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Case Report

Dedifferentiated Pleural Liposarcoma

Dediferansiye Plevral Liposarkom

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ABSTRACT

Ninety percent of primary pleural malignancies are malignant mesotheliomas. Pleural liposarcoma (LS) is a rare form of primary pleural malignancy. Among pleural LSs, the dedifferentiated subtype is even rarer and is considered a negative prognostic factor. In this report, we present the case of a 56-year-old male patient who was evaluated for the etiology of a right-sided pleural effusion. Over time, the disease progressed to involve the entire right hemithorax, circumferentially, with radiological findings resembling malignant mesothelioma. The diagnosis was confirmed by a tru-cut biopsy performed under thoracic ultrasonography guidance. This patient's prognosis remained poor during follow-up, and we present this case in light of the current literature.

Keywords: Liposarcoma, pleural malignancy, dedifferentiated pleural liposarcoma

ÖZ

Plevral primer malignitelerinin %90'ını malign mezotelyoma oluşturur. Plevranın liposarkomu oldukça nadir görülen bir primer plevral malignitedir. Plevral liposarkomlar içerisinde dediferansiye plevranın liposarkom ise daha da nadir görülen ve görülmesinin plevral liposarkomlar içerisinde negatif prognostik faktör olduğu kabul edilen bir alt tiptir. Bu yazıda; sağ plevral sıvı etyolojik incelenmesi için tetkik edilen, ilerleyen zaman içerisinde sağ hemitoraksı çepeçevre tutan ve radyolojik olarak malign mezotelyoma ile benzeşen, tanısını torasik ultrasonografi eşliğinde tru-cut biyopsi ile alan 56 yaşında takiplerinde prognozu çok kötü seyreden bir olgu güncel literatür eşliğinde paylaşıldı.

Anahtar Kelimeler: Liposarkom, plevral malignite, plevral dediferansiye liposarkom

INTRODUCTION

Liposarcoma (LS) is one of the most common types of sarcoma, accounting for 15-25% of all sarcoma types, and represents one of the most challenging entities in diagnostic pathology. The World Health Organization classifies LS into five main subtypes: well-differentiated LS (WDLS), dedifferentiated LS (DDLS), myxoid LS, pleomorphic LS (PLS), and myxoid PLS. These tumors most commonly originate from deep soft tissues, such as the retroperitoneal area, extremities, and intramuscular fascia. Intrathoracic LSs are extremely rare, accounting for only 1-2% of all LSs. LSs are aggressive tumors characterized by a high rate of recurrence and metastasis (1,2).

The exact incidence of primary pleural LS, a very rare pleural malignancy, remains unclear, as most cases have been reported as isolated case studies in the literature. It is believed to be more common in males and in individuals over the age of 50. The symptoms in most cases are non-specific, typically consisting of chest pain, cough, and dyspnea (3).

A review of the English literature reveals approximately 50 reported cases to date, and, due to its rarity in pulmonary practice, we present a case of dedifferentiated pleural LS located in the right hemithorax, accompanied by a brief review of the literature (4).

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CASE REPORT

A 56-year-old male patient presented to the emergency department with complaints of shortness of breath and was subsequently referred to the pulmonology clinic for further evaluation of pleural effusion (PE). His medical history included hypertension, coronary artery disease, and a prior cerebrovascular event. The patient had a 40 pack-year smoking history, with no notable family history. On physical examination, bilateral early inspiratory rales were auscultated. Laboratory test results are summarized in Table 1. A posteroanterior chest X-ray revealed PE in the right lung, volume loss in the right hemithorax, and multiple metastatic nodules in both lungs (Figure 1). Chest computed tomography (CT) demonstrated mediastinal pleural thickening in the right hemithorax, along with PE and multiple bilateral pulmonary nodules suggestive of metastasis (Figure 2).

