

# Large cervicothoracic myxoinflammatory fibroblastic sarcoma with brachial plexus invasion: A case report and literature review

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**Abstract.** Myxoinflammatory fibroblastic sarcoma is a rare sarcoma that develops in patients of all ages, which usually presents as a slow-growing painless mass in the distal extremities. To date, myxoinflammatory fibroblastic sarcoma with invasion of the brachial plexus has rarely been reported in the literature. In this study, a case of large cervicothoracic sarcoma, which invaded the brachial plexus, is presented. The patient reported no sensory disturbance or dyskinesia. The tumor was completely resected without injury of the brachial plexus. The postoperative histological diagnosis was myxoinflammatory fibroblastic sarcoma. Follow-up examination performed 24 months after surgery revealed no tumor recurrence and no sensory disturbance or dyskinesia was reported. This study presents a rare case of large myxoinflammatory fibroblastic sarcoma with brachial plexus invasion that was successfully managed by surgery.

## Introduction

Myxoinflammatory fibroblastic sarcoma is a rare neoplasm, accounting for 1% of all adult malignancies (1). The tumor most commonly occurs in the distal extremities and usually affects males and females equally in the fourth and fifth decades of life. Myxoinflammatory fibroblastic sarcomas usually present as a slow-growing painless mass that can mimic infection, ganglion or benign tumors. Accurate diagnosis is based on the postoperative pathological result. Wide local excision is the first choice of treatment. For acral tumors with multiple recurrences, amputation may be considered. The efficacy of chemotherapy and radiotherapy

remains unclear. Myxoinflammatory fibroblastic sarcoma is a low-grade sarcoma with a low rate of mortality and metastasis, demonstrating a long-term clinical course. However, the local recurrence rate is extremely high (2).

Tumors that invade the brachial plexus present a significant challenge as surgical procedures may lead to sensory disturbance and dyskinesia (2).

In this study, the rare case of large myxoinflammatory fibroblastic sarcoma with invasion of the brachial plexus, which that was successfully managed with surgery, is presented.

## Case report

A 54-year-old man, who had suffered from a right cervicothoracic mass for nearly 16 years and did not obtain any medical treatment, was referred to the Department of Hand Surgery (Huashan Hospital, Fudan University, Shanghai, China) in January 2013. The patient presented with a huge mass with no pain and no sensory or motor dysfunction. The patient previously presented to Shanghai Changhai Hospital (Shanghai, China) in November 2012. Magnetic resonance imaging (MRI) and fine-needle aspiration was performed. Subsequently, the patient was referred from Shanghai Changhai Hospital to Huashan Hospital.

Physical examination revealed a right cervicothoracic mass that was palpable from the supraclavicular fossa to the infraclavicular region. The supraclavicular and infraclavicular portions of the mass were ~10x5x3 cm and 25x18x8 cm in size, respectively. On palpation the mass was hard with no tenderness, clear boundaries and negativity for Tinel's sign. The active and passive activity of the right upper limb was normal and no abnormal sensation in the affected limb was reported.

B-ultrasound examination revealed a low echo-level mass located between the right neck and shoulder, extending from the right anterior chest wall to the right anterior axillary region. A chest radiograph identified a dense shadow at the right clavicular region. MRI revealed a large occupying lesion at the supraclavicular and infraclavicular region (Fig. 1). A pre-operative pathological examination performed at Shanghai Changhai Hospital in November 2012 indicated the diagnosis of a benign mesenchymal tumor.

