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Case Report

Recurrent Chest Wall Dedifferentiated Liposarcoma, Huge Size at Presentation, Case Report and Review of the Literature

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Received: July 12, 2020; **Published:** September 30, 2020

Abstract

Liposarcoma is the most common soft tissue sarcomas. The disease was first described by Rudolf Virchow in 1857 in a patient with a tumor of the thigh [1]. We present a case of a 37-year old male patient who presented with a recurrent large chest wall soft tissue liposarcoma due to late presentation because of the COVID-19 Pandemic and country lockdown. The disease was not metastatic at presentation and the pathology showed High grade Dedifferentiated Liposarcoma. Wide local resection was performed and defect was covered by Latissimus dorsi flap.

Keywords: *Recurrent; Chest Wall; Dedifferentiation; Soft Tissue Liposarcoma*

Abbreviations

COVID-19: Corona Virus Disease of 2019; MRI: Magnetic Resonance Image; PET: Positron Emission Tomography; FNCLCC: Fédération Nationale des Centres de Lutte Contre le Cancer

Introduction

Soft tissue sarcomas are rare disease entities which usually present in the lower extremity as a growing mass in the deep soft tissues. There are many histological subtypes of soft tissue sarcomas of which liposarcoma is the commonest.

Liposarcoma is considered the most common soft tissue sarcoma, accounting for approximately 17% of all soft tissue sarcomas. The annual incidence is approximately 2.5 cases per million population. The tumor originates from the adipocytes with peak incidence seen in the ages between 40 and 60 years old. There is a slight male predominance. Prognosis depends on the histological subtype as well as the stage of the disease at presentation.

Case Presentation

A 37 years old male patient referred to the orthopedic oncology clinic for further evaluation and management regarding a large chest wall mass. Patient is previously healthy with no past medical diseases, his recent complaints dates back to three years ago when he presented with a soft tissue mass that was excised marginally in another country and the pathology showed atypical lipomatous tumor. Six months prior to presentation he started to feel a mass at the same surgical site that is progressively increasing in size associated with pain and heaviness. Magnetic resonance imaging and CT scan was done and Tru cut biopsy and showed dedifferentiated liposarcoma, he was offered surgical resection but he elected to go back to his home country for further management. Tumor continued to grow rapidly

and was referred to our clinic for further management but due to the country lockdown due to the COVID19 pandemic he was delayed to present for approximately three months. On presentation to the clinic, tumor has grown to an enormous size and started to fungate. Physical examination showed the mass involving most of the left sided chest, huge in size, it was mobile and deep to the fascia, with redness over the skin and impending to fungate (Figure 1). The mass was firm and tender on palpation. MRI with contrast was done and showed a large multi-lobulated heterogeneously enhancing left chest wall/axillary mass with scattered fatty/necrotic component (Figure 2), PET scan was done and showed a large avid lesion in the chest wall and no evidence of nodal or distal metastasis (Figure 3). Tru-Cu needle biopsy shows an extensively necrotic high grade dedifferentiated liposarcoma. During his admission and investigation, the tumor was growing and eventually fungating through his skin. After a multidisciplinary meeting with: Thoracic, Orthopedic, Vascular, Plastic, Musculoskeletal radiology and Interventional radiology teams, the decision was made to do wide local resection followed by adjuvant radiation therapy. Preoperative angioembolization was performed prior to the surgical resection and the tumor was excised and the defect was reconstructed by the plastic surgeon using latissimus dorsi flap (Figure 4). The surgical wound and flap healed nicely three weeks post operatively. The final pathology showed the mass that weighs 3200 gm and measures 28 x 24 x 15 cm and the diagnosis was dedifferentiated liposarcoma with negative margins (Figure 5). Patient was referred for adjuvant radiotherapy.

