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Rare Case Of Primary Pulmonary Dedifferentiated Liposarcoma

Tuoro A[1*], Aversa V[2] and Cassina EM[1]

[1]Thoracic Surgery Unit, "San Gerardo" Hospital, Monza, Italy

[2]University "Magna Graecia" of Catanzaro, Catanzaro, Italy

[*]Corresponding author:

Antonio Tuoro,
Thoracic Surgery Unit, "San Gerardo" Hospital,
Monza, Italy, E-mail: aversa.vincenzo@hotmail.it

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1. Abstract

Liposarcoma is a relatively common soft tissue sarcoma, but primary pulmonary liposarcomas are extremely rare, expecially for dedifferentiated subtype. We report the case of a 44-year-old African woman, healed from SARS-Covid, who presented with a 8 cm right lung ilary mass and underwent to a right intrapericardial pneumonectomy with diaphragmatic and pleural resections. Histological test confirmed diagnosis of dedifferentiated primary pulmonary liposarcoma. The patient recovered well having an uncomplicated postoperative course and was discharged after 7 days. On four months follow up the patient was still alive and without evidence of disease. More studies have to be conducted because very few cases are reported in literature.

2. Introduction

Primary pulmonary liposarcoma 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 subtypes, dedifferentiated is the most uncommon with only four cases descripted in literature.

3. Case Presentation

A 44-year-old moroccan woman arrived to our Hospital because of recurring hemoptysis. She underwent about a month before to a Torax-TC that exposed a right lung ilary mass (7,2 cm) treated with antibiotic. An EBUS was performed without bioptic sample because of suspected Echinococcus disease. Once excluded, a transthoracic biopsy was performed, but samples were not clinically adeqate because of nectotic tissue. During hospitalization she developed pain associated with hyperpyrexia and increased inflammation indices, treated with analgesic and antibiotic therapy. A Body-TC [Figure 1, 2] exposed an increment of known mass (8 cm), and body-PET [Figure 2] reported an uneven captation, in particular at the origin of upper right bronchus (SUVmax = 5.84), at middle right bronchus (SUVmax = 6.09) and at middle right bronchus biforcation (SUVmax = 6.03) and no other place worth of notice. So it was decided to perform a right thoracotomy in order to reach the mass. Extemporaneous histological test confirmed malignity of the mass, so it was dedided to perform a right pneumonectomy with diaphragmatic and pleural resections due to infiltration [Figure 4, 5]. Bronchial stump was covered by intercostal muscle previously isolated. A 32 CH thoracic dreinage was placed. Postoperative period was uneventful, drainage was removed after 2 days and the patient was discharged after 7 days asynthomatic and in good general conditions. On 10 days follow up the patient was still asynthomatic with normal outcomes of pneumonectomy. Histological test confirmed for primary pulmonary liposarcoma dedifferentiated with S100+ and MDM2.

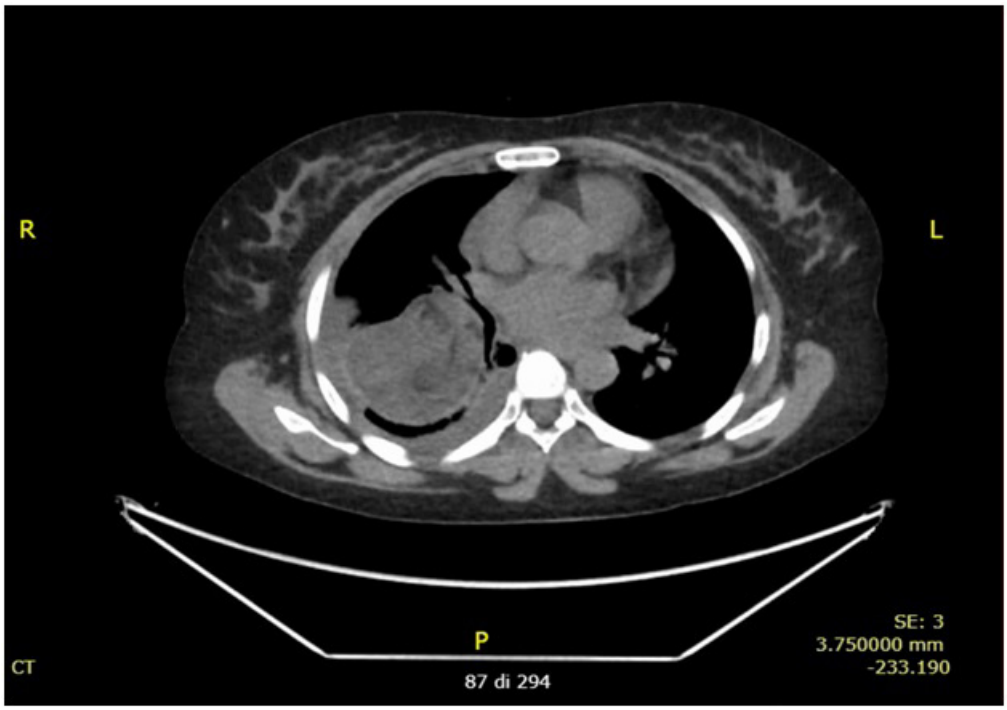

Figure 1: body-TC showing large mass uneven capitation.

