An Atypical Presentation in Journal of a Rare Giant Chest Wall Tumor

Abstract

Pleomorphic liposarcoma (PLS) is a rare high-grade sarcoma that originates commonly in the extremities and on the retroperitoneum. PLS originating in the chest wall is infrequently reported in literature. One such case in an elderly man who presented with a giant anterior chest wall tumor is reported herein. The computed tomography scan of the chest helped in delineating the mass. The diagnosis was confirmed on histopathology. The tumor was managed with a radical excision with reconstruction. The patient underwent follow-up for 1 year during which no local recurrence or metastasis was found. PLS should be considered in the differential diagnosis of primary chest wall tumor.

Keywords: *Giant chest wall tumor, high-grade sarcoma, pleomorphic liposarcoma*

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Introduction

Liposarcoma (LS) is one of the most common types (>20%) of soft tissue sarcoma.[1] It has a number of different variants: Well-differentiated and myxoid/round cell and pleomorphic LS (PLS). PLS is the rarest (<5%) but most aggressive variant.[2] PLS can be discriminated from other high-grade sarcoma by the presence of pleomorphic lipoblast.[3] It is usually seen in late middle age, with slightly male predominance. The lower extremity, particularly the thigh, is the most common location, followed by the upper extremity and the retroperitoneum. Uncommon sites affected include the mediastinum, abdominal cavity, pelvic cavity, paratesticular area, scalp, and orbit. LS originating in chest wall is rare (3%), and most of them are well-differentiated variant (>70%). PLS again is infrequently reported variant in the chest wall.[3]

Herein, we describe a rare case of PLS presenting as giant swelling in an unusual chest wall location that was managed with a radical excision.

Case Report

A 72-year-old male presented with a 25-year history of a painless, giant swelling in the right, anterior chest wall. The swelling had an insidious onset and shown a sudden, rapid increase in size, pain since last 1 month. The patient reported no history of fever, weight loss, loss of appetite, hemoptysis, and trauma or tuberculosis. When questioned on his delayed presentation, the patient stated that since slowly expanding swelling was not affecting his routine activities, he sought medical advice only when localized pain appeared and became unbearable associated with the appearance of dry cough. Local physical examination showed a giant, 15 cm × 15 cm sized, well-defined, oval swelling localized in the right, anterior chest wall just crossing over the sternum. The swelling was nontender, firm, smooth, free from overlying skin but fixed to underlying structures with a narrow pedicle. There was a focal area of scarring on the overlying skin where warm leaves were applied previously as a traditional treatment practice [Figure 1]. The systemic examination was unremarkable except for slight dullness and diminished air entry at the right lung base. Routine laboratory and biochemical tests were unremarkable. The chest radiograph showed nonspecific homogeneous opacity. Ultrasonography (USG) of the swelling revealed a heterogeneous echogenic, well-defined mass. Mammography demonstrated a high-density mass lesion. Computed tomography (CT) scan chest demonstrated a well-defined, round to oval, heterogeneously enhancing

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mass lesion of 14.7 cm × 9.1 × cm 12.5 cm size, with multiple, nonenhancing, hypodense areas consistence with necrosis. Multiple fat containing areas and flecks of internal calcifications also noted. Indistinct fat planes with possible invasion to underlying chest wall muscle, underlying thickened pleura in the right parasternal area and gross pleural effusion at the right lung base was noticed. The sternum and ribs were free from the tumor. The findings were consistent with a malignant soft tissue tumor [Figure 2].

The core needle biopsy of the swelling revealed the presence of varying number of pleomorphic lipoblasts arranged in storiform growth pattern against a background of high-grade malignant fibrous histiocytoma (MFH) which was also confirmed later on by histological examination of the excised specimen [Figure 3].

Immunohistochemical (IHC) markers examination showed varying degree of positivity [Figure 4].

a. S 100 (4+) in lipoblasts
b. DESMIN (1+) in occasional neoplastic cells
c. SMA (3+) in spindle cells
d. VIMENTIN (4+) in spindle cells and lipoblasts.

