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Case Report
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A Case Report of Localized Pleural Malignant Mesothelioma: A Great Pretender Mimicking a Thymoma

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Abstract

Localized pleural Malignant Mesothelioma (LMM) is a rare neoplasm arising from mesothelial cells and histologically undistinguishable from diffuse Malignant Pleural Mesothelioma [1]. LMM is generally presented as a well-circumscribed lesion, frequently capsulated but always without gross or microscopic diffuse pleural spreading. Because of these characteristics, LMM might be clinically and radiologically confused with other pleural or mediastinal lesions, and clinical diagnosis may be therefore challenging. We report the case of a patient with an asymptomatic finding of an anterior mediastinum mass suspected for thymoma and treated with a radical excision *via* sternotomy. Histological examination showed a Localized Malignant Mesothelioma (LMM).

Introduction

Localized Malignant Mesothelioma (LMM) is a rare disease arising from the mesothelial cells which was firstly recognized as a separate entity in 2004 by WHO [2]. Despite its indolent behavior, LMM is histologically undistinguishable from diffuse malignant pleural mesothelioma [3]; it is usually characterized as a well circumscribed and capsulated lesion with neither gross nor microscopic pleural spreading. Due to these characteristics, when LMM arises from the parietal pleura, it can be clinically and radiologically difficult to be distinguished from other benign tumors of the pleura; diagnosis may be even more challenging when lesions arise from the mediastinal pleura or pericardium. Computed Tomography (CT) cannot allow a correct differential diagnosis, and FDG PET/CT, which is positive in most cases, helps to exclude a benign lesion.

Case Presentation

A 42-year-old female Caucasian patient affected by Graves' disease was referred to our department due to a mediastinal shadow in the right upper part, detected on a routine chest radiograph (Figure 1). She only complained of malaise, weakness and reported a paroxysmal hypertensive crisis. A chest CT scan, performed without iodine contrast agent because of her pathology, showed a 40 mm mediastinal mass with well-defined edges closed to the left anonymous vein and the pericardium, seemingly of thymic relevance (Figure 2). Radiological features and clinical presentation raised the doubt of a thymic tumor rather than another mediastinal lesion. Neurologic evaluation as well as the absence of acetylcholine receptor antibodies excluded concomitant Myasthenia Gravis. However, the positron emission tomography with a 2-deoxy-2-fluorine-18fluoro-D-glucose (FDG-PET/CT), revealed a significant FDG uptake of the mass with 5.5 of maximum standardized uptake value (SUV max), highly suggestive of malignancy. Surgery was then planned to remove the mass; an extended thymectomy *via* sternotomy was performed. Surprisingly, during the operation a well capsulated lesion arising from the left mediastinal pleura, adherent to the thymic tissue with no evidence of invasion of the neighboring structures was found; a complete resection of the lesion en-bloc with left mediastinal pleura, thymus and peri-thymic fatty tissue was achieved. Post-operative course was uneventful. Histopathologic examination of the specimen revealed a biphasic LMM, with fuso-cellular components within epithelioid cells organized in solid and tubular aggregates with an immunohistochemically pattern suggestive for malignant mesothelioma (Table 1 and Figure 3). After a multidisciplinary discussion, it was decided for no adjuvant therapy. To date, the patient had no signs of local recurrences or distant metastases after a follow-up of 60 months.

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Table 1: Immunohistochemical markers.

Markers	Expression
CK-pan[1], Calretinin[1], Vimentin[1]	Positive
CK7[2], CA125[2], CK5/6[2], WT-1[2]	Positive
t(X;18) (SYT;SSX) translocation	Negative
BerEP4[1], p40[1], p63[1], CD34[1], bcl-2[1], CD99[1], CEA[1], TTF-1[1], S-100[1], ER[1], Inhibin-A[1], MART-1[1], CD117[1]	Negative
CD3[3], CD5[3], CD20[3]	Positive
Ki-67	50%
Nr. of mitoses x 10 high power fields	9

[1]Markers expressed on epithelial and lymphocytic cells
[2]Markers expressed only on epithelial cells
[3]Markers expressed only on lymphocytic cells

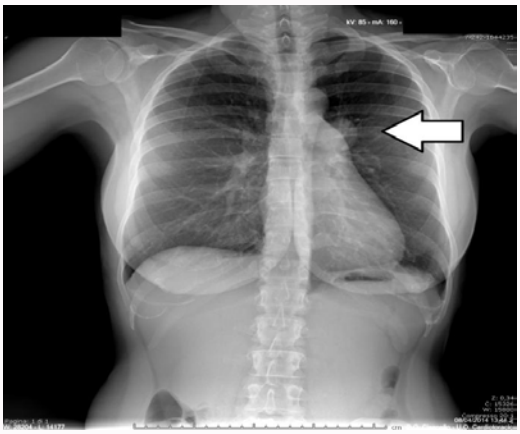

Figure 1: Chest radiograph showing a projection of the left mediastinal shadow.

