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Case Report

A combined, supraclavicular, infraclavicular, transaxillary, and posterior subscapular approaches for *en bloc* resection of giant myxofibrosarcoma

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ABSTRACT

Background: Myxofibrosarcoma (MFS) is a rare and locally infiltrative tumor that commonly occurs in extremities in older adults; however, truncal and head and neck cases have been reported. They are characterized by multinodular growth, incomplete fibrous septa, and myxoid stroma. Surgical resection is the mainstay of treatment.

Case Description: The authors report a case of a combined, supraclavicular, infraclavicular, transaxillary, and posterior subscapular approaches for resection of giant MFS.

Conclusion: The anatomical complexity and rarity of tumors involving the brachial plexus impose many challenges onto surgeons performing surgical resections. Treatment choices and surgical outcomes rely heavily on meticulous multidisciplinary planning, anatomical knowledge, careful dissection, and extent of resection. This case is unique in utilizing four different approaches to the brachial plexus to resect one tumor.

Keywords: Brachial plexus, Infraclavicular, Myxofibrosarcoma, Posterior subscapular, Soft-tissue sarcoma, Supraclavicular, Transaxillary

INTRODUCTION

Myxofibrosarcomas (MFSs) are malignant tumors of myofibroblasts. The World Health Organization defines MFS as a malignant connective tissue neoplasia of fibroblastic origin.[11] They are low- or high-grade sarcomas that arise in soft tissue or bone, typically presenting as a painless slow-growing mass.[9] They are infiltrative tumors, usually in superficial soft tissue and are tumors of older adults.[7] They primarily affect the extremities and limb girdles and are very rarely in the head and neck.[14] MFS is characterized by a high risk of local recurrence that is thought to be related to an infiltrative growth pattern with tumor spread along vascular and fascial planes.[4,13] It has a range of microscopic appearances; however, all display at least focal nuclear pleomorphism.[8] High-quality magnetic resonance imaging (MRI) is critical to preoperative planning. Surgery is the mainstay of treatment and adjuvant radiotherapy is typically indicated to improve outcome.[2,3]

CASE REPORT

The patient was a 74-year-old former smoker with no other significant medical history who presented with a palpable mass in his right axilla and right neck. MRI of his chest was performed and demonstrated a large infiltrative solid heterogeneous mass centered in deep right lateral chest wall fat with infiltration into the right lower neck region [Figures 1a-e]. The mass measured up to 20 × 17 × 5 cm at the time of initial imaging. The divisions and cords of the right brachial plexus approached this large lesion at the level of the mid distal clavicle, although without evidence of invasion or encapsulation [Figures 1d and e]. There was no abnormal enhancement of the brachial plexus. The right axillary vein was mildly effaced as it coursed at the superior aspect of this lesion. Radiographically, it was felt to most likely represent a sarcomatous lesion such as a myxoid sarcoma. The patient underwent multidisciplinary evaluation, appropriate staging studies, and percutaneous biopsy confirming a high-grade spindle cell sarcoma most suspicious for MFS. His treatment plan was established in collaboration with radiation oncology. Given the tumor size and proximity to the brachial plexus and axillary vessels, the patient initially received neoadjuvant radiation therapy through 50Gy in 25 fractions of intensity-modulated radiation therapy. Restaging demonstrated again a local tumor with no evidence of regional or distant spread and no direct invasion of important structures. Given the complex anatomy, a multidisciplinary surgical team was established to ensure adequate intraoperative expertise. Preoperatively, no neurologic deficits were noted.

After consent was obtained, approximately 2 weeks after radiation therapy, the patient was brought to the operating room and was positioned in a left lateral decubitus manner with free mobility of the arm to facilitate various exposures during different phases of the operation [Figures 2a and b]. The areas prepped included: The right neck anteriorly and posteriorly, from the ear to clavicle; the circumferential right arm; and the ipsilateral chest wall to midline anteriorly and posteriorly. The right arm was placed in a stockinette. Neuromonitoring was initiated and during the procedure, the physiologic status of the brachial plexus was monitored by means of spontaneous and evoked motor potentials. Before incision was made, needle electrodes were placed in the deltoid, biceps, triceps, brachioradialis, flexor carpi radialis, flexor carpi ulnaris, abductor pollicis brevis, hypothenar eminence, and the first dorsal interosseous. Nerves were stimulated using a bipolar probe and the procedure was continuously monitored by a neurophysiologist.

