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
CT : Computed Thomography
EBUS : Endo-Bronchial Ultra-Sound
¹⁸F-FDG PET/CT : ¹⁸F-FluoroDeoxyGlucose Positrone-Emission Tomography/Computed Tomography
SUV : Standardized Uptake Value
Fr : French
S100+ : S100 protein positive
MDM2+ : MDM2 protein positive

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 hemoptyis. 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 

Because of the suspected Echinococcus disease, a transthoracic biopsy was performed, but the samples were not adequate due to necrotic tissue. During hospitalization, the patient 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 increase in the size of the known mass (8 cm), and a total-body $^{18}$F-FDG PET/CT (Fig. 3) reported uneven metabolic activity, particularly at the origin of the upper right bronchus ($\text{SUVmax} = 5.84$) and at the middle right bronchus very close to the bifurcation with the lower bronchus ($\text{SUVmax} = 6.03$). 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 $\text{S100}^+$ and $\text{MDM2}^+$ as markers. On twenty-four months follow up, the patient was still alive and without evidence of recurrence disease.

**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

![Chest-X ray showing a right large opacity.](image1)

![Chest-CT showing a large mass with an uneven density.](image2)
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.
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.

**REFERENCES**


