Liposarcoma of the chest wall

Transformation of dedifferentiated liposarcoma from a recurrent lipoma

Pinar Bicakcioglu, *MD,* ***Serpil D. Sak,*** *PhD,* ***Abdullah I. Tastepe,*** *PhD.*

ABSTRACT

تعد الساركومة الشحمية ثاني أكثر الساركومات الشحمية في الأنسجة الطرية انتشاراً وذلك بعد ورم المنسجات الليفي الخبيث لدى البالغين. وعادة ما تظهر هذه الساركومة في الأطراف وفي الحيز خلف الصفاق، غير أنها نادراً ما تظهر في جدار الصدر. نستعرض في هذا المقال حالة نادرة لساركومة شحمية ضخمة وُجدت في جدار الصدر، وقد ظهرت متحولة من ورم شحمي متنكس. لقد قمنا بقطع جدار الصدر وإعادة بناؤه بتغيير موضع العضلة الظهرية العريضة بواسطة بضع الصدر الخلفي الوحشي. لقد تلقى المريض 4 مراحل من العلاج الكيميائي المساعد وذلك بعد إجراء العلمية الجراحية وتشخيص الحالة على أنها ساركومة شحمية فاقدة التمايز. ولم يشكو المريض من أية مضاعفات بعد العملية، كما أنه كان معافى من الأمراض لمدة 30 شهراً.

Liposarcoma is the second most common soft tissue sarcoma after malignant fibrous histiocytoma in adults. It is frequently found in the extremities and retroperitoneum; rarely it can be seen in the chest wall. We report a rare case of giant liposarcoma originating from the chest wall representing a transformation of a relapsing lipoma in the same region. We performed chest wall resection, reconstruction with latissimus dorsi muscle transposition via posterolateral thoracotomy. The patient received 4 series of adjuvant chemotherapy after the postoperative diagnosis of dedifferentiated liposarcoma. The patient had no postoperative complication and has remained disease-free for 30 months.

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From the Department of Thoracic Surgery (Bicakcioglu), Ataturk Training and Research Hospital for Chest Diseases and Thoracic Surgery, Kecioren, Department of Pathology (Sak), Ankara University Medical Faculty, and the Department of Thoracic Surgery (Tastepe), Gazi University Medical Faculty, Ankara, Turkey.

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Address correspondence and reprint request to: Dr. Pinar Bicakcioglu, Department of Thoracic Surgery, Ataturk Training and Research Hospital for Chest Diseases and Thoracic Surgery, Kecioren 06280 Ankara, Turkey. Tel. +90 (312) 3552110. Fax. +90 (312) 3552135. E-mail: piyaren@gmail.com

Soft tissue tumors originating from primitive mesenchymal tissues are rare tumors, accounting for only 1% of malignant neoplasms.[1,2] Liposarcomas account for 15% of soft tissue sarcomas and is frequently seen in the extremities and retroperitoneum, rarely it is seen in the head, neck, and inguinal region. Chest wall involvement is reported to be rare, approximately 3%.[3] We present this case because of rarity of the localization, and this type of relapse. Initially, it appeared as a lipoma, then relapsed as lipoma and then well differentiated liposarcoma, and at the last relapse it was transformed to dedifferentiated liposarcoma.

Case Report. A 66-year-old male patient presented to our clinic in July 2009, with a recurrent giant, painless mass, which has been growing for 2 years in his left chest wall. He had undergone surgical resection of a subcutaneous lipoma of the same site in 1999. The lesion relapsed again in the same region in 2002 and 2006, and excisional biopsies revealed lipoma and well-differentiated liposarcoma, respectively. His past medical history revealed hypertension and heavy smoking of 40 packs a year. On physical examination, there was previous incisional scars in the left axillary region, and a giant, lobulated, fixed and hard mass palpated in the same region. A computed tomography scan of the chest revealed a mass in the left hemithorax; intersection with the muscles were indistinct, there was intrathoracic extention with costal involvement and having calcific, necrotic areas (**Figure 1A**).