Incisions were planned to achieve a supraclavicular, infraclavicular, transaxillary, and posterior subscapular approaches [Figures 2c and d]. The operation began through transcervical approach by the otolaryngology (ENT) team members. A curvilinear cervical incision was made along a relaxed skin tension line, extending from the right supraclavicular fossa to the posterior neck [Figure 2a]. The incision was carried through platysma anteriorly, and just

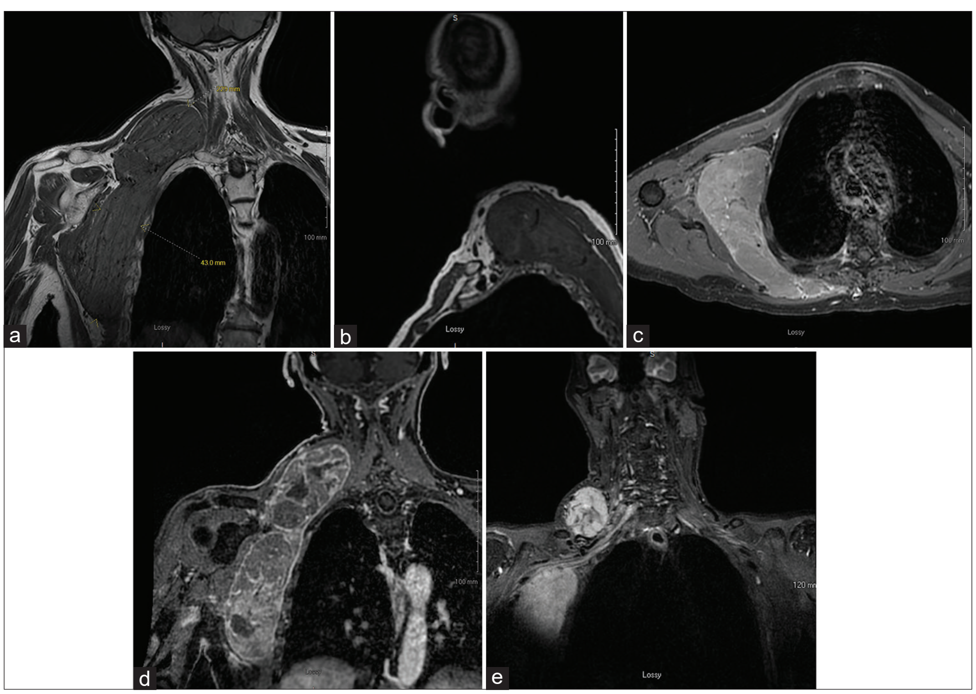

Figure 1: Preoperative magnetic resonance imaging demonstrating an irregular, large bilobed T1 hypointense (a and b), T2 hyperintense (c-e), heterogeneously enhancing mass (c-e) involving the right chest wall, abutting the brachial plexus, and extending into the right neck.

above trapezius muscle posteriorly. Subplatysmal flaps were elevated superiorly and inferiorly down to the clavicle. With this, the capsule of the mass was easily identified superiorly, lying between the posterior border of sternocleidomastoid (SCM) and anterior border of trapezius at its highest extent in neck [Figure 3a]. There was a thin overlying muscular pseudocapsule noted, and the plane overlying the true capsule was carefully identified and exposed. Both blunt and sharp dissection were used circumferentially around the mass, with careful identification and ligation of feeding vessels to the large, necrotic, and vascular mass. These vessels included branches of internal and external jugular vein, and the transcervical artery and vein. Cranial nerve (CN) 11 was identified and preserved.