The mass was extremely large, with a supraclavicular portion measuring 10x5x3 cm and an infraclavicular portion measuring 25x18x8 cm (Fig. 2). The mass was exposed via supraclavicular incision and the supraclavicular portion of the

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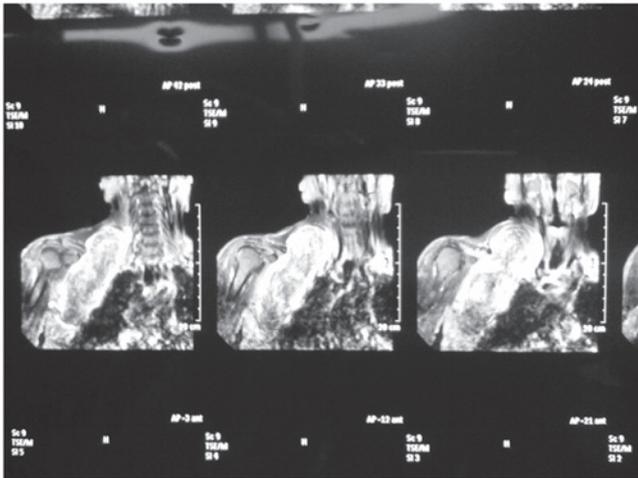


Figure 1. Magnetic resonance imaging revealing large occupying lesion at the supraclavicular and infraclavicular regions.

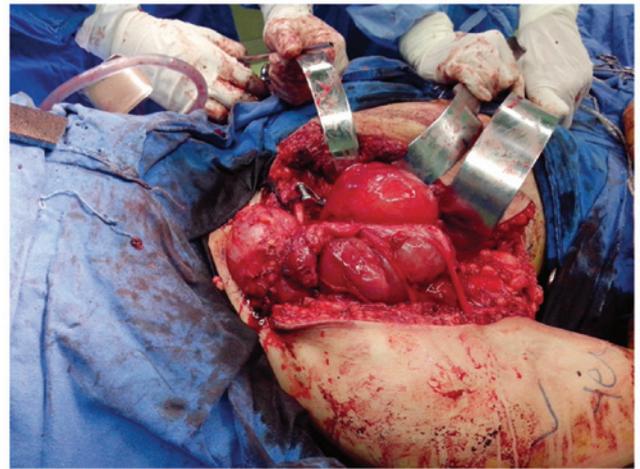


Figure 3. The mass was wrapped and separated by the medial, lateral and posterior cords and the axillary, musculocutaneous, median and ulnar nerves.



Figure 2. The supraclavicular portion of the mass was 10x5x3 cm and the infraclavicular portion of the mass was 25x18x8 cm prior to surgery.

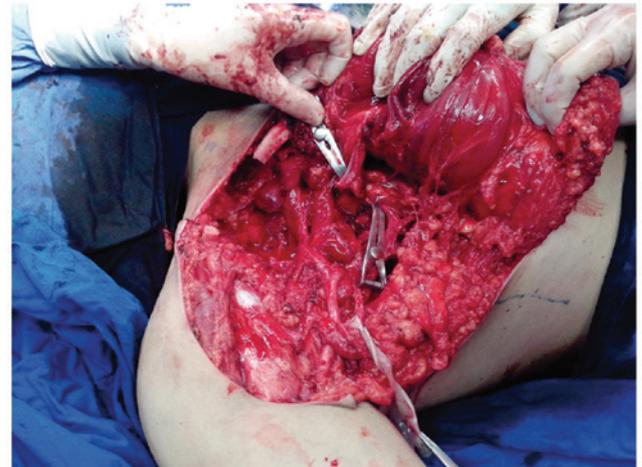


Figure 4. The mass was gradually resected to maintain the integrity of the nerves.

hard mass, measuring 10x5x3 cm, with clear boundaries and an abundant blood supply, was identified. The tumor oppressed the brachial plexus and vessels, and could not be separated by only supraclavicular incision. The mass extended from the supraclavicle to the infraclavicle and could not be completely exposed by only supraclavicular incision. An additional incision from the infraclavicle region to the deltopectoral interval and midaxillary line was made, and the infraclavicular portion of the hard mass, sized 25x18x8 cm, with clear boundaries and an abundant blood supply, was identified. The tumor was lobulated. The mass was wrapped and separated by the medial, lateral and posterior cords and the axillary, musculocutaneous, median and ulnar nerves (Fig. 3). The mass was evidently adhered to the aforementioned nerves and could not be easily separated. The mass was gradually resected carefully to maintain the integrity of the nerves (Fig. 4). Clavicotomy was performed prior to separation of the nerves and vessels surrounding the mass. Finally, the mass was completely resected (Fig. 5). Tumor invasion of the clavicle without bone destruction was observed. Intraoperative electromyography recorded the somatosensory evoked potential by stimulating

the axillary, musculocutaneous, median, radial and ulnar nerves. Following surgery, the activity and sensation of the right upper limb were normal.

