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Case Report

Primary pulmonary dedifferentiated liposarcoma: A case report and literature review

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ABSTRACT

Liposarcoma is a relatively common soft tissue sarcoma, but primary pulmonary liposarcomas are extremely rare, especially for the dedifferentiated subtype. We report the case of a 44-year-old African woman, healed from SARS-CoV-2, who presented a 8 cm right lung hilar mass and underwent a right intrapericardial pneumonectomy with diaphragmatic and pleural resection. The histopathologic examination confirmed the diagnosis of dedifferentiated primary pulmonary liposarcoma. The patient recovered well having an uncomplicated postoperative course. This case study confirms the effectiveness of pneumonectomy as treatment for primary pulmonary dedifferentiated liposarcoma. However, more studies have to be conducted on its best treatment because very few cases are reported in literature.

KEYWORDS: liposarcoma, primary pulmonary liposarcoma, dedifferentiated liposarcoma, liposarcoma of the lung, rare lung cancer.

ABBREVIATIONS

SARS-CoV-2	:	Severe Acute Respiratory Syndrome Corona Virus 2
CT	:	Computed Thomography
EBUS	:	Endo-Bronchial Ultra-Sound
^{18}F-FDG PET/CT	:	^{18}F-FluoroDeoxyGlucose Positrone-Emission Tomography/Computed Tomography
SUV	:	Standardized Uptake Value
Fr	:	French
S100+	:	S100 protein positive
MDM2+	:	MDM2 protein positive

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INTRODUCTION

Primary pulmonary liposarcoma (PPL) is an extremely rare malignancy with very few cases reported in literature. Intrathoracic liposarcomas are uncommon, and generally located in the mediastinum. Malignant degeneration of a pulmonary lipoma and pleuropulmonary asbestosis have been considered as possible pathogenetic factors [1]. Of all the subtypes, the dedifferentiated liposarcoma is the most uncommon with only four cases reported in literature.

CASE REPORT

A 44-year-old Moroccan woman came to our hospital because of recurring hemoptysis. About a month before, she underwent a chest X ray for a suspected SARS-CoV-2 pneumonitis (Fig. 1) showing a voluminous right lung opacity. Afterwards, chest CT scan demonstrated a right lung hilar mass (7.2 cm) which was initially treated with antibiotic. An EBUS was performed without bioptic sample

because of a suspected Echinococcus disease. Furthermore, a transthoracic biopsy was performed, but samples were not adequate because of necrotic tissue. During hospitalization she developed pain, high fever and increased inflammation indices; consequently she was treated with analgesic and antibiotic therapy. A total-body CT scan (Fig. 2) showed an increment of known mass (8 cm), and a total-body ^{18}F-FDG PET/CT (Fig. 3) reported an uneven metabolic activity, in particular at the origin of the upper right bronchus (SUVmax = 5.84) and at the middle right bronchus very close to the biforcation with the lower bronchus (SUVmax = 6.03). Having excluded other methods to perform a diagnosis, it was decided to perform a right thoracotomy. Intraoperative frozen section confirmed malignancy of the mass, and hence it was decided to perform a right pneumonectomy with diaphragmatic and pleural resections due to their involvement (Fig. 4, 5). Bronchial stump was covered by intercostal muscle previously retrieved. A 32 Fr chest drain was placed. Postoperative period was uneventful, the chest drain was removed after 2 days and the patient was discharged after 7 days. On 10 days follow up the patient was still asymptomatic with normal outcomes after pneumonectomy. Histological examination confirmed a primary pulmonary dedifferentiated liposarcoma with S100+ and MDM2+ as markers. On twenty-four months follow up, the patient was still alive and without evidence of recurrence disease.

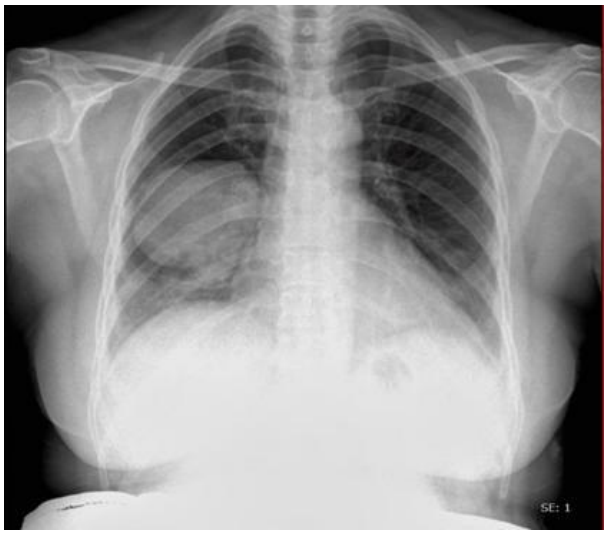

Fig. 1. Chest-X ray showing a right large opacity.

