) have been shown to be useful for achieving disease stability in DF (Fiore *et al.*, 2016; Yao *et al.*, 2014). Both our patients received adjuvant Tamoxifen for 5 years.

Malawer's classification of shoulder girdle resections including scapulectomy comprises of six categories including intra-articular proximal humeral resection, partial scapular resection, intraarticular total scapulectomy, extra-articular total scapulectomy and humeral head resection (classical Tikhoff–Linberg resection), extra-articular humeral and glenoid resection, and extra-articular humeral and total scapular resection (Malawer, 1991). Japanese Musculoskeletal Oncology Group (JMOG) have classified scapulectomy into five categories in terms of resection area as follows; 1. Total scapulectomy (include extra-articular resection), 2 Glenoid preserved, 3. Acromion preserved, 4. Both acromion and glenoid preserved and 5. Resection of the lower half of the scapula (Hayashi *et al.*, 2014). Both the patients underwent Type II shoulder resection (Partial Scapulectomy) as per the Malawer's classification. As per the JMOG classification, one patient underwent Type IV resection and the next patient type V resection.

Reconstruction of the rotatory muscles of the shoulder be attempted, though not always achieved. The scapula is the key link to arm stability because of its attachment to the chest wall by the insertion of the periscapular muscles. Reconstruction of this linkage is vital to obtain a satisfactory upper extremity and a stable shoulder girdle which are essential for normal elbow and hand function. The key surgical steps required for soft tissue reconstruction following limb salvage of the scapular have been described in detail by some authors (Öztürk *et al.*, 2019). The deltoid, trapezius, rhomboids and latissimus should be retained. Secondary adhesion of the serratus anterior muscle to the posteriorly applied rhomboids and latissimus dorsi forms a muscle envelope that provides good scapular stability. Soft tissue reconstruction by tenodesis of remaining muscles around the shoulder joint was done in our cases.

Limb-sparing surgery procedures can be done for

malignant tumors of the shoulder girdle and soft tissue. Reconstructive procedures and reconstructive methods such as prosthetic replacement, autoallograft, and soft tissue reconstructions should be specified in each case. These resection and reconstruction methods are reliable and applicable procedures for local tumor control, pain control, and functional outcomes (Kiss *et al.*, 2007). Preserving the glenoid or the acromion lead to better function compared to total scapulectomy (Hayashi *et al.*, 2014). The amount of remaining bone influenced functional outcome. Resection of 80% of the scapula has a modest effect on function and complete or partial preservation of the glenohumeral joint enables excellent functional results. If the deltoid is reattached to the scapular remnant and trapezius, the rotator cuff can be removed without resulting in a severe functional deficit (Gibbons *et al.*, 1998). Patients can compensate extremely well by using the preserved joints and the contralateral upper limb; therefore, patient satisfaction does not rely on shoulder function alone (Vahanan *et al.*, 2007). In a study, it was showed that retention of the glenohumeral articulation gave superior functional results in a study on fifteen patients who underwent total scapulectomy compared to a group who had their glenoid retained (Veth *et al.*, 2003). Gleno humeral joint was preserved in both the patients and had reasonably good functional result facilitating them to have reasonable shoulder movements with compensation from the other upper limb to perform their professional work.

Apart from achieving good oncological and functional results, Limb-saving surgery should achieve good psychological outcomes. As treatment tend to produce long-term side-effects, assessment of quality of life is important (Vitale and Jimenez, 2009). Studies assessing the quality of life in survivors after scapulectomy are few as most studies concentrate on oncological and functional results (Vahanan *et al.*, 2007). Involvement of rehabilitation therapists is integral to attaining acceptable emotional reactions to scapulectomy. Patients who have undergone total scapulectomy generally have a decreased shoulder range of motion (ROM), especially abduction. Elbow and hand movements are usually retained. A number of patients have gait abnormalities in leaning and walking with higher risk of falling (Vitale and Jimenez, 2009). Patients should consult rehabilitation therapists for a personalized rehabilitation plan. The usual recommendations include a sling immobilization for 4 weeks and ongoing physical therapy. Our patients were educated by our rehabilitation team providing them the physical therapy and emotional support. Both the patients have achieved excellent social and psychological well-being at the end of years.

Only few cases of Scapulectomy as part of surgical clearance for fibromatosis has been described in the world literature. Subtotal scapulectomy for one case of aggressive fibromatosis was shown in a study on functional outcomes after subtotal scapulectomy (Gibbons *et al.*, 1998). Excision of large myofascial fibromatosis involving the shoulder girdle in a 11-year old girl in which the tumor was shaved off lateral border of scapula was described in a study (Bhaskar *et al.*, 1994). Both our patients underwent scapulectomy for fibromatosis of periscapular muscles for better tumor clearance. Scapulectomy should be considered for fibromatosis of periscapular muscles as it provides good disease-free survival consequent to the radical procedure, though it needs to be validated by further studies with significant number of patients.

CONCLUSION

Scapulectomy for symptomatic fibromatosis of the scapular muscles gives acceptable functional results and good disease-free interval. Scapulectomy should be considered as part of resection for better tumor clearance in fibromatosis involving muscles around the scapula. Further studies are needed to evaluate and validate the need for radical surgery in this setting.

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Conflict of Interest

Authors declare no conflict of interest.

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