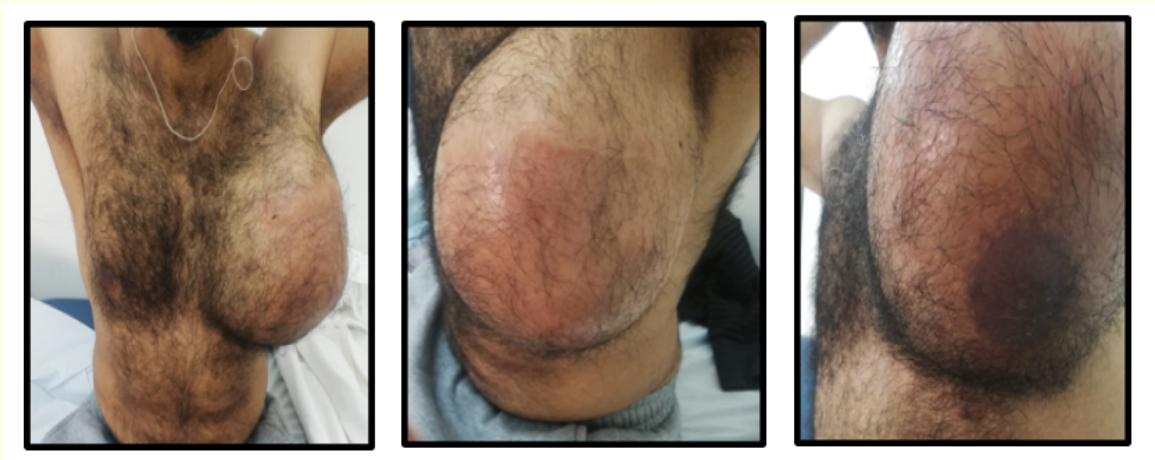

Figure 1: *Clinical images of the tumor.*

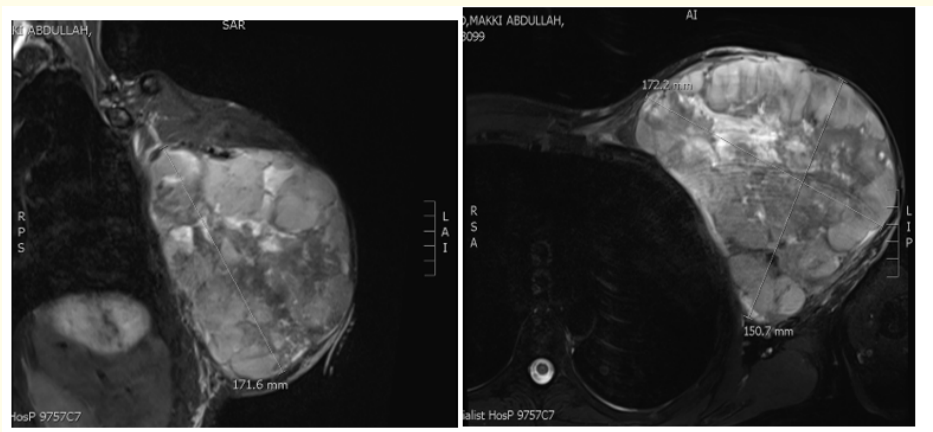

Figure 2: *MRI of the tumor. Coronal (right) and axial (left) plane, post-contrast enhanced T1 weighted images showing a large multi-lobulated heterogeneously enhancing left chest wall mass.*

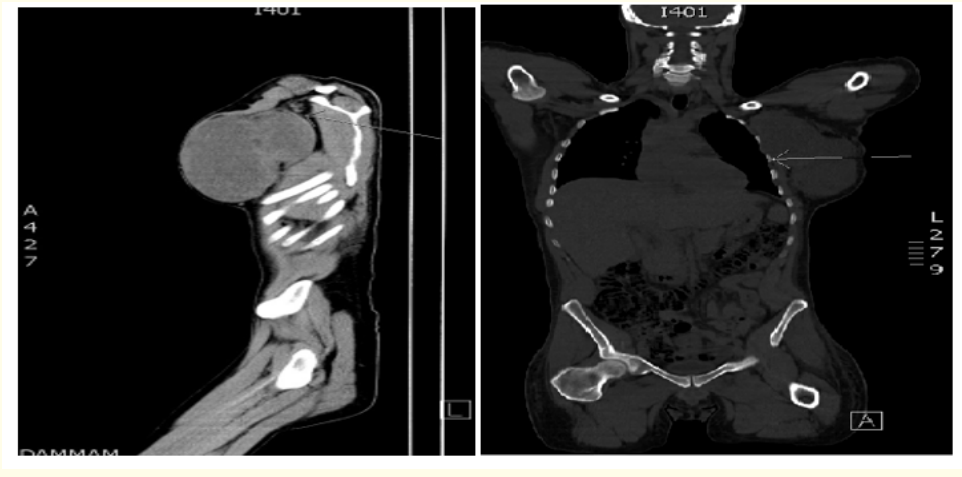

Figure 3: *Sagittal (left) and coronal (right) CT images showing a large, multi-lobulated left chest wall mass, extending to the left axilla, with invasion of the left 3rd rib. PET/CT showed no evidence of distant metastatic disease.*