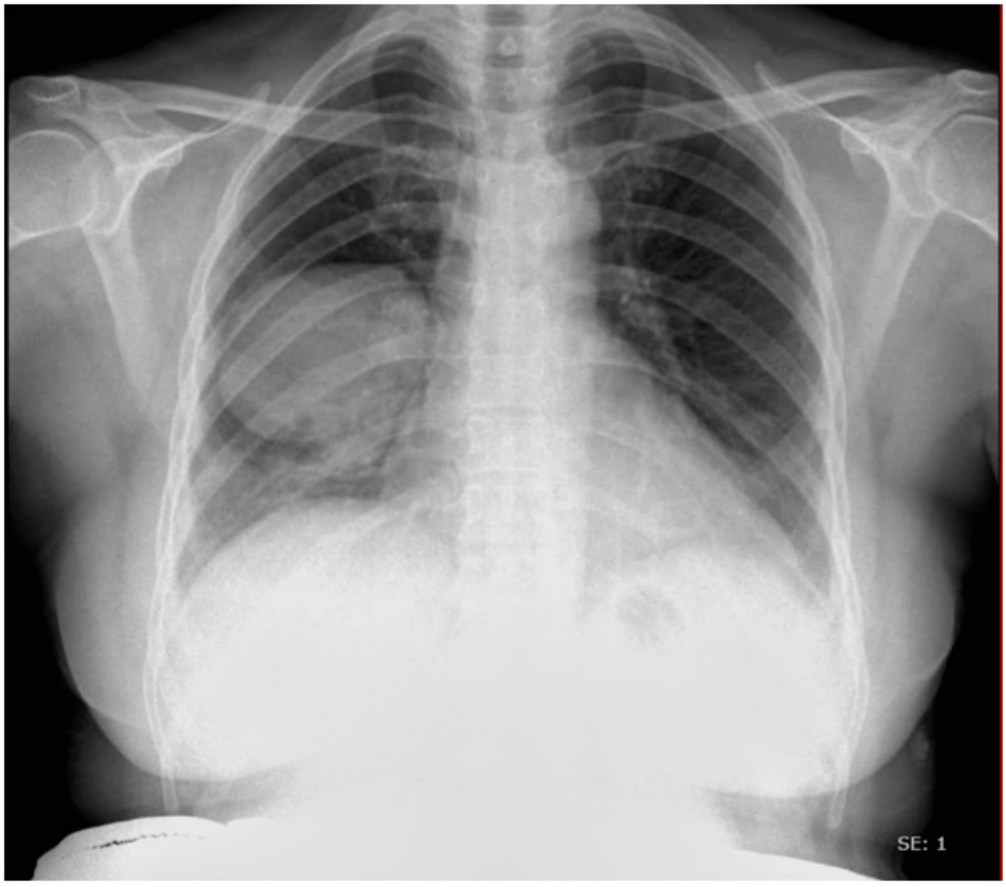

Figure 2: Thorax-RX.