The tissue used for histological analysis was embedded in paraffin and sectioned. The histological results showed a prominent mixed inflammatory infiltrate, macrophages, Touton-type giant cells and myxoid matrix, which confirmed the pathological diagnosis of myxoinflammatory fibroblastic sarcoma (Fig. 6) (3). Histopathologically, the lesion demonstrated numerous poly-morphonuclear leukocytes, each with a large vesicular nucleus. The tumor tissue was composed of atypical cells with boundaries that could not be clearly defined.

Immunostaining was performed by two certified pathologists in the present department who were blinded to the clinical characteristics of the patient. Micrometer-thick tissue sections were autoclaved in buffer, and incubated with antibodies. Immunostaining was performed using the biotin-free horseradish peroxidase enzyme-labeled polymer of the Envision Plus detection system. The antibodies used were as follows: anti-vimentin antibody (clone, EPR3776; catalog



Figure 5. The mass was completely resected.

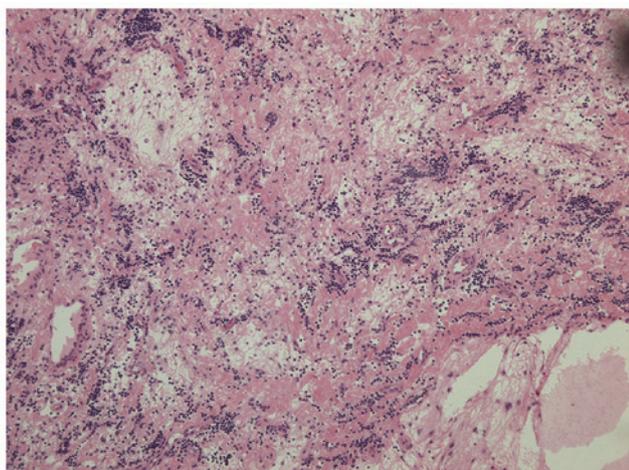


Figure 6. Postoperative histological results showing prominent mixed inflammatory infiltrate, macrophages, Touton-type giant cells and myxoid matrix, indicating a diagnosis of myxoinflammatory fibroblastic sarcoma.

no., ab92547; dilution, 1:200; 60 min at room temperature; Abcam, Cambridge, UK); anti-CD68 antibody (clone, KP1; catalog no., ab955; dilution, 1:200; 60 min at room temperature; Abcam); anti-CD34 antibody (clone, EP373Y; catalog no., ab81289; dilution, 1:250; 60 min at room temperature; Abcam); anti-S100  $\beta$  antibody (clone, EP1576Y; catalog no., ab52642; dilution, 1:500; 60 min at room temperature; Abcam); anti-cytokeratin antibody (clone, AE1/AE3; catalog no., M351501; dilution, 1:200; 60 min at room temperature; Dako, Glostrup, Denmark); anti-EMA antibody (clone, 2F6; catalog no., ab156947; dilution, 1:150; 60 min at room temperature; Abcam); and anti-Desmin antibody (clone, D33; catalog no., ab8470; dilution, 1:200; 60 min at room temperature; Abcam). The Periodic Acid Schiff (PAS) Stain kit (Baso, Zhuhai, China) was also used. Immunohistochemical staining revealed positivity for vimentin, periodic acid-Schiff, cluster of differentiation (CD)68, CD34, S100 and negativity for cytokeratin, epithelial membrane antigen and desmin.

Follow-up MRI examination 24-months after surgery revealed no evidence of tumor recurrence and no sensory disturbance or dyskinesia.

The patient was followed up every month in the first 3 months. Subsequently, the patient was followed up every 3 months until April 2016. Subsequently, the patient will be followed-up every 6 months.

