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Primary Myxoid Liposarcoma of the Middle Mediastinum About a Case (Primary Myxoid Liposarcoma of the Middle Mediastinum)

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Abstract **Case Report**

Myxoid liposarcomas "LPS" are rare tumors, especially in their mediastinal location. We report a case of a myxoid liposarcoma of the middle mediastinum, in a 51-year-old patient, with no particular pathological history, admitted with a NYHA stage V dyspnea picture. A chest X-ray made objectifying a mediastinal widening of water density, with external edges convex towards the lung and internal edges merged with the mediastinum, the chest CT scan of which links it to a lesion process of the middle mediastinum well defined with clear heterogeneous contours of mixed tissue, fat and water density without calcifications, The punctureCT-guided transparietal biopsy with the result of an anatomopathological study is compatible with a myxoid liposarcoma. Myxoid LPS of the mediastinum is a very rare entity and a few sporadic cases have been reported in the literature. Surgery appears to be the treatment of choice. Radiotherapy and chemotherapy retain their place in very specific indications.

Keywords: Liposarcoma, Rare Tumor, Myxoid, Rare Location, Middle Mediastinum, Rare, Chest Scan.

INTRODUCTION

Liposarcomas (LPS) are rare tumors, representing approximately 15 to 20% of all sarcomas. They occur most often in the lower limbs and less in the retroperitoneum. Mediastinal localization remains exceptional, comprising less than 1% of all mediastinal tumors [1]. We report the case of a myxoid liposarcoma of the middle mediastinum with a review of the literature.

PATIENT AND OBSERVATION

A 51-year-old patient, with no particular pathological history, not a known smoker, admitted with NYHA stage V dyspnea.

Physical examination found a conscious patient with respiratory unstable HR 65 cycles/min, Sao2 95% on oxygen, and signs of respiratory struggle.

Hemodynamically stable.

In whom a frontal chest X-ray shows a well-circumscribed opacity of water tone, starting in the mediastinum with a bilateral endothoracic component more marked on the right with external edges convex towards the lung and internal edges merging with the mediastinum. The chest CT scan without and after injection of contrast product shows a heterogeneous lesion process of the middle mediastinum of mixed tissue density, fat and water in places without calcifications, well limited and with clear contours. It pushes back and compresses the heart forward as well as the right pulmonary artery which remains permeable with the main bronchi and their bronchi and behind compresses the esophagus, the descending aorta and the prevertebral spaces, below rests on the diaphragm which remains intact with above it compresses with elongation of the aortic arch this mass is responsible for a collapse of the middle lobe, right basal and lingular with a diffuse interstitial syndrome (Figure 1). The extension assessment (abdominal CT scan) is without abnormality. The transparietal biopsy puncture guided by the radiology department reveals a morphological appearance of a mesenchymal proliferation with a myxoid background and a lipoblastic component suggesting a myxoid liposarcoma.