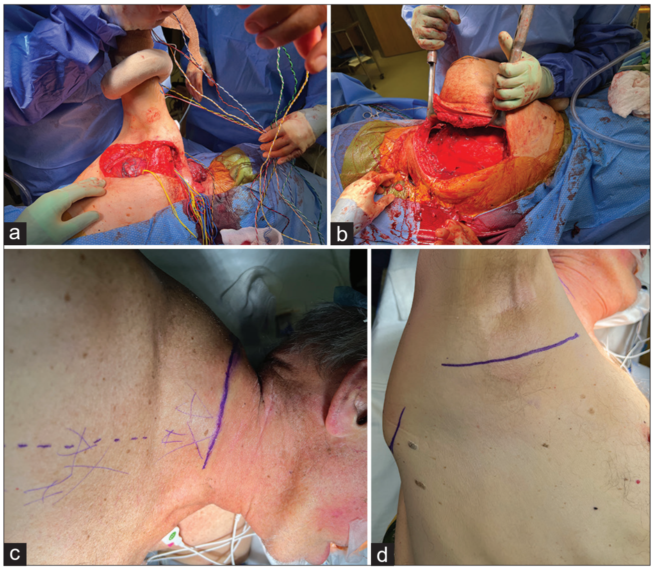

Figure 2: Intraoperative lateral decubitus positioning demonstrating free mobility of the right arm and exposure of neck and anterior (a) and (b) posterior chest wall. (c) Planned supraclavicular and infraclavicular incisions. (d) Planned axillary and posterior subscapular incisions.

Neurosurgery team members then identified the brachial plexus, exiting between the anterior and middle scalene muscles, and descending anterior to the mass. CN 11 was further dissected and confirmed with nerve stimulator running over the posterior superficial portion of mass. The nerve was bluntly dissected free of soft-tissue attachments connecting it to the mass. A flap was elevated containing the nerve for protection throughout remainder of procedure, the nerve was not skeletonized to avoid excessive traction. At this point, we began dissecting the mass from surrounding soft-tissue attachments circumferentially using combination of LigaSure™ (Medtronic) cautery and blunt dissection. Neurosurgical team members continued exposure and identification of the brachial plexus in the neck. The upper and middle trunks of the brachial plexus were identified posterior to the anterior scalene and secured with vessel loops. The omohyoid muscle was identified and divided. The mass was able to be elevated off the spinal vertebrae deeply and all circumferential soft-tissue attachments.

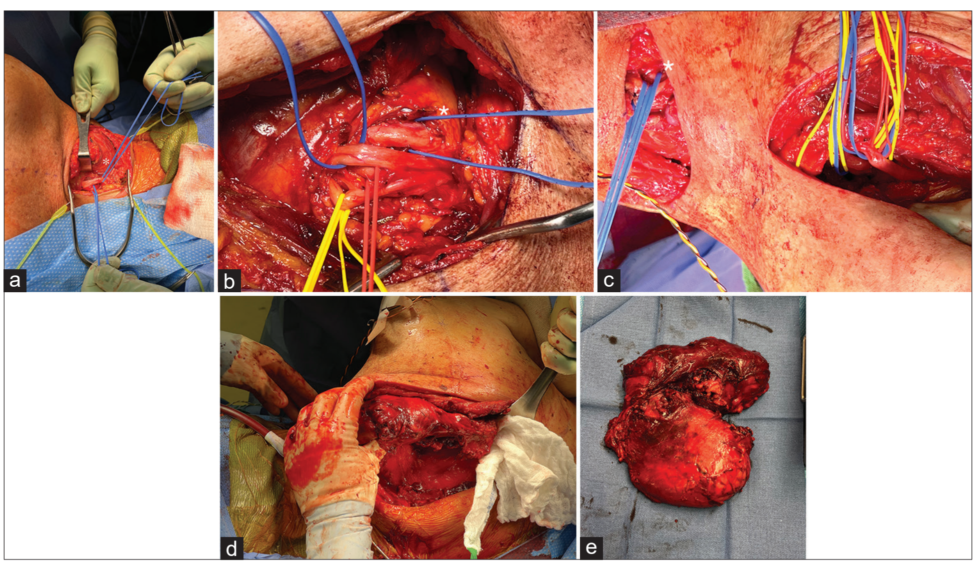

Figure 3: (a) Supraclavicular incision with mass retracted laterally, dissection flap containing spinal accessory nerve, and upper trunk (*blue loop). (b) Infraclavicular exposure with medial cord (yellow loop) lateral cord (*blue loop) and posterior cord (blue loop), and axillary artery (red loop) with underlying mass. (c) Axillary and infraclavicular incisions with proximal and distal brachial plexus exposure including musculocutaneous nerve (*blue loop) and median nerve (blue loop). (d) Posterior subscapular incision with dissection off posterior chest wall and lateral mass retraction. (e) Pathology specimen of gross tumor.