## Discussion

Myxoinflammatory fibroblastic sarcoma, originally termed 'acral myxoinflammatory fibroblastic sarcoma' was first identified by Meis-Kindblom and Kindblom (4) in 1998. In the same year, Montgomery *et al* (5) reported an inflammatory myxohyaline tumor of the distal extremities with virocyte or Reed-Sternberg-like cells, while Michal (6) reported inflammatory myxoid tumor of the soft parts with bizarre giant cells. In addition, Jurčić *et al* (7) demonstrated that occurrence of such tumors in the proximal regions of the limbs and thus the term 'myxoinflammatory fibroblastic sarcoma' was coined. Myxoinflammatory fibroblastic sarcoma develops in patients of all ages with no clear gender predilection.

Laskin *et al* (2) analyzed 104 myxoinflammatory fibroblastic sarcoma patients, which revealed that in 61% of cases, the tumor occurred in the fingers, hands and feet and in 73% of cases the tumor occurred in the dorsal soft tissue involving distal acral sites. Other affected regions included the knees and lower leg, elbow and forearm, ankle, upper leg, upper arm, shoulder and the inguinal region (2). To the best of our knowledge, the present case of a cervicothoracic myxoinflammatory fibroblastic sarcoma with brachial plexus invasion is the first to be reported in the literature. The largest myxoinflammatory fibroblastic sarcoma reported previously was 16 cm in size (8), while the supraclavicular and infraclavicular masses identified in this case were 10x5x3 cm and 25x18x8 cm, respectively.

A painless slow-growing mass or swelling is the most common initial complaint in patients with myxoinflammatory fibroblastic sarcoma. A number of individuals also present with pain, tenderness or dysfunction of the affected area (8). The symptoms observed in the patient in the present case are consistent with those reported in the literature (2).

Preoperative auxiliary examinations indicated a benign tumor in the present study; however, postoperative histological analysis diagnosed low-grade malignant myxoinflammatory fibroblastic sarcoma. We hypothesize that in the context of diagnosis and treatment of large tumors, which are commonly benign, malignancy must always be suspected and therefore the use of pre-operative biopsy may improve diagnosis and treatment.

In the present case, the tumor was carefully resected to protect the medial, lateral and posterior cords, as well as the median, axillary and musculocutaneous nerves, which surrounded the tumor. The postoperative activity and sensation of the limb was normal. Surgery was difficult due to the large tumor size and invasion of the brachial plexus. Therefore, the tumor was separated carefully as injury of the brachial plexus may lead to dysfunction of the upper limb.

The histological differential diagnosis of myxoinflammatory fibroblastic sarcoma may be associated with the myxoid, inflammatory and atypical features. Differential diagnosis includes tenosynovitis, giant cell tumor of the tendon sheath, inflammatory myofibroblastic tumor, liposarcoma, epithelioid sarcoma and myxoid malignant fibrous histiocytoma (9).

Lombardi *et al* (10) performed immunohistochemical analysis in 138 myxoinflammatory fibroblastic sarcoma patients, which revealed that vimentin was strongly positive in all lesions. A total of 84 and 57% of tumors exhibited focal positivity for CD68 and CD34, respectively. Focal positivity for smooth muscle actin, S-100 protein, activin receptor-like kinase 1 and keratin was also observed in 6-16% of patients (10). These results were consistent with the immunohistochemical staining results observed in the patient of the present case: Vimentin(+), CD68(+), CD34(+) and S100(+).

Wide resection is generally accepted as the first choice of treatment for myxoinflammatory fibroblastic sarcoma. At present, the efficacy of chemotherapy and radiotherapy remains unclear and the rate of local recurrence is high (11,12). In the present case, MRI performed during follow-up 24 months after surgery revealed no tumor recurrence. This case reported the successful surgical management of a huge myxoinflammatory fibroblastic sarcoma, which invaded the brachial plexus.

For such a large tumor with invasion of the brachial plexus, the neurological function of the brachial plexus may be preserved through a precise surgical procedure. The association between adjuvant therapy and the prognosis require observation in additional cases.

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