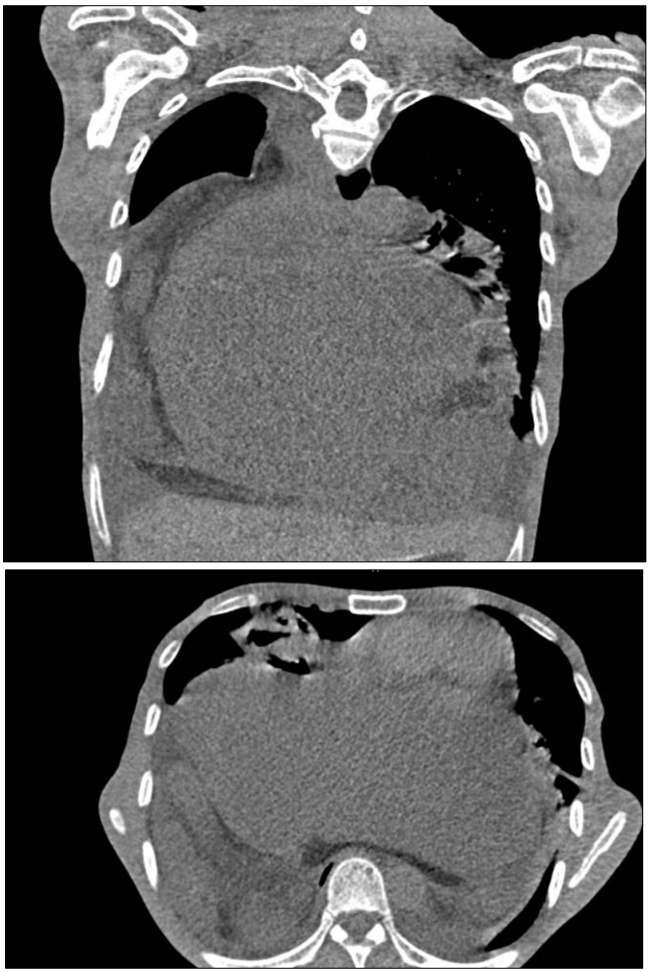

Figure 1: Thoracic CT scan: hetero-dense tissue mass occupying the right hemi-thorax, 10x9x6cm, site of areas of fatty density, and pushing back the VCS, and coming into contact with the trachea, the esophagus, and the vertebral bodies without bone lysis, Mohammed VI University Hospital, Marrakech

DISCUSSION

Liposarcomas are soft tissue sarcomas with adipocytic differentiation. They represent approximately 15% of malignant soft tissue tumors in adults and the myxoid variety represents 40 to 50% of LPS, all locations combined [1, 2]. They usually occur in the lower limbs (75%) and less frequently in the retroperitoneum [2]. Primary mediastinal liposarcomas, with less than 200 cases reported in the literature to date, are extremely rare tumors, representing less than 1% of all mediastinal tumors [2]. Myxoid LPS generally affects adults with a peak incidence during the 4th and 5ththdecade. Myxoid LPS can be asymptomatic and discovered radiologically in approximately 12% of cases [4]. It becomes symptomatic by the compression it causes on the mediastinal structures while it has acquired a significant volume, in particular on the tracheobronchial axis, the heart and the vessels, and the esophagus. This results in a functional limitation such as cough, dyspnea, wheezing, wheezing, dysphagia, angina pectoris or superior vena cava syndrome [4].

Radiological images of LPS are not specific but suggestive of the diagnosis. Chest radiography usually shows mediastinal widening, or deviation of the tracheal axis or vessels [2].

On chest CT scan: mediastinal LPS appears as a heterogeneous, moderately limited mass containing areas of mixed fatty and solid density. With possible areas of necrosis.

Magnetic resonance imaging (MRI) can provide determining elements in resectability, and precisely show the limits of the tumor with the elements of the mediastinum, its locoregional extension.

Differential diagnoses are immature teratomas, lymphomas [2-4]. The diagnosis of myxoid LPS is histological, by the discovery of a tumor with myxoid stroma, presenting a vascularization with a characteristic "branched" appearance consisting of small vessels and the presence of lipoblasts, the absence of nuclear atypia, and rare mitoses [1].

The WHO classification recognizes 5 categories of LPS which are distributed as follows [2]. (1) Well-differentiated LPS (adipocyte-type LPS (atypical lipoma), sclerosing LPS, inflammatory LPS

spindle cell LPS); (2) Dedifferentiated LPS; (3) Myxoid round cell LPS; (4) Pleomorphic LPS; (5) Combined mixed LPS. Myxoid and well-differentiated LPS are low-grade sarcomas of malignancy, most often encapsulated and rarely metastasize. While round cell, pleomorphic and dedifferentiated LPS are of high malignancy with a tendency to local recurrence and metastasize early [1-4]. Due to the low incidence of mediastinal LPS, therapeutic strategies are extrapolated from similar tumors in other locations.

The treatment of myxoid LPS is radical surgical resection with negative margins (R0), One centimeter is chosen as a threshold in some studies. Furthermore, these tumors tend to invade adjacent structures making their complete excision difficult [3-5]. Surgery also remains the treatment of choice for local recurrences [5].

The interest of systematic complementary radiotherapy and chemotherapy in the event of complete resection is controversial. It remains reserved for poorly differentiated forms, unresectable tumors and recurrent tumors [1-7].

CONCLUSION

Myxoid liposarcomas of the middle mediastinum are very rare tumors. The presumptive diagnosis can be obtained by radiological investigations and the confirmation is histological. Radical surgery remains the treatment of choice in patients with myxoid liposarcoma and allows to obtain the best survivals

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