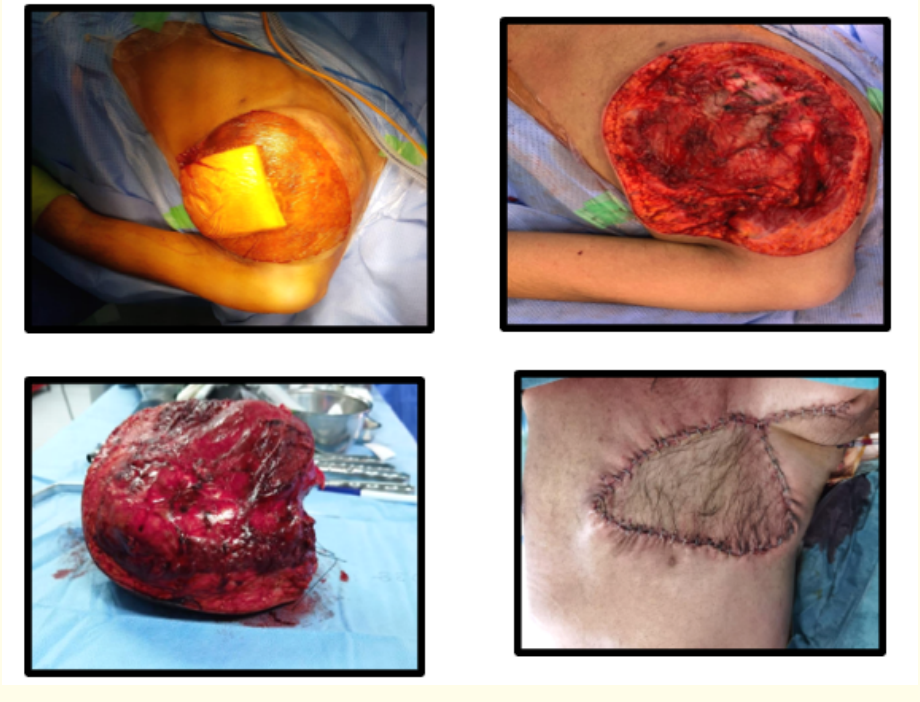

Figure 4: *Intraoperative pictures showing the tumor, the resected specimen. And the soft tissue reconstruction.*

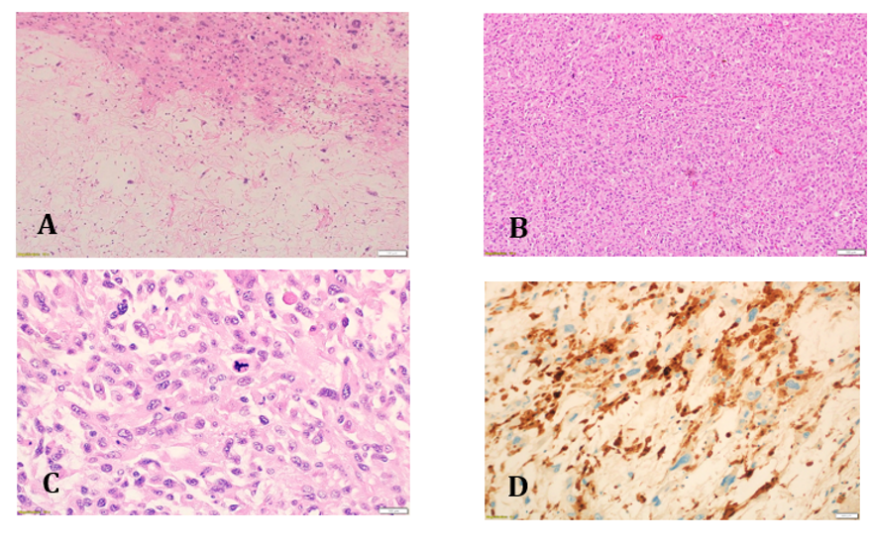

Figure 5: *(A) Medium power magnification showing myxoid areas (lower left) and a cellular pleomorphic areas (upper right) resembling myxofibrosarcoma (Hematoxylin and eosin 200x magnification). (B) Medium power showing non-lipogenic cellular sarcoma with an epithelioid/spindled morphology (Hematoxylin and eosin 200x magnification). C) High power magnification with pleomorphic nuclei, abundant eosinophilic cytoplasm and atypical mitosis (arrow) (Hematoxylin and eosin 400x magnification). D) S100 immunostain positive in the tumor cells (200 x magnification).*

Discussion

Liposarcoma is a soft tissue malignancy that arises from the fat cells of the body either in the extremities or the retroperitoneum. The disease was first described by Rudolf Virchow in 1857 in a patient with a tumor of the thigh [1]. Liposarcoma normally appears as a slowly growing, painless, nonulcerated subcutaneous mass in a middle-aged person, but some lesions grow rapidly and become ulcerated early. Peak incidence is between 40 and 60 years of age, and there is a slight male predominance. Liposarcoma is the most common soft tissue sarcoma, accounting for approximately 17% of all soft tissue sarcomas with an incidence of 2.5 cases per million population diagnosed every year.

Histologically the disease can either be well differentiated, pleomorphic, myxoid or dedifferentiated liposarcoma.