DISCUSSION

The first case of PPL was reported by Latienda in 1946 [2]. Till then very few cases have been reported in the literature. This tumor originates

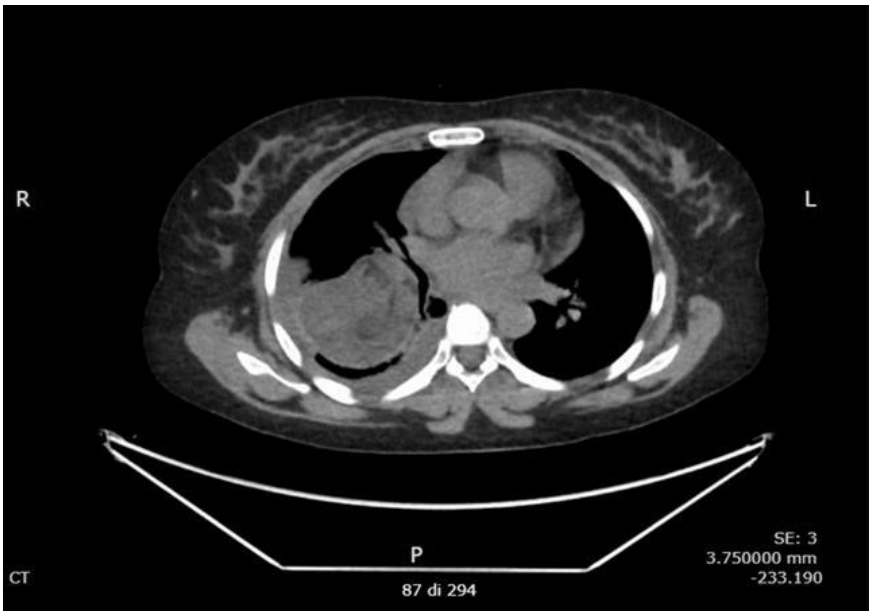

Fig. 2. Chest-CT showing a large mass with an uneven density.

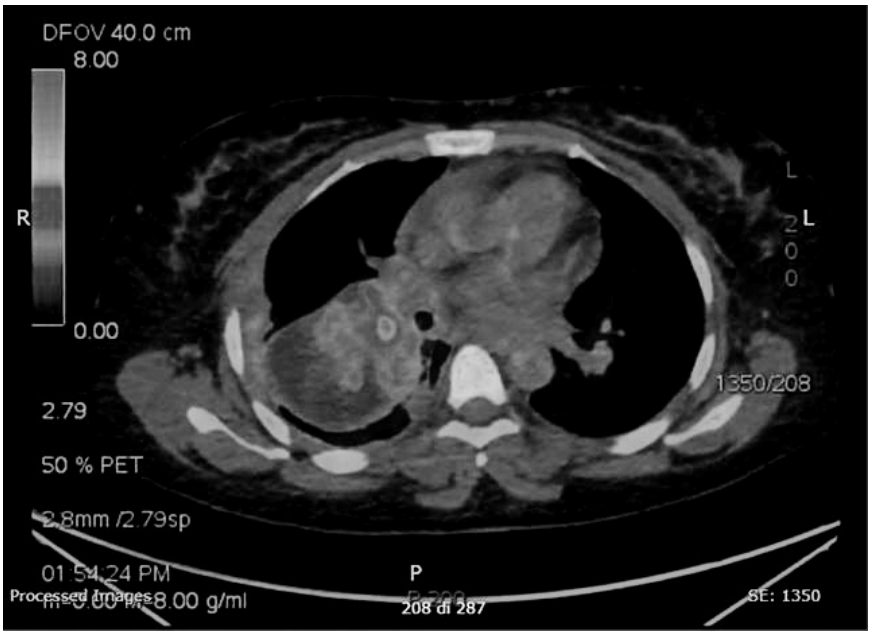

Fig. 3. PET scan showing a very low metabolic activity.

Fig. 4. Right lung.