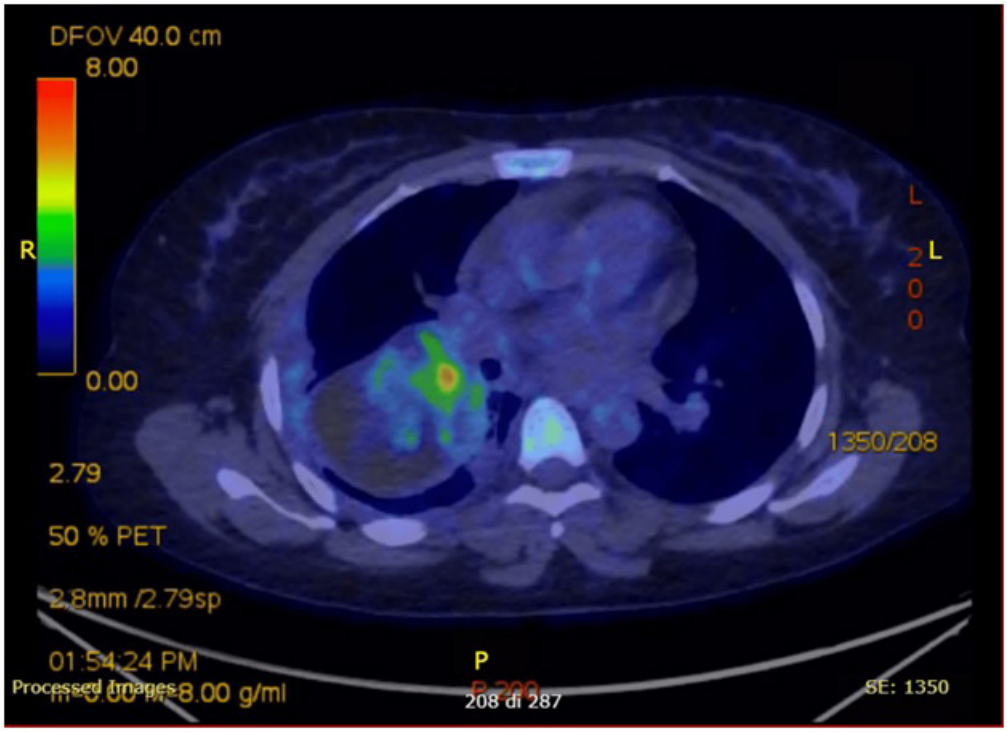

Figure 3: PET showing.

