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