Thoracentesis was performed under thoracic ultrasonography (USG) guidance. Cytological analysis of the pleural fluid revealed atypical cells suspicious for malignancy; however, the cell count was insufficient to establish a definitive diagnosis. A positron emission tomography-CT scan was performed, which showed multiple nodular lesions that were intensely hypermetabolic in both lungs (SUV_{max}: 9.4). In the right hemithorax, PE was noted along with intense hypermetabolism on the pleural surfaces, and a minimally hypermetabolic lesion was observed in the posterior aspect of the right lower lobe, measuring 47 mm (SUV_{max}: 12.5). In the mediastinum, intense hypermetabolic lymph nodes were detected in the right upper, bilateral lower paratracheal, subcarinal, and right hilar lymphatic stations, with the largest node located in the right hilar region, suggesting metastasis (SUV_{max}: 12) (Figure 3).

An informed consent form was obtained. For diagnostic purposes, a tru-cut biopsy of the right hemithorax was performed under thoracic USG guidance (Figure 4). Histopathological examination revealed tumoral cell infiltration embedded within dense fibrous stroma, with evidence of muscle and adipose tissue invasion at the periphery. The majority of the areas showed spindle cells without visible nucleoli. Extensive tumor necrosis was present. A comprehensive immunohistochemical panel was conducted, with no staining observed for smooth muscle actin, desmin, *SOX10*, cluster of differentiation 31 (*CD31*), *CD34*, Cam5.2, osteoclast associated receptor, cytokeratin, epithelial membrane antigen, D2-40, *SATB2*, WT1, *CALB2*, *STAT6*, anaplastic lymphoma kinase, *CD163*, immunoglobulin G4, *S100*, *CD45*, and *CD68*. Focal weak positivity was noted for cyclin-dependent kinase 4 and MDM2. The Ki-67 proliferation index was approximately 60% in the hotspot area. *BAP1* expression was retained. The differential diagnosis included pulmonary artery intimal sarcoma, sarcomatoid mesothelioma, DDLS, solitary fibrous tumor, malignant peripheral nerve sheath tumor, synovial sarcoma, inflammatory myofibroblastic tumor, malignant melanoma, IgG4-related diseases, histiocytic lesions, hematologic malignancies, extraskeletal osteosarcoma, and pleomorphic carcinoma with spindle cell features. Based on histopathological and immunohistochemical findings, the final diagnosis was consistent with DDLS (Figure 5).

A medical oncology consultation was requested for the case. However, as the patient and their relatives expressed that they do not wish to pursue oncological treatment despite initially considering it, the patient was advised to apply to the medical oncology outpatient clinic if they reconsider, and were subsequently discharged. Unfortunately, two months after the diagnosis, the patient passed away without receiving any oncological treatment.

Table 1. Laboratory values of the case

Parameter	Conclusion	RR
WBC (10^3 U/L)	12.4	4.00-10.00
Hemoglobin (g/dL)	6.6	13-17
Hematocrit (%)	22	40.00-54.00
Urea (g/dL)	35	16.6-48.5
Kreatinin (mg/dL)	0.39	0.7-1.2
ALT (U/L)	80	0-40
AST (U/L)	73	0-41
LDH (U/L)	224	135-225
CRP (µg/L)	138	0-5
Procalcitonin (mg/dL)	0.111	<0.5
Sedimentation mm/hour	26	0-15

ALT: Alanine aminotransferase, AST: Aspartate aminotransferase, CRP: C-reactive protein, LDH: Lactate dehydrogenase, mm: Millimeters, RR: Reference range, WBC: White blood cell

