

Primary Mediastinal Liposarcoma

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Summary

Liposarcomas (LPS) are uncommon and present diagnostic and therapeutic difficulties. Computed tomography and magnetic resonance imaging allow for a diagnostic orientation, allowing differentiation from other more common tumors of the anterior

mediastinum. Surgery is the treatment of choice and has the advantage of being curative. We report a case of well-differentiated liposarcoma of the anterior mediastinum.

Key Words: Liposarcoma, Mediastinum, Surgery

Introduction

Liposarcomas (LPS) are very rare tumors especially in the mediastinum. They are characterized by a high histologic and prognostic heterogeneity and present lots of diagnostic challenges because of their typical clinical latency, the absence of specific radiological data, but also therapeutic challenges because of the lack of a consensus outside the classic cancer surgery even in the face of recurrence.

We report a case of well-differentiated liposarcoma of the mediastinum

Case Report

A 52 years old man of general good health presented with a two months history of chest pain and dyspnea. A chest radiograph was unremarkable. However a chest computed tomography (CT) scan showed an anterior mediastinal tumor of tissue and fatty components with minimal bilateral pleural and pericardial effusions. The patient underwent an anterior mediastinotomy and the mass biopsied. Histology confirmed the lesion was a liposarcoma. After staging, and in consultation with the Thoracic Oncology Group, a surgical resection of the mass, the thymus and involved surrounding structures including the pericardium, the right and left mediastinal pleura, the innominate trunk and left ventral segment of the right upper lobe was performed by sternotomy. Histological analysis of the surgical specimen that measured 14cm x 11cm x 3.5 cm and weighed 191g, found a tumor process of fatty differentiation with immunologic labeling; MDM2 positive, confirming a well-differentiated liposarcoma grade 1 (according FNLCC). The

immediate post-operative period was marked by an infection of the surgical site without mediastinitis. This was managed locally and with antibiotics. Adjuvant radiotherapy is ongoing. Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Figure 1 : Chest CT axial section: tumor mass of thymic compartment dual component (tissue and fat) associated with dense septa.

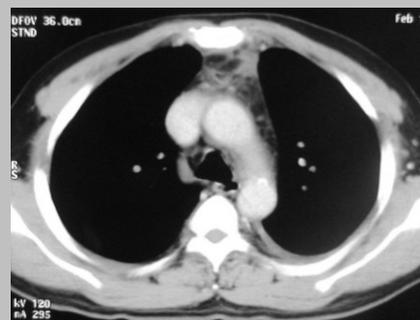


Figure 2 : Chest CT axial section: anterior mediastinal tissue mass infiltrating the pericardium with bilateral pleural effusion.

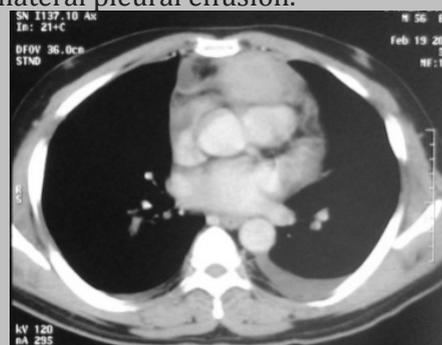


Figure 3 : Well differentiated liposarcoma (HES X 10) tumor proliferation of immature fat cells “lipoblasts” associated with an inflammatory infiltrate.

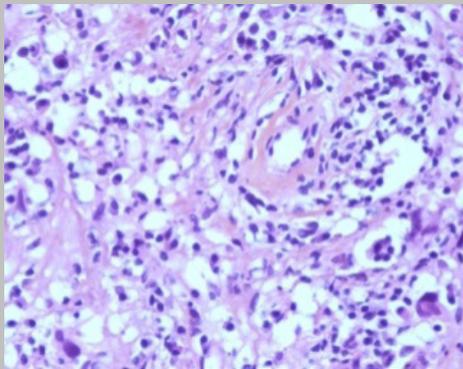
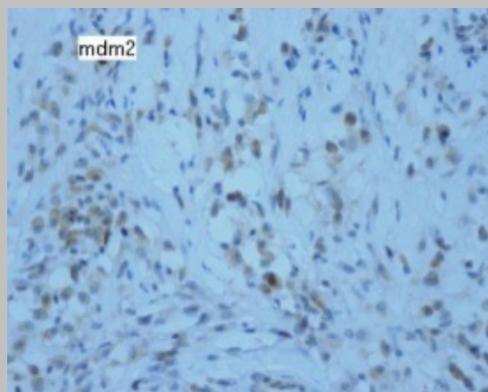


Figure 4 : Positive immuno marking by MDM2



Discussion

Sarcomas are rare mesenchymal tumors representing less than 1 % of all malignancies. Liposarcomas are however, one of the most common subtypes of adult soft tissue sarcomas. They are rarely found in the mediastinum (1). Liposarcomas do not arise from malignant transformation of an existing lipoma despite being found in previous lipoma excision sites (2). Liposarcomas affect both sexes equally with an average age at diagnosis around the fourth decade, but the disease can affect all age groups. According to the WHO classification of soft tissue tumors, liposarcoma are divided into well-differentiated LPS, LPS less differentiated, myxoid LPS, pleomorphic LPS and mixed type LPS. They are usually located in the anterior mediastinum (2-4).