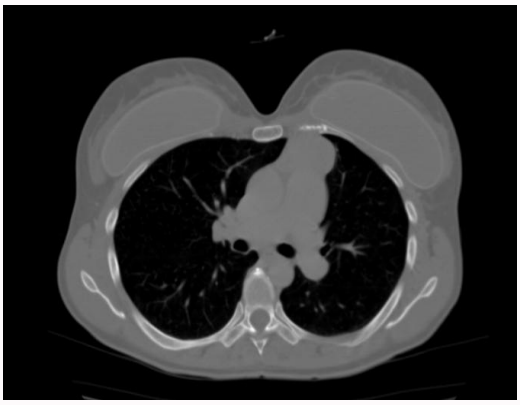

Figure 2: Computed Tomography (CT) image showing an anterior mediastinal tumor adjacent to the ascending aorta, pericardium, and left anonymous vein.

Discussion

Localized pleural malignant mesothelioma is a rare disease, firstly reported and recognized in 1994 by Crotty et al. [3] and in 2004 included in the WHO classification as a separate entity [2]. This tumor has identical histopathologic and immunohistochemically features of "classical" malignant pleural mesothelioma, but without a pleural spreading [3]. Because of the small number of cases available in the literature, the epidemiologic, clinical and pathological characteristics are not well known. Unlike the classical MPM, the role of asbestos exposure in the development of the pathology is not clear [1,3]. Considering radiological features, a correct diagnosis is often challenging; in fact, clinical and radiological characteristics of LMM are not peculiar and they might be misdiagnosed; in particular it is often confused with Solitary Fibrous Tumor of the Pleura (SFTP). Both SFTP and LMM appear on CT scan present as a homogeneous, solitary and well-circumscribed tumor arising from the pleural surface, with a dimension up to 20 cm of diameter, with no pleural effusion, even though a central necrosis or hemorrhage has been reported in lesions bigger than 10 cm. While differential diagnosis based on CT scan is usually difficult, FDG-PET can better discriminate benign SFTP from either malignant SFTP or LMM based on levels of FDG uptake. As reported by Park et al. [4], benign SFTP usually shows a SUV max lower than <2.5 while both to malignant SFTP and LMM often have higher values. In our case, differential diagnosis was based on radiological features of the mass and the clinical history of the patient, who were affected by weakness and autoimmune diseases, which could be related to thymic neoplasm [5].

LMMs of the mediastinum are extremely rare and they are more likely to arise from the mesothelial cells of the pericardium [6]. To the best of our knowledge, there is only a single case of suspected LMM of the anterior mediastinal pleura [7]; in that report, a core biopsy was performed to make a diagnosis and the patient was treated with definitive radiotherapy but no further clinical and oncological data are available. Akamoto et al. described a case of LMM in the middle mediastinum arising from the pericardium, considered in the beginning, as metastatic lymph node with no radiological evidence of the primary cancer that was surgically resected but relapsed after 1 year [6]. The largest case series of LMM available, it is that by Allen and colleagues who report clinical and pathological data of 21 patients treated with surgical intervention, but no discussion regarding diagnostic path and differential diagnosis was reported [1].

In our case a radical R0 resection has been achieved through a sternotomy; after multidisciplinary discussion, no adjuvant therapy was proposed, and the patient is currently disease-free after 4 years from surgery. Allen et al. [1] report that complete surgical resection proved to be curative in 10 out 21 of the patients from a cohort with a follow-up of 4.8 years, while in 10 out of 21 patients the LMM showed an aggressive behavior with either early local recurrence or distal metastasis, even though none of them developed a diffuse pleural disease. In a systematic review published by Gelvez-Zapata et al. [8], survival for LMM was estimated to be longer than those of classical MPM suggesting that a complete surgical excision may be curative, especially thanks to the possibility of a R0 resection. The role of chemotherapy and radiotherapy is still not clearly established and needs further evaluation [1,4,8].

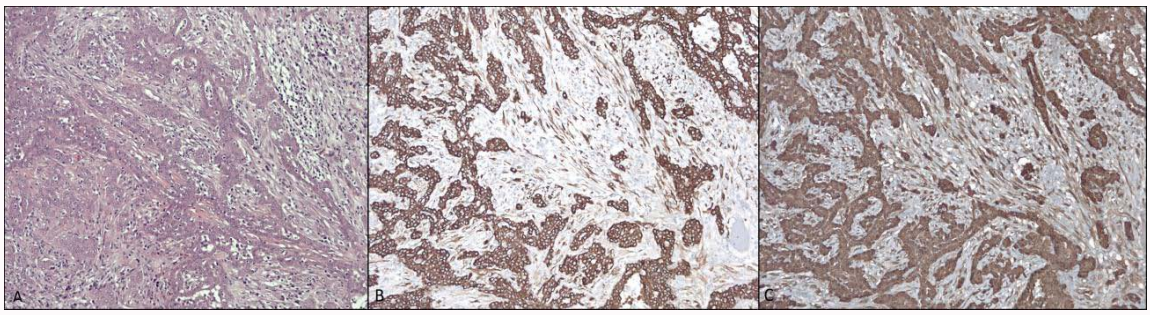

Figure 3: Photomicrograph (x10) of the surgical specimen; A: hematoxylin & eosin; B: positive reaction for calretinin; C: positive reaction for CK pan.

In conclusion, LMM is a rare tumor and differential diagnosis is not always easy. Radical surgical resection and careful follow up are usually the treatment of choice, whenever feasible.

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