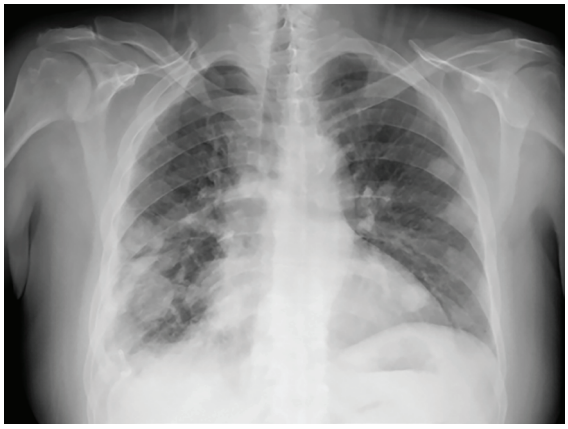

Figure 1. Lung X-ray of the case

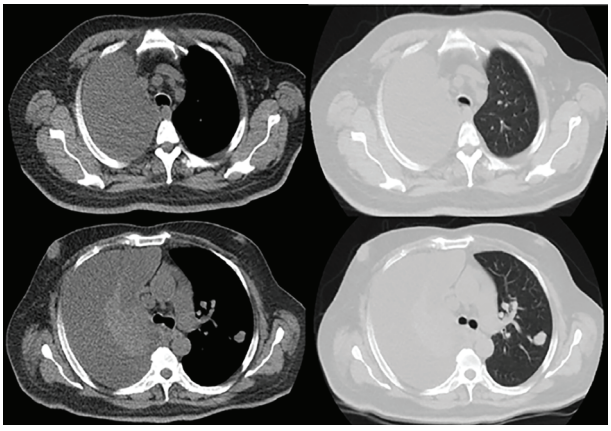

Figure 2. A computed tomography scan: mediastinal pleural thickening in the right hemithorax, circumferential involvement of the pleura in the right hemithorax, pleural effusion and multiple bilateral metastatic nodules

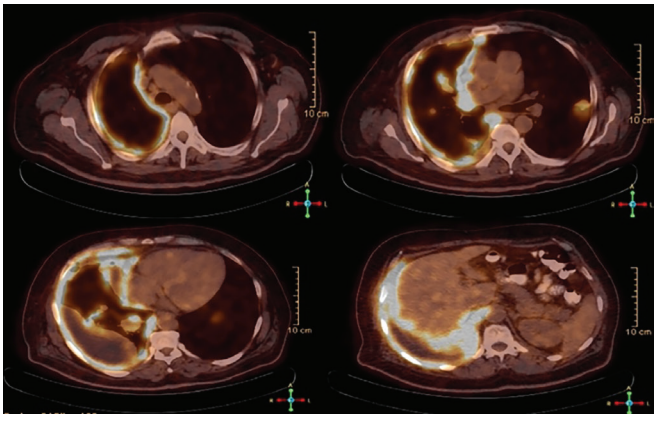

Figure 3. Positron emission computed tomography of the case

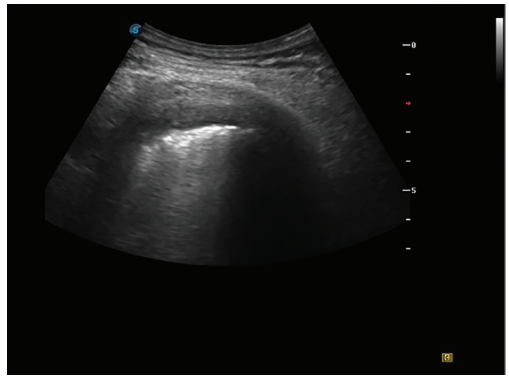

Figure 4. Appearance of thickened pleura on thoracic ultrasonography

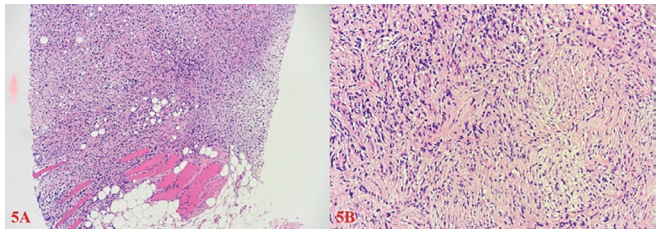

Figure 5. A) The tumor shows infiltration of adipose and muscle tissue at its periphery (hematoxylin and eosin x200). **B)** Within the fibrous stroma, tumoral infiltration with spindle-shaped cells and hyperchromatic nuclei lacking visible nucleoli was observed (hematoxylin and eosin x400)

DISCUSSION

This article presents a case of a 56-year-old male pleural LS, a rare and aggressive malignancy characterized by frequent recurrence and metastatic potential, which was diagnosed through a tru-cut biopsy guided by thoracic USG.