At this point, a descending limb of the incision between the middle and lateral thirds of the clavicle was created and extended caudally along the deltopectoral groove to allow for dissection to continue infraclavicularly [Figure 2c]. The cephalic vein was identified and traced to the deltopectoral groove. The pectoralis minor was transected. More distal plexus components were palpated and identified just deep to the fat pad [Figure 3b]. The clavipectoral fascia was incised which exposed the distal plexus elements which were dissected and separated from the mass [Figure 3b]. The cords of the brachial plexus were then exposed. The lateral cord was seen first and was secured with a vessel loop. The musculocutaneous nerve and then lateral contribution to the median nerve were identified and secured with vessel loops. Medial to the lateral cord was the axillary artery and it was secured with a vessel loop in addition the axillary vein. Once the axillary artery was found [Figure 3b], the posterior cord was identified just posterior to it and secured. Finally, the medial cord was seen medial to the axillary artery. The mass was carefully separated from each of these neurovascular structures.

After maximal safe dissection from the clavicular incisions, our thoracic surgery colleagues made an incision into the axilla for eventual mobilization of the thoracic component of the tumor. Neurosurgery identified the ongoing segments of the brachial plexus, including the terminal branches [Figure 3c]. These structures were looped and intraoperative neuromonitoring confirmed continued function. The tumor abutted these structures but was easily separable with no gross evidence of invasion. The axillary incision was carried down to the chest wall between the posterolateral border of the pectoralis major muscle and the anterolateral border of the latissimus dorsi [Figure 2c]. The tumor was encountered and again, a well-defined capsule was identified. Dissection along this capsule proceeded inferiorly until the inferior edge of the tumor was mobilized, anteriorly until the dissection plane around the brachial plexus was encountered, and posteriorly as far as possible until further dissection was limited by the scapula. At this point, it was clear that a posterior approach would provide the best access to the remaining thoracic component of the tumor which extended both underneath the scapula and posteriorly up into the neck.

The curvilinear cervical incision was then extended posteriorly between the spinous processes and the medial border of the scapula to expose the trapezius muscle [Figure 3d]. The trapezius muscle was incised vertically extending from the cervical dissection far enough inferiorly to provide sufficient exposure to the remaining portions of the tumor. The tumor was dissected along the capsule in the plane between the tumor and the chest wall as well as the more superficial plane between the tumor and the scapula. There were focal areas of adhesions to both the medial border of the scapula as well as the super anterior border of the scapula. After ensuring integrity of critical neurovascular structures and their dissection from the mass, ultimately complete mobility of the tumor was obtained, and it was delivered through the posterior incision. The tumor was extremely large, lobulated, and surrounded along majority of its surface by an intact capsule [Figure 3e].

After copious irrigation and hemostasis confirmation, a Jackson Pratt drain and a blake drain were placed in the cervical portion of the dissection bed and anterior-inferior aspect of the dissection bed, respectively. The trapezius was closed with running suture. The platysma was closed in a running manner after several stitches were used to approximate supraclavicular fat over the proximal portions of the brachial plexus. The axillary incision was closed in layers with care to approximate both muscle and fat over the exposed brachial plexus to separate it from the overlying skin incision. The skin at all sites was closed in multiple layers.

Postoperatively, the patient had pain limited right upper extremity weakness which, however, had intact intrinsic hand function, palpable radial and ulnar pulses, and intact sensation. On histological examination, much of the tumor had morphology of an undifferentiated pleomorphic sarcoma. However, there were several foci of myxoid stroma with associated curvilinear vasculature. These features, in conjunction with the tumor's infiltrative growth pattern, support the diagnosis of a high-grade MFS [Figure 4]. Margins were positive at the deep and superficial aspects.

At his 3-week postoperative visit, the patient had pain related right-sided 4/5 deltoid weakness and resting asymmetry with the right shoulder lower than the left. However, he had 5/5 strength everywhere distally and is working with physical therapy outpatient to improve mobility.

DISCUSSION

MFS is one of the most common soft-tissue masses arising in older adults. Due to MFS's propensity for recurrence and distant metastasis, accurate diagnosis, surgical management, and proper adjuvant therapies are crucial to patient outcomes. Preoperative staging and imaging with computed tomography (CT), ultrasound, and MRI can assist with determining the anatomical complexity of the mass. The optimum strategy for treating MFS is surgical resection with margins.[11] Obtaining margins can be challenging due to MFS multidirectional spreading along fascial plains.