The well-differentiated type and most myxoid types have favorable prognoses, with 100% and 88% 5-year survival rates, respectively [2]. Although they rarely metastasize, repeated local recurrences may cause the tumor to evolve into a higher grade of sarcoma or to Dedifferentiate, in which case metastasis is possible. However, in the other hand of prognosis Round-cell and poorly differentiated types have a non-Favorable prognosis and they tend to metastasize quickly and widely, especially in poorly differentiated liposarcomas [3]. The lungs and the liver are the most common sites of metastasis.

Dedifferentiated liposarcoma represents progression of Well differentiated from an indolent form to more locally aggressive lesion, rapidly growing disease with metastatic potential [4-6].

Five-year disease-specific survival in patients with Dedifferentiated Soft tissue Liposarcoma is 44% compared with 93% in patients diagnosed with pure Well-Differentiated Liposarcoma [4].

High-grade Dedifferentiated Soft tissue Liposarcoma is managed primarily with surgical resection. The tumor is removed with adequate margins of normal tissue or a major fascial barrier circumferentially. Tumor invasion to a major neurovascular structure may require resection with arterial reconstruction. Neoadjuvant radiation therapy is used to reduce the risk of local recurrence in cases of high-grade dedifferentiated Liposarcoma that is greater than 5 cm in diameter or after resection that cannot be improved without causing major morbidity [7,8].

We reviewed the relevant literature on the topic by using PubMed search tool. We found 6 cases reported about chest wall Liposarcoma that been published. We found that the Mean age for these cases was 74.5 year. Female to male ratio was 1:5 and the commonest histopathology type was published is well-differentiated (4 cases) followed by pleomorphic (2 cases), the biggest size of tumor which been reported before our case is 19.0 X 15.0 X 8.5 cm (Table 1).

Case	Age	Gender	Histopathology	Size
Mark W Maxfield; "The surgical management of a giant pleomorphic liposarcoma of the chest wall" (2015, Vol. 23(6) 726-728)	69	Male	Pleomorphic liposarcoma	5 X 14 X 9 cm
Yasir Khan; "Pleomorphic liposarcoma of chest wall: a rare entity with challenging management" (2019, Vol. 27(4) 310-312)	37	Male	Pleomorphic liposarcoma	2.17 X 1.24 X 2.06 Cm
Santosh Shenoy; "Core Needle Biopsy for Diagnosis of Giant Thoracic Liposarcoma" (2010 May;76(5):E33-4.)	89	Male	Well Differentiated	20 X 19 X 6 cm
Yaron Shoham; "Immediate reconstruction of the chest wall by latissimus dorsi and vertical rectus abdominis musculocutaneous flaps after radical mastectomy for a huge pleomorphic liposarcomas" (2013; 47: 152-154)	53	Female	Pleomorphic liposarcomas epithelioid type of the Breast*	15 x 17 x 11 cm
Takeshi Uenotsuchi; "Large subcutaneous liposarcoma arising from the chest wall" (2005; 15 (1): 43-5)	63	Male	Well-differentiated liposarcoma, adipocytic type	16 × 14 × 10 cm
Jin Yong Jeong; "Liposarcoma of the chest wall: a case potentially transformed from a recurrent lipoma ((2011) 59:310-311)	73	Male	Well-differentiated liposarcoma, which showed typical lipoblasts and a dedifferentiated component	16.7 X 13.2 X 4.5 cm
Tsuyoshi Shoji; "Giant primary liposarcoma of the chest" ((2009) 57:159-161)	58	Female	Well-differentiated liposarcoma	19.0 X 15.0 X 8.5 cm
Our case	37	Male	Dedifferentiated Liposarcoma.	26 X 23 X 15 cm

Table 1

To our knowledge, this is the largest soft tissue liposarcoma occurring in the chest wall reported in the English literature, and the reason was the delay in presentation due to the country lockdown and the COVID-19 pandemic during that period which rendered the patient from quick access to care and management.

Conclusion

Liposarcoma is a common soft tissue tumor that can rarely be found in the chest wall. The tumor may be rapidly growing and can present initially with huge size if delayed to presentation and treatment. Such a delay, like in our case, may result in skin contamination and tumor fungation in which the skin has to be sacrificed with the resection. The optimum treatment is wide local resection and soft tissue reconstruction and, in the chest wall, the latissimus dorsi is a good option for coverage.

Summary

Liposarcoma is the commonest soft tissue sarcoma. The disease usually presents in the lower extremity in the deep soft tissues which usually resulted in large size at presentation. Soft tissue sarcomas which originate in an accessible site tend to present earlier with smaller site at presentation, however there are some reports of tumors that present with huge size at presentation in a site where they can be easily detected like the anterior chest wall.

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Volume 11 Issue 10 October 2020