Malignant mesothelioma accounts for the majority of pleural malignant lesions (90%). The remaining cases include rare entities such as PLS, malignant solitary fibrous tumor, localized malignant mesothelioma, pleuropulmonary blastoma, synovial sarcoma, angiosarcoma, and pleural lymphoma. Although the exact etiology of PLS remains unclear, it is believed to arise from residual primitive mesenchymal cells. Additionally, although not definitive, the malignant transformation of pre-existing pleural lipomas is also suspected (5).

Upon reviewing the patient's chest CT from two years prior to admission, no findings suggestive of lipoma, pleural lipoma, or pleural pathology were identified. The most comprehensive study to date on pleural LS was conducted by Kawai et al. (4) and published in 2023. In their review of 45 reported cases, the authors aimed to evaluate the clinicopathological and immunohistochemical

characteristics of PLS and to identify prognostic factors. The results of the study indicated that the average patient age was 51, and 66% of cases were male. Dedifferentiated PLS was identified as the rarest histologic subtype with only five reported cases. *CDK4-MDM2* positivity was emphasized as a key marker for definitive diagnosis. The WDLS subtype was associated with a more favorable prognosis. Our case involved a 56-year-old male with *CDK4-MDM2* positivity and was consistent with dedifferentiated PLS, a rare and aggressive variant. This case supports previous findings highlighting the diagnostic value of *CDK4/MDM2* immunostaining and the poor prognosis associated with the dedifferentiated subtype.

Due to the rarity of PLS, it is not possible to provide definitive data regarding average tumor size, tissue density, or SUV_{max} values based on the current literature. In a study by Matsukuma et al. (6), which included their own case report and a review of 31 PLS cases, tumor sizes were found to range from 3.5 cm to 29 cm. Similarly, Chen et al. (7) reported tumor sizes ranging from 4 cm to 39 cm in a series of 23 cases. SUV_{max} values were reported to vary between 3.3 and 10.2 (5,6). Regarding CT density, Carrillo et al. (8) reported a density of 18 Hounsfield units (HU) in one PLS case on chest CT, while Prabhakar et al. (9) observed a density of 10 HU in their cases. Our case involved a dedifferentiated PLS, circumferentially surrounding the right hemithorax, with a maximum diameter of 10 cm, an SUV_{max} of 12.5, and a CT density of 18 HU.

One of the largest studies on PLS, conducted by Matsukuma et al. (6), emphasized that dedifferentiated histology is a negative prognostic factor. A review of the literature suggests that overall survival in PLS cases ranges between seven months and eight years (10). Our patient did not receive any oncological treatment and passed away two months after the diagnosis. Following the oncology consultation, no oncological treatment was planned for the patient due to the refusal by the patient and their relatives. The primary reasons for this decision may include the patient's poor overall performance status at the time of diagnosis, the presence of metastatic lesions in both lungs, and a high tumor burden in the right hemithorax. As emphasized in the literature, this is consistent with the limited response to treatment and generally poor prognosis observed in cases of dedifferentiated pleural LS (11).

CONCLUSION

In conclusion, the most common primary malignancy of the pleura is malignant mesothelioma. Although rare, other primary pleural malignancies that may radiologically mimic mesothelioma can also be encountered. There is a significant gap in the literature regarding these rare entities. Studies involving large case series of pleural malignancies, such as PLS, are needed to better understand their clinical, radiological, and pathological characteristics. However, even individual case reports, like this one, provide valuable contributions to the growing body of literature on rare pleural tumors.

ETHICS

Informed Consent: An informed consent form was obtained.

FOOTNOTES

Authorship Contributions

Surgical and Medical Practices: D.Y., Concept: C.D., Design: C.D., B.B., Data Collection or Processing: D.Y., A.N.T.Y., Analysis or Interpretation: B.B., Literature Search: D.Y., Writing: C.D.

Conflict of Interest: No conflict of interest was declared by the authors.

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