Whole body scan showed no distant metastases then surgery was performed. Left posterolateral thoracotomy was performed from seventh intercostal space. Intraoperative exploration revealed a lobulated, hard mass on the 3rd, 4th, 5th, and the 6th rib; infiltrating the serratus anterior, extending towards the lower end of the scapula, nipple, axilla and partly the pectoralis major muscle. Mass was extending to intrathoracic extrapulmonary region, lung tissue was normal (**Figure 1B**). En-bloc resection of the mass together with

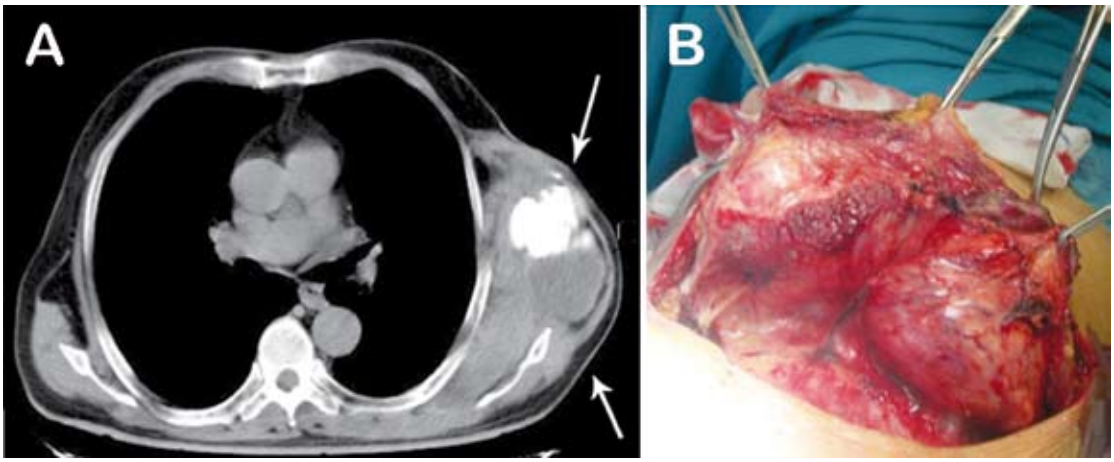

Figure 1 - Image of **A)** mass in thorax computed tomography. **B)** Intraoperative view of the mass.

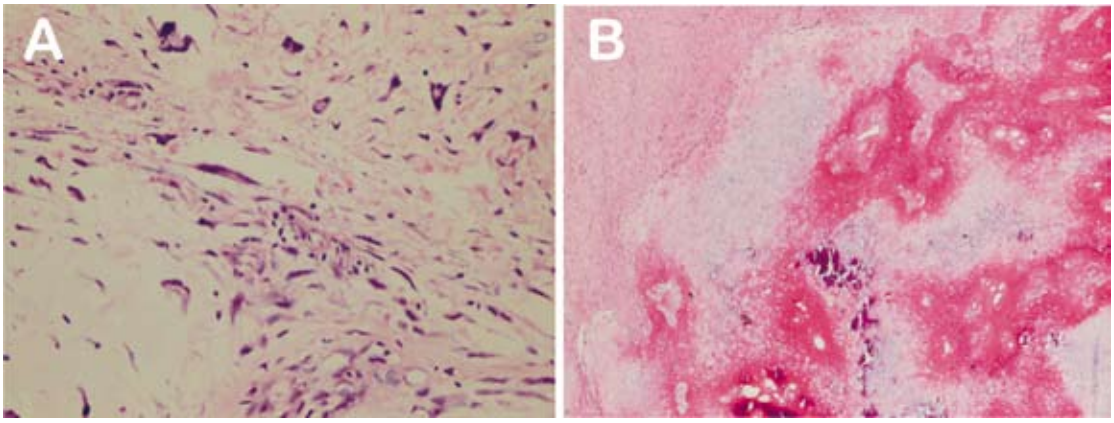

Figure 2 - **A)** Histopathology of dedifferentiated liposarcoma with myxofibrosarcoma-like areas, (H-E x400). **B)** Histopathological view with heterogynous elements of dedifferentiated liposarcoma involving osteo/chondrosarcomatosis areas.

the adjacent chest wall was performed and defect was repaired with a 20x30 cm polytetraflouroethylene mesh patch and latissimus dorsi muscle flap with assistance of plastic surgeon. Histopathological diagnosis of the 25x25x10 cm mass was reported as dedifferentiated liposarcoma. All lymph nodes were reactive with negative surgical margins.