Morphologic and IHC markers findings were consistent with a PLS (FNCLCC grade 3).

USG-guided aspiration of right pleural effusion reported a dark colored, serosanguineous, nonmalignant reactionary fluid. Based on these findings, a radical surgical excision with reconstruction was planned.

Surgical intervention revealed a large, encapsulated multilobulated, grayish mass weighing 3 kg with areas of necrosis and clot, having focal adhesion to underlying muscle and pleura which was also excised followed by reconstruction. The patient had uneventful postoperative recovery [Figure 5]. The patient was kept on regular follow-up and shown no local recurrence or metastasis at 1 year.

Discussion

The chest wall PLS is rare and there is infrequent case report available in literature.[4,5] Our case may represent the largest PLS in the anterior chest wall. PLS usually present as asymptomatic growing mass over a variable duration of time until they invade nerves causing pain or produces other symptoms by compressing adjacent

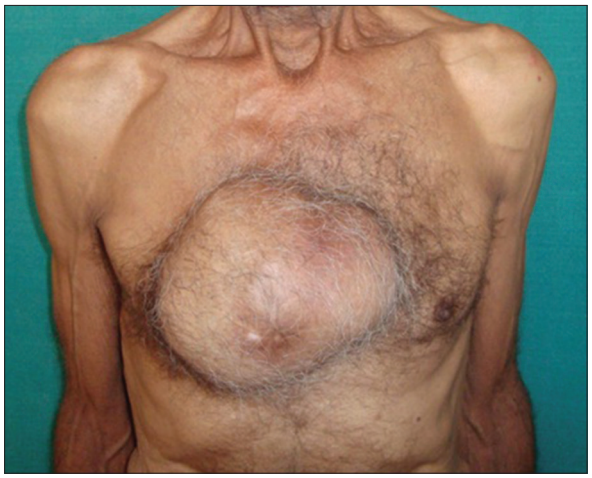

Figure 1: A giant anterior chest wall tumor

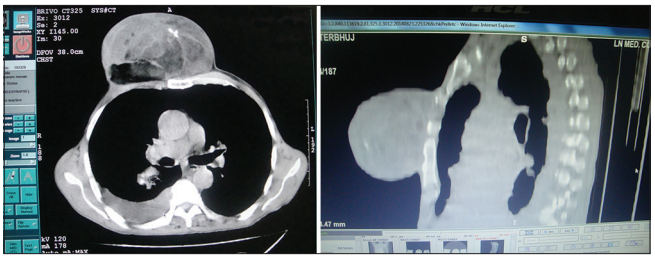

Figure 2: Computed tomography chest shows a well-defined round to oval heterogeneous, enhancing mass lesion with multiple, nonenhancing hypodense areas consistence with necrosis, multiple fat containing areas, and flecks of internal calcifications

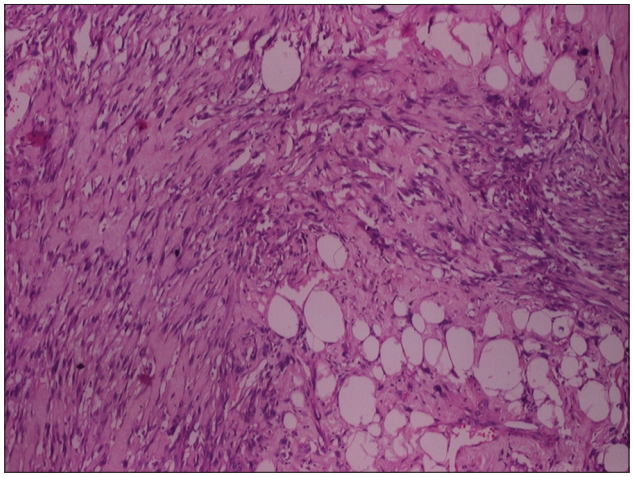

Figure 3: Histopathology examination photograph showing varying number of pleomorphic lipoblasts in a background of a malignant fibrous histiocytoma-like high-grade sarcoma