LPS are often encapsulated tumors, however, some tumors are large, aggressive, infiltrating surrounding structures and causing compression or invasion of mediastinal organs. It may thus be responsible for dyspnea, cough, dysphagia or superior vena cava syndrome. However, LPS is characterized by a long period of clinical latency, explaining diagnosis

at an advanced stage, especially as general health condition is rarely altered. 15 % of cases is of radiologic discovery (Fortuitous). In our case, the patient, an adult over 60 years, presented with an exertion dyspnea and chest pain. CT scan showed a large invasive anterior mediastinal process (5-9). Chest radiography was nonspecific, CT scan suggest the diagnosis by showing a lesion; lipomatous in nature with a density varying between -50 and -150 Hounsfield units. The distinction between lipoma and liposarcoma is a common dilemma because of the similarities between these two lesions. Some radiological features however permits this distinction: the presence of thick septa, nodular and / or globular lesions, non-fat and decreased fat composition areas of lesions strongly suggest liposarcoma. Furthermore, chest MRI revealed: a heterogeneous tumor with a fat component (hyper signal T1 and hypo signal T2) with a tissue component (hypo signal T1, hyper signal T2). This is the gold standard of Liposarcomas. In addition to their role in the diagnosis, CT scan and MRI helps study both local and regional extensions of the tumor, its relations with surrounding structures, the search for metastases (10,11).

As with any non-surgical tumor of the mediastinum, histologic diagnosis of mediastinal LPS can be done preoperatively. Trans -parietal biopsy is difficult to interpret. Biopsy by a minimally invasive surgery to obtain a tumor fragment sufficiently representative is necessary so that histologic grading is not underestimated. The biopsy should avoid areas of necrosis. In our case, The CT scan showed an invasive lesion of the anterior mediastinum with bilateral pleural and pericardial effusion in favor of a hematologic disease, but histology on large biopsy fragments obtained by an anterior mediastinotomy confirmed for LPS. The tumor is usually large, often soft, yellow and characterized by necrotic and hemorrhagic beaches. Under optical microscopy, the LPS consists of a proliferation of spindle cells; often of a fascicular architecture, sometimes associated with epithelioid cells, round cells, and multinucleated giant cells. Hyper staining and nuclear monstrosity dominates cytoplasmic and nuclear anomalies. Mitotic activity is usually high; most LPS show extensive necrosis.

The immunohistochemical study is useful for the differential diagnosis, emphasizing the usual negativity of anti-cytokeratin anti-EMA antibodies in the sarcomatous areas (2-4). LPS should not be confused with other lesions may include

“lipoblaste-like” cells, ie fibrous mediastinitis, fibro-inflammatory post-infectious alterations, Hodgkin’s lymphomas and germ cell tumors. The differential diagnosis may also arise with a thymus-liposarcoma, the latter is distinguished by the presence of thymic tissue of varying abundance within the tumor (4). Cytogenetic studies are of value in the diagnosis of LSP, in fact, they often show polyploidy and complex chromosomal rearrangements, genomic studies show a third LSP, MDM2 amplification (8).

Surgery remains the standard method of treatment and has the advantage of being curative. However, these tumors are often large, invasive by their extensions into the spaces between muscles and along the neurovascular sheath; extra-capsular resection is rarely achievable with a surgical mortality about 15%. Surgery remains equally the treatment method in local recurrent cases. Our patient underwent oncologic surgery, extended to the left innominate vein, pericardium with wedge resection of the right ventral segment, followed by radiotherapy is ongoing (12,13).

The role of radiotherapy and adjuvant chemotherapy in the treatment of LPS remains to be determined. Randomized trials cannot be achieved because LPS are generally rare. However, meta-analysis data advises the benefit of postoperative radiotherapy and chemotherapy with doxorubicin in reducing local and regional recurrence of sarcoma. These recurrences (about 50% of cases) occur most often within 24 months after surgery, even in cases where complete resection was practiced (14).

The histological grade of the tumor is the main prognostic factor. It is associated with local recurrence, the presence of pleuro-pulmonary metastases. In all reported studies, the survival of non-operated patients is less than 18 months (6). The 5-year survival rate is 30 %. It is significantly lower than that of other thoracic sarcomas (survival rate of 40-60 % at 5 years) (1, 3, 9).

Conclusion

Liposarcomas are hardly symptomatic hence their late diagnosis. CT and magnetic resonance imaging help in establishing diagnosis. Biopsy by a minimally invasive surgery helps diagnosis, allows grading of non-surgical tumors. Surgical resection remains the only therapeutic means, even in cases of local and regional recurrences.

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