The clinical course of MFS can vary significantly, and they are often initially asymptomatic. Our patient presented with a painless subcutaneous palpable mass of his neck and chest wall which led to further workup. With lower-grade masses, patients typically experience expansile growth, whereas higher-grade tumors often cause invasion and compression of key surrounding structures, such as airway structures in the neck and brachial plexus structures in the axilla. In our

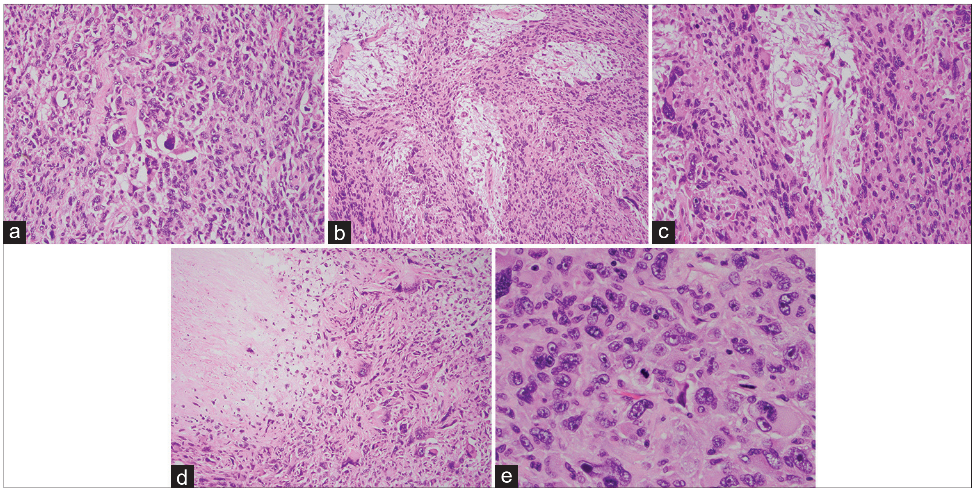

Figure 4: Tumor histology (a) The tumor has high cellularity and severe cytologic atypia (hematoxylin and eosin, ×200 magnification). (b and c) The tumor also had focal areas with loose myxoid stroma and associated delicate vessels (hematoxylin and eosin, ×100 and ×200 magnification. (d) Necrosis was also present (upper left of image; hematoxylin and eosin, ×100 magnification). (e) Mitotic figures were also present (center of image; hematoxylin and eosin, ×400 magnification).

case, key features of higher-grade MFS included the mass's infiltrative growth pattern, its association with and abutment of neurovascular structures, and its histopathological features including high cellularity, severe nuclear atypia, and tumor necrosis. The tumor in this case demonstrated adjacency, without disruption, of neurological function of the brachial plexus.

Multiple studies have demonstrated that extent of resection and tumor size is directly proportional to patient outcomes. Zumarraga *et al.* examined 75 patients with appendicular MFS and reported that overall survival was correlated with positive surgical margins, local recurrence, and distant metastasis.[16] In their series of 158 patients, Sanfilippo *et al.* found that small tumor size and negative margins were the overall best predictors of survival.[12] Lin *et al.* reported that patients with lower mitotic activity and negative margin status were more likely to have distant metastasis-free survival after MFS resection.[5] Each of these studies support safe wide local resection with margins for the best prognosis.

Adjuvant therapies for MFS are appealing due to their ability to prevent recurrence. While data are limited, multiple retrospective studies have shown an association of radiation therapy and decreased rate of recurrence.[6,10] In addition, randomized trials evaluating the clinical benefit of radiation on all subtypes of high-grade sarcoma have shown an improvement in local control.[1,15] The role of chemotherapy as an adjuvant therapy for MFS is more limited due to a lack of randomized clinical trials; however, limited retrospective data have shown no benefit in metastatic control or overall survival.[6,12] Due to the mass location and size, our patient had preoperative radiation therapy through 50 Gray of radiation in 25 intensity-modulated radiation therapy sessions to the right chest wall and neck. In discussions with his oncology team, he decided to forego chemotherapy treatment with doxorubicin. As of 3 months postoperatively, he remains clinically and radiographically disease free on surveillance MRI and CT imaging.

CONCLUSION

We present a case on the resection of a large MFS involving the neck, axilla, and chest wall through a combined team of thoracic surgery, neurosurgery, and otolaryngology-head and neck surgery. Although rare, tumors involving the brachial plexus require adequate anatomical knowledge and complex surgical planning. The anatomical complexity and rarity of tumors involving the brachial plexus impose many challenges onto surgeons. Multidisciplinary teams, each with their own expertise, allow for safe and maximal resection. This case combines four common approaches to the brachial plexus for one giant tumor resection.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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