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An unusual case of recurrent huge primary mediastinal dedifferentiated liposarcoma

Noni Novisari Soeroso[a,*], Andika Pradana[a], Marshal Djaka[b], Maulidya Ayudika[b], Soekimin Ngadimin[c], Luhur Soeroso[a]

[a] Department of Pulmonology and Respiratory Medicine, Faculty of Medicine, Universitas Sumatera Utara, Universitas Sumatera Utara Hospital, Jl. Dr Mansyur No. 66, Medan 20154, Indonesia
[b] Department of Thoracic Surgery, Faculty of Medicine, Universitas Sumatera Utara, Universitas Sumatera Utara Hospital, Jl. Dr Mansyur No. 66, Medan 20154, Indonesia
[c] Department of Anatomic Pathology, Faculty of Medicine, Universitas Sumatera Utara, Universitas Sumatera Utara Hospital, Jl. Dr Mansyur No. 66, Medan 20154, Indonesia

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ABSTRACT

INTRODUCTION: Primary mediastinal dedifferentiated liposarcoma is an extremely rare malignant mesenchymal neoplasm composed of lipogenic tissue. It can be huge enough to compress heart and surrounding organs leading to clinical symptoms.
PRESENTATION OF CASE: We present a case of huge primary mediastinal dedifferentiated liposarcoma in a 54-year-old man, confirmed by immunohistochemistry staining, who received surgical removal. However, six months later, the tumor recurred, and the patient underwent chemotherapy for 6 cycles resulting in stable disease. Six months after the last visit, the tumor showed no sign of recurrence anymore.
DISCUSSION: Among the various subtypes, the dedifferentiated mediastinal liposarcoma is the least found type of liposarcoma and often leads to misdiagnosis. Challenges occurred not only in diagnosis but also in treatment since it frequently grows back and chemotherapy may be needed after surgery.
CONCLUSION: This is a highly rare case of huge mediastinal liposarcoma that recurred after surgery and gave a satisfying outcome after chemotherapy.

1. Introduction

Liposarcomas are malignant mesenchymal tumors with varying degrees of atypia. It mostly found in lower limbs and retroperitoneum. Mediastinal liposarcoma (ML) is an extremely rare entity, accounting for about 1% of all mediastinal tumors, predominantly in posterior part, and less than 2% of all liposarcomas [1]. Dedifferentiated liposarcoma is the least common subtype of liposarcoma, particularly found in male over the fourth decades [2] and appear to be challenging to diagnose.

Due to its slow progression, around 15% of patient may be asymptomatic. The symptoms are related to tumor size and its invasion to adjacent organs. Most patients present with dyspnea, chest discomfort or cough. Patients may also present with superior vena cava syndrome [2] Most similar reported cases were successfully treated by complete surgical excision alone [1,2], but in this case, after removal of the giant tumor, it recurred 6 months later and leading for a need of chemotherapy.

All the works reported in this article were in line with the Surgical Case Report (SCARE) guideline [3].

2. Case report

A 54-year-old male presented with shortness of breath for 4 months, gradually worsened so severely that upon admission, he could no longer lying down, along with cough and swallowing difficulty. He was light smoker. Physical findings showed diminished breath sound in the left hemithorax.

Complete blood count and serum tumor markers (AFP, Beta-HCG, LDH and CEA) were all within normal limit. Chest X-ray showed consolidation in almost the whole left hemithorax, shifting the trachea and mediastinum to the right. Thoracic CT-scan revealed a fat-containing mass with heterogenous density extending into the anterior mediastinum, enhanced with contrast, causing left lung atelectasis, suggestive for mediastinal teratoma with right minimal pleural effusion (Fig. 1).

Cytology of transthoracic lung biopsy were inconclusive. Patient was then scheduled for median sternotomy, open biopsy and surgical removal. Upon surgery, a giant capsulated mass measuring 50 × 30 cm was removed (Fig. 2A). However, due to its invasion to

* Correspondence author.
E-mail address: noni@usu.ac.id (N.N. Soeroso).

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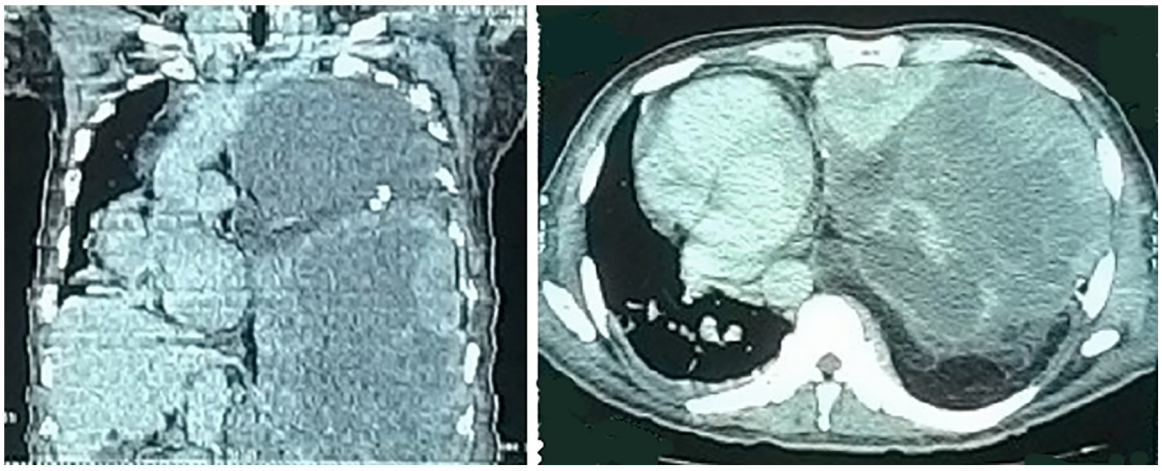