In the second recurrence, tumor consists of the fibrotic zones, trabeculae intersecting fat, fibrotic areas contained collagen fibrils in which were embedded scattered spindle and multipolar hyperchromatic stromal cells. In the third recurrence, tumor contained myxofibrosarcoma-like areas, and osteo-chondrosarcomatosis areas (**Figures 2A & 2B**). The patient was discharged on the fifth postoperative day. Four series of adjuvant chemotherapy were given without radiotherapy. The patient has remained disease-free on the 30th postoperative month and recent thoracic magnetic resonance imaging revealed no relapse.

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Discussion. Liposarcoma is the second most common soft tissue sarcoma in adult life.[1,2] According to the last classification of the World Health Organization, it is histologically divided into 4 subtypes: (a) atypical lipomatous neoplasm (ALN)/well differentiated liposarcoma (WDL), (b) myxoid/round cell, (c) dedifferentiated liposarcoma (DDL), and (d) pleomorphic liposarcoma.[4] Atypical

lipomatous neoplasm/WDL is the most common form of liposarcoma encountered in the sixth and seventh decades of life. Men and women are equally affected.[2] Liposarcoma is frequently seen in the deep muscle of the extremities (75%) and in the retroperitoneum (20%). The reminder was divided between the head and neck, groin, spermatic cord, and miscellaneous areas.[5] Also, liposarcoma of the chest wall is uncommon.[6] Unlike ALN/WDL, DDL most commonly occurs in the retroperitoneum and inguinal regions.[7] We present a case that demonstrates transformation of dedifferentiated liposarcoma from lipoma in the chest wall. Liposarcomas are generally huge lesions and gradually grow to reach giant scales with local invasion. It can be determined as a growing mass or asymptomatic mass detected by coincidence on routine chest radiography. Pain can be the main symptom if the nerve is invaded.[3] In our case, the patient had a growing mass in the chest wall that was painless.

Traditionally, DDLs were defined as ALN/WDL juxtaposed to areas of high-grade unlipogenic sarcoma, usually resembling either a fibrosarcoma or malignant fibrous histiocytoma. Dedifferentiation was believed to represent after a latent period of several years. These views have now been modified. Whereas, most DDLs display high-grade dedifferentiation, a small number contain exclusively low grade areas or a combination of both areas (secondary dedifferentiation). Second, it can be seen in the original biopsy or excision material (de novo or primary dedifferentiation).[4,8] In our case, the DDL was originated from WDL with secondary dedifferentiation after lipoma, and involving osteo/chondrosarcomatosis areas.

Dedifferentiation most commonly occurs in the retroperitoneum with aggressive progression.[7,8] Henricks et al[9] reported that, 133 of the 155 DDL to be de novo with most of them (106 cases) located in the retroperitoneum. In their report, 41% of the patients had local recurrence, 17% of had distant metastasis and 28% of died from DDL. The most important prognostic factor is anatomic location, with retroperitoneal lesions having the worst clinical behavior, which could be because of a longer latent period in the well-differentiated components at these sites. In addition, it is often impossible to achieve a wide surgical excisional margin in the retroperitoneum.

Lipomas are completely benign, but they may recur locally, fewer than 5%. Malignant changes are virtually unheard of, and only a few cases have been reported in the literature.[4] There were 2 cases of lipomas that differentiated to liposarcomas as far as we have found in the literature; this is the third case. Treatment in liposarcoma is curative surgery with maintaining negative surgical margins. The efficacy of radiotherapy and chemotherapy is disputable and it is noted that the best response is with the myxoid type.[7] Prognosis of DDL is generally poor, whereas tumors located in soft tissues having relatively better prognosis compared to those located in the retroperitoneum.[8] Although dedifferentiated liposarcoma has poor prognosis, it can have a better prognosis if it is resectable with negative margins.[10] Despite complete resection was achieved in our case, adjuvant chemotherapy without radiotherapy had been given after resection, due to the giant size of the tumor and to prevent relapses.

In conclusion, although lipomas are benign lesions, in rare locations like thoracic wall they may have malignant potential. Complete resection of the tumor and long term follow up is recommended.

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