Giant atypical lipomatous tumor of the mediastinum

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ÖZET

Mediastenin dev atipik lipomatöz tümörü

İyi diferansiye liposarkom olarak da adlandırılan atipik lipomatöz tümörler adipositik tümörlerin ara veya lokal agresif formudur. Bu tümörler mediastende nadiren yerleşir ve eksizyon sonrası potansiyel nüks riski taşır. Bu olgu sunumunda mediastende lokalize olan ve cerrahi olarak eksize edilen dev atipik lipomatöz tümörlü bir hasta sunuyoruz. Hasta nüks gelişmeden postoperatif 17. ayda asemptomatik olarak izlenmektedir. Cerrahi, mediastinal liposarkomların tedavisinde en iyi seçenektir.

Anahtar Kelimeler: *Lipomatöz tümör, liposarkom, mediasten.*

SUMMARY

Giant atypical lipomatous tumor of the mediastinum

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Atypical lipomatous tumors, so-called well differentiated liposarcomas are the intermediate or locally aggressive form of adipocytic tumors. Mediastinum is an uncommon localization for these tumors and they have a potential risk of recurrence fol-

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lowing excision. We herein report a case with a giant atypical lipomatous tumor located at the mediastinum that was surgically excised. The patient is free of recurrence and asymptomatic at postoperative 17 months. Surgery remains as the treatment of choice for mediastinal liposarcomas.

Key Words: *Lipomatous tumor, liposarcoma, mediastinum.*

Although liposarcomas are the single most common sarcomas of adulthood, accounting for approximately 20% of all soft tissue sarcomas, a mediastinal liposarcoma is an uncommon tumor representing around 1% of all malignancies and 9% of primary sarcomas of the mediastinum (1,2).

The intermediate or locally aggresive form of adipocytic tumors is the well differentiated liposarcoma, the so-called atypical lipomatous tumour. These tumors constitute the larger subgroup of all liposarcomas with a rate of 40-45% which tend to occur equally in the retroperitoneum or the limbs followed by the paratesticular area, the mediastinum, and very rarely in the subcutaneous tissues. Atypical lipomatous tumours most often occur in adults between fifth and seventh decades with a slight male predominance (3). Although surgical excision is the optimal treatment of choice for these tumors, the recurrence rates may range from 50 to 90% following surgery (2).

We herein report a 52-year-old male patient with a giant atypical lipomatous tumor located in the mediastinum, who underwent total excision of the tumor through a left posterolateral thoracotomy.

CASE REPORT

A 52-year-old man with two months cough admitted with a huge left-sided intrathoracic mass diagnosed with chest X-ray that was confirmed by computed tomography which showed a mediastinal tumor with the density of fat (Figure 1). He had a history of gastric bleeding, atrophic gastritis, vitamin B-12 deficiency, and Gilbert syndrome for 15 years. Laboratory data were within the normal limits. Physical examination revealed decreased breath sounds in the left lower zone. The patient underwent a left posterolateral thoracotomy which showed a tumor attached to the mediastinum with a small pedicle. The tumor was totally excised with ligation of its pedicle (Figure 2). Macroscopically, the tumor was well capsulated with congested vascular structures on its surface. It measured 24 x 19 x 11 cm in diameter and weighed 2370 grams. Histological examination revealed an atypical lipomatous tumor showing features of prolifera-

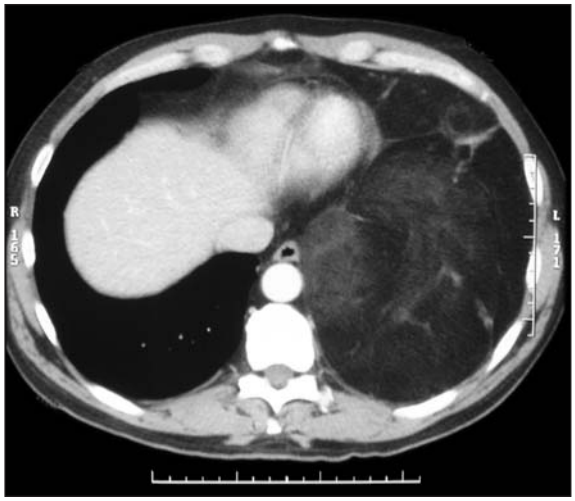

Figure 1. Computed tomography showing a huge sized tumor with fat density involving the left lower zone.