Fig. 1. The coronal and axial view of chest CT Scan revealed an inhomogeneous fatty mass in the left hemithorax, shifting heart and trachea to the right.

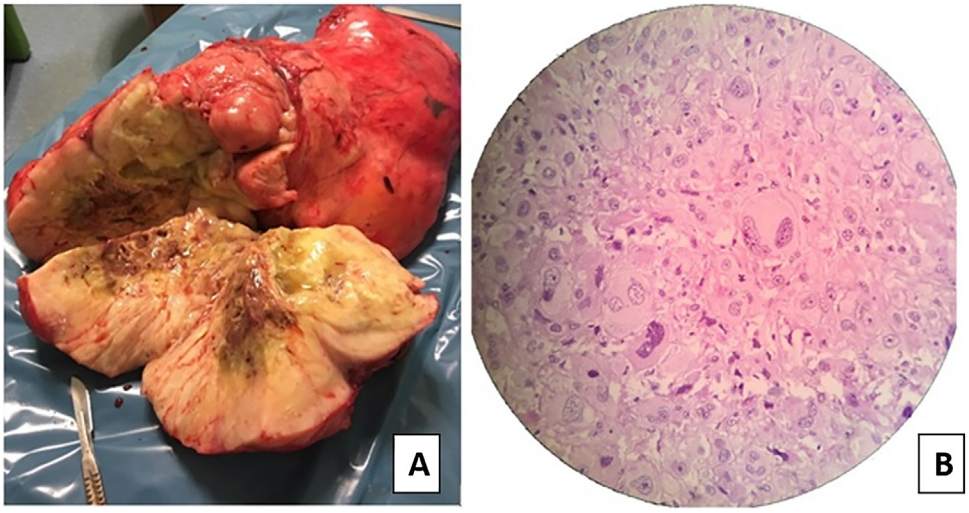

Fig. 2. (A) Macroscopic view of the tumor, measured 50 × 30 cm. (B) Microscopic view shows spindle-like cells and lipoblast cells.

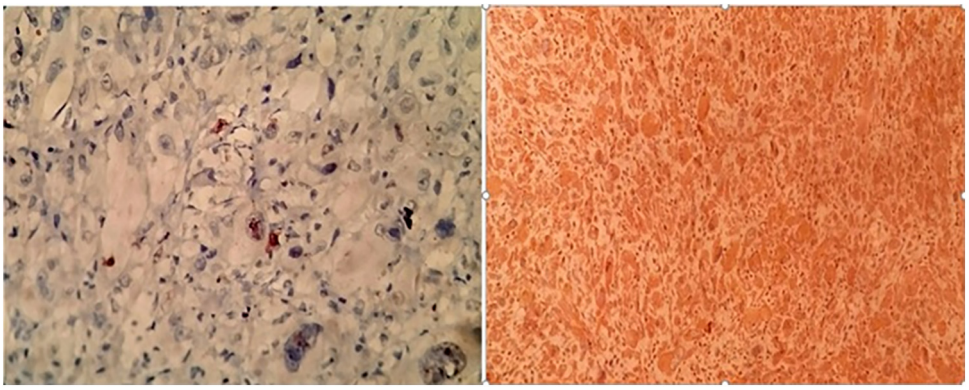

Fig. 3. Immunohistochemistry of S100 (left) and Vimentin (right).

the great blood vessels, it was not possible to remove all the tumor tissues, and we leave about 3 × 5 cm mass around innominate vein. Histopathologic findings revealed solid mass, composed of spindle-like cells forming herring-bone appearance, with enlarged nuclei and mitotic activity. In some spots, lipoblast cells were found (Fig. 2B). Immunohistochemistry staining of Pancytokeratin and CD-30 were both negative but turned out to be positive with Vimentin and S-100, confirming the diagnosis of primary dedifferentiated liposarcoma (Fig. 3). Post-operative chest X-ray showed the lung had fully re-expanded and symptoms resolved.

Because some little malignant tissues were still left in the mediastinum, the liposarcoma had high tendency for recurrence. Hence, we recommended the patient to undergo chemotherapy. Unfortunately, he never showed up until six months later, he came back with shortness of breath again. Chest x-ray and Thoracic CT scan revealed soft tissue mass and pleural effusion again in the left