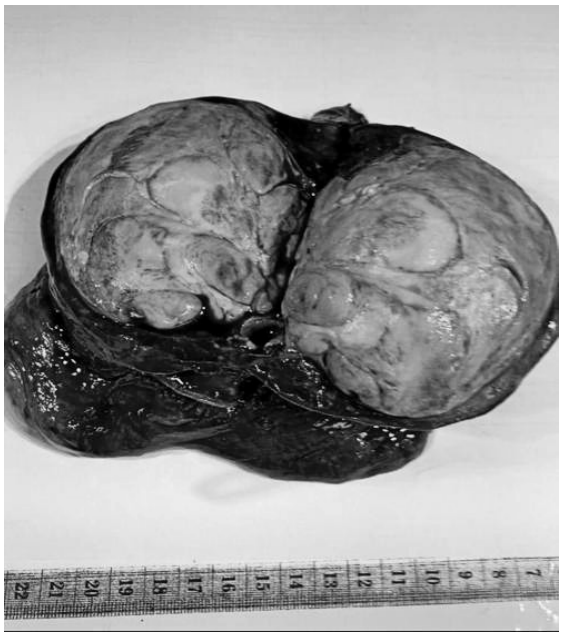

Fig. 5. Macroscopic view of the tumor sectioned in the median plane.

from primitive mesenchymal cell and it is commonly found out in the extremities and retroperitoneum.

PPL generally originates in the mediastinum and rarely arises from the lung tissue, chest wall or pleura. Possible risk factors associated with the development of PPL are pleuropulmonary asbestosis or malignant degeneration of pulmonary lipoma [3]. In our case the patient had domestic asbestos exposure, but there was no sign of asbestosis in her lungs.

Table 1. Literature review.

Author	Age	M/F	Tumor site	Tumor size (cm)	Subtype	Lung resection	Rec	Mets	Adj	DFS (m)	Status
Loddenkemper	49	F	LLL	9	DPPL	L	No	No	None	16	A
Chen	59	F	NA	7	DPPL	L	Yes	No	RT	6	D
Longano	74	M	LUL	10	DPPL	L	No	No	None	12	A
Our case	44	F	RLL	8	DPPL	P	No	No	None	24	A

M: male; F: female; LLL: left lower lobe; NA: not available; LUL: left upper lobe; RLL: right lower lobe; DPPL: dedifferentiated primary pulmonary liposarcoma; RT: radiotherapy; DFS: disease free survival; m: months; A: alive; D: dead; L: lobectomy; P: pneumonectomy; Rec: recurrence; Mets: metastasis; Adj: adjuvant treatment.

No pathognomonic signs and symptoms are attributable to PPL but they are the same as any other pulmonary tumors, such as cough, sputum production, hemoptysis, dyspnea, chest pain and loss of weight. This nonspecific symptoms can lead to further delays in diagnosis. Also, because these tumors are slow growing, they often present as a large mass [4, 5]. Diagnosis is based on the histological analysis.

The World Health Organization (WHO) classifies liposarcomas into four subtypes [6]:

- Atypical well-differentiated, probably benign
- Myxoid/round cell, locally aggressive frequently with metastasies
- Dedifferentiated, very aggressive and highly malignant
- Pleomorfic, malignant

In literature, we found only four cases of dedifferentiated PPL [7-9] (Table 1). In those patients the median age was 56.5 and M:F ratio was 1:3. Tumor size ranged from 7 to 10 cm, without any preference of pulmonary localization. Three patients underwent a lobectomy; our case underwent a right pneumonectomy extended to the pleura and diaphragm due to their neoplastic involvement. Only one patient [7] had a recurrence disease and was treated with radiotherapy. The same patient died after 6 months after diagnosis. The other patients had absence of disease at follow up. None of the patients had metastasies.

According to other authors [7-9], radical excision with lymphonodal dissection is the best treatment for PPL, and when R0 resection is obtained, no other treatment is required. Chemotherapy and radiotherapy as adjuvant treatment have not shown great success even in patients with residual disease, inoperable cases or metastatic disease.

CONCLUSION

PPL is an extremely rare tumor with an attitude to lead to a local invasion rather than generate a metastatic disease. The best treatment is surgical margin free excision, even if it is a local advanced tumor. Till now, radiotherapy and chemotherapy are believed to be ineffective therapeutic modalities for improving the survival. In the light of the small number of cases reported in the literature, more data have to be collected in order to have some more evidences to establish the best treatment and the potential role of the new oncologic drugs.

CONFLICT OF INTEREST STATEMENT

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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