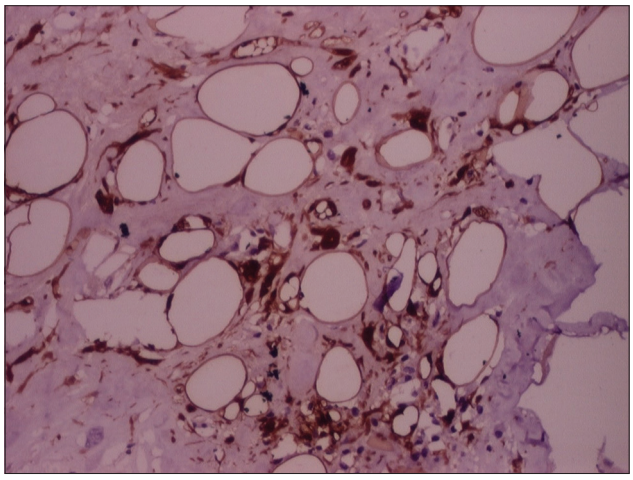

Figure 4: High resolution image showing lipoblast strongly positive for Immunohistochemical marker) S 100

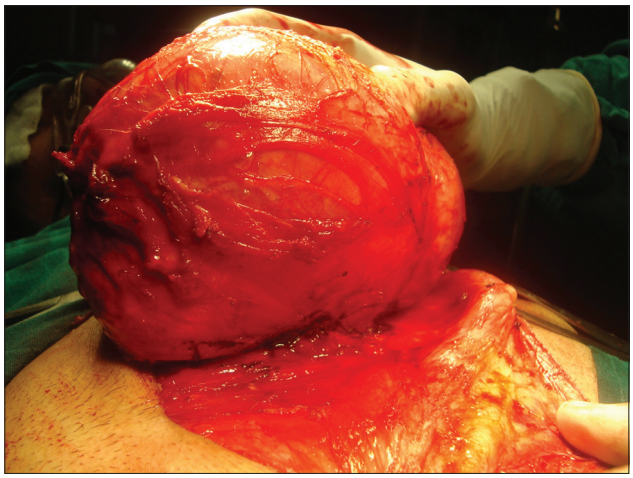

Figure 5: Excised specimen showing large encapsulated, multilobulated, mass with narrow pedicle

structures. Hence, they often diagnosed late and delayed diagnosis leads to larger tumor at presentation.[6] The definitive diagnosis is made only at a histological examination by carefully demonstrating the presence of pleomorphic lipoblasts against a background of high-grade sarcoma which commonly resemble MFH and rarely epithelioid carcinoma. Strong positivity of IHC markers S-100 and vimentin helps in excluding the PLS from the other nonlipogenic sarcoma and carcinoma with pleomorphic cells.[7]

Clinically, it is often difficult to differentiate LS from other soft tissue swellings of the chest wall. CT or magnetic resonance imaging is equally effective in determining the complete extent of the mass, differentiating and making an operative decision, but CT is more effective in demonstrating the metastasis. The presence of calcifications and fat in a heterogeneously enhancing mass lesion is an important differential for LS.[8]

Prognosis of PLS is affected by older age, larger size, central and deep location, higher histological grade, positive surgical margins, surgical procedure, metastasis at presentation.[9,10]

Complete surgical excision is the preferred therapeutic choice. The role of adjuvant radiotherapy or chemotherapy is controversial with no reported benefit found in literature.[10]

Conclusion

PLS is a rare high-grade sarcoma of late adulthood that frequently metastasizes to lung and pleura. It affects commonly the deep soft tissue of extremities. Complete surgical excision is the preferred treatment modality, and overall prognosis is poor. Our case may represent the largest PLS in unusual anterior chest wall location with relatively less aggressive behavior.

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Conflicts of interest

There are no conflicts of interest.

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