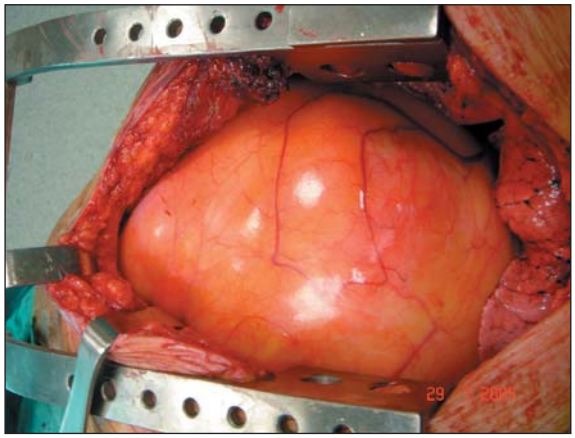

Figure 2. Intraoperative view of the tumoral mass.

ting adipocytes of different size with nuclear hyperchromasia (Figure 3). The patient is free of recurrence and asymptomatic at 17 months follow-up proven by control chest tomographies.

DISCUSSION

Adipocytic tumors may be classified as benign, intermediate and malignant tumors according to WHO classification of soft tissue tumors (4). The benign group of adipocytic tumors consists of lipoma, lipomatosis, lipomatosis of nerve, lipoblastoma/lipoblastomatosis, angiolipoma, myolipoma, chondroid lipoma, while the malignant group consists of liposarcomas such as dedifferentiated, myxoid, round, pleomorphic and mixed cell types. Well differentiated liposarcomas, the so-called atypical lipomatous tumours, comprise the intermediate or locally aggressive form of adipocytic tumors.

Atypical lipomatous tumours can be categorized histologically as adipocytic (or lipoma-like), sclerosing, spindle cell and inflammatory type. The most common forms are adipocytic (or lipoma-like) and sclerosing types, whereas spindle cell variant is much rarer (1). Adipocytic (or lipoma-like) liposarcomas are usually well-circumscribed, lobulated masses that may attain huge dimensions as in our case. Macroscopically, they may be indistinguishable from ordinary benign lipomas, which make microscopic examination essential for the differential diagnosis. However, an atypical lipomatous tumour is microscopically composed of a relatively mature adipocytic proliferation with significant variation in cell size compared with a benign lipoma. In addition, atypical lipomatous tumours have focal adipocytic nuclear atypia showing hyperchromasia and scattered hyperchromatic multinucleate stromal cells.

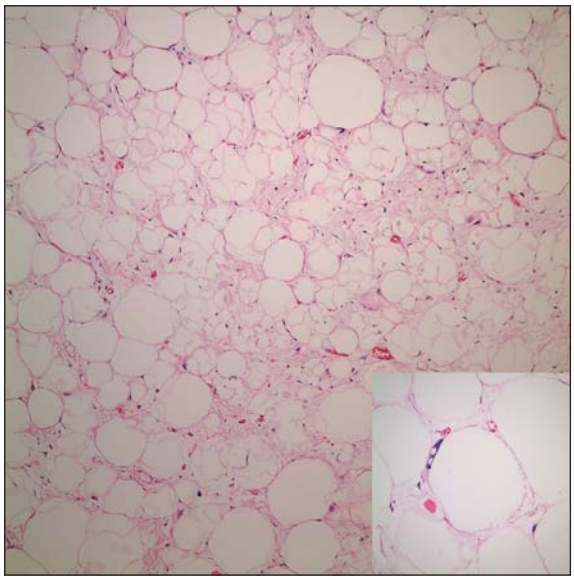

Figure 3. The neoplasm is characterized by proliferation of adipocytes with marked variation in cell size (HE, x100). Atypical cellular features, such as nuclear hyperchromasia were present (inset) (HE, x400).

Among the malignant liposarcoma subtypes, pleomorphic and round cell types are more aggressive which show a greater risk of local recurrence and metastasis. Moreover, patients with well differentiated liposarcomas those without an encapsulated tumor have a similar risk of recurrence and metastasis. On the other hand, our patient is free of recurrence and asymptomatic at postoperative 17 months. It has also been shown that well differentiated liposarcomas of the groin and retroperitoneum transform to dedifferentiated liposarcomas with a relatively greater rate (26%) compared to those tumors located at the trunk and extremities (1%) (5).

Mediastinal liposarcomas appear as a fat containing mass to a solid mass on computerized tomography with low attenuation values ranging from -50 to -150 Hounsfield units (HU) (6). Greater values may be related to the necrosis, heterogeneity and soft tissue component of the liposarcomas. On magnetic resonance imaging, the signal intensity is comparably higher in T1-weighted images than in T2-weighted images showing the fatty component of these tumors (7).

Surgical excision appears as the optimal treatment of choice in patients with mediastinal liposarcomas because either chemotherapy or radiotherapy has a limited effect on survival for liposarcomas (2). Radiotherapy is very likely to result in mediastinal fibrosis at this location, and the effect of chemotherapy has yet to be determined. Furthermore, it has been reported that patients did benefit from repeated surgical resections in case of recurrence (2,8,9).

In conclusion, atypical lipomatous tumours are quiet rare tumors of the mediastinum which are very likely to recur following excision and surgery remains as the optimal treatment of choice for these tumors.

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