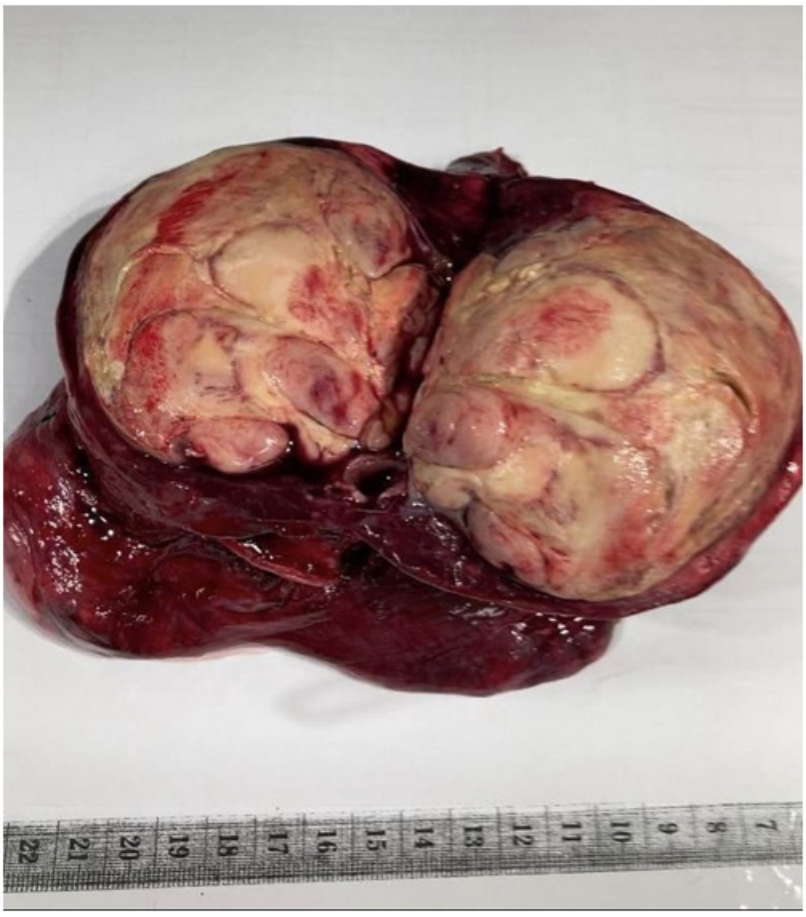

Figure 4

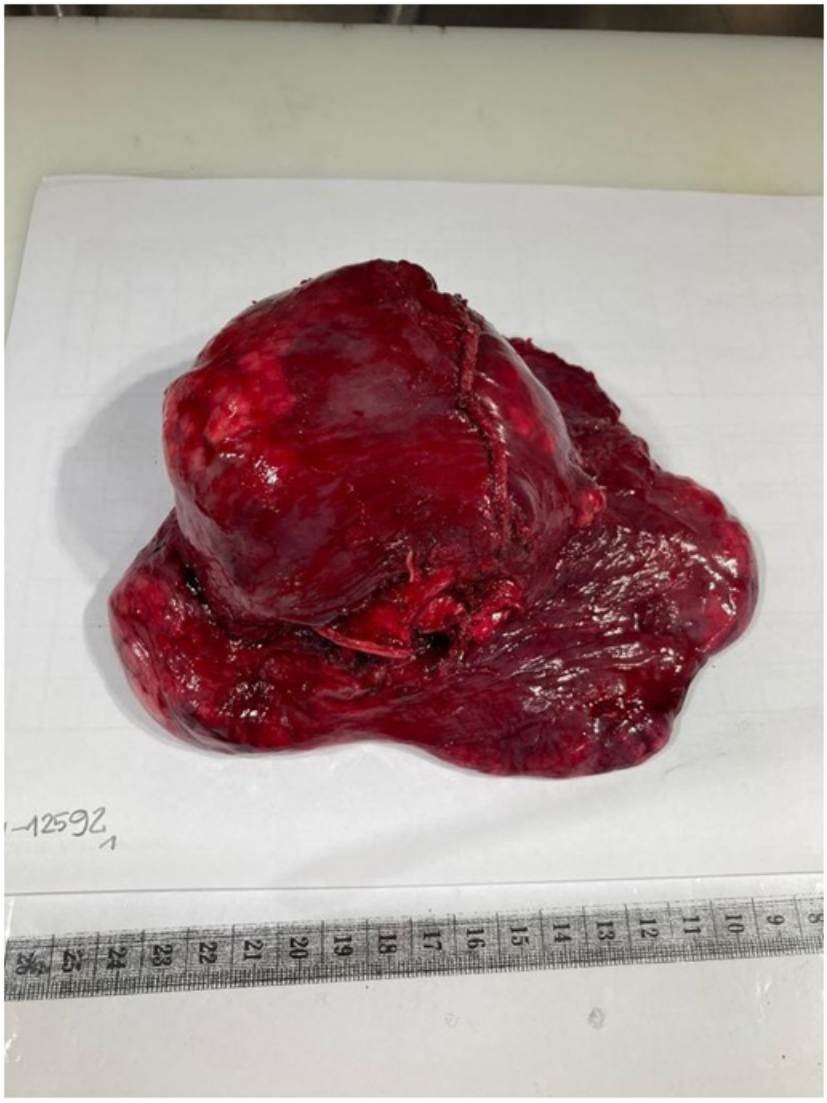

Figure 5

4. Discussion

The first case of PPL was reported by Latienda in 1946 [2]. Till then very few cases have been reported in literature. It originates from primitive mesenchymal cell and is common in extremities and retroperitoneum. PPL generally originates in the mediastinum, rarely arises from parenchyma, chest wall or pleura. Possible risk factors associated with development of primary pulmonary liposarcoma are pleuropulmonary asbestosis or malignant transformation of pulmonary lipoma [3]. In our case the patient had contact with asbestos because of the house where she lived, but there was no sign of asbestosis. No particular characteristic symptoms and signs are attributable to PPL but are the same of any other pulmonary tumors, such as cough, sputum production, hemoptysis, dyspnea, chest pain, and loss of weight and appetite. This can lead to further delays diagnosis. Also, because these tumors are slow growing, they often present as a large mass [4]. Diagnosis is based on histological test. The World Health Organization (WHO) classifies liposarcomas into four subtypes [5]:

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

In literature we found only four cases of dedifferentiated Primary pulmonar liposarcoma [6,7,8] **[**Table 1**]**. In those patients the middle age was 56,5 and the ratio M:F of 1:3. Tumor size was from 7 to 10 cm, without particular pulmonary localization. Three patients underwent to lobectomy, our case underwent to pneumonectomy because of pleural and diaphragmatic infiltration. Only one patient had recurrency and was treated with RT therapy. The same patient died after 6 months after diagnosis. Other patients had absence of disease at follow up after 16, 12 and 4 months rispectively. None of the patient had metastasies. According to other authors [recensioni trattamento], radical excision with lymphnode dissection is the best treatment for PPL, and when R0 resection is obtained, no other treatment is required. Chemotherapy and radiotherapy as adjuvant treatment haven't shown great success even in patients with residual disease, inoperable cases or metastasies.

Table 1

Author	Age	Gender	Localizzation	Tumor Size	Subtype	Intervent	Reccurrence	Metastasies	Treatment	DFS months	Status
Loddenkemper	49	F	LLL	9,00 cm	Dediff pulmonar	Lobectomy	no	no	none	16	AoD
Chen	59	F	NA	7,00 cm	Dediff pulmonar	Lobectomy	yes	no	RT	6	Dead
Longano	74	M	LUL	10,00 cm	Dediff pulmonar	Lobectomy	no	no	none	12	AoD
Tuoro	44	F	RLL	8,00 cm	Dediff pulmonar	Pneumonectomy	no	no	none	4	AoD

5. Conclusion

Primary polmunar liposarcoma is a extremely rare disease with potential of local complications and distant metastases. The best treatment is radical excision. In consideration of the small number of cases reported in the literature, more studies have to be conducted in order to have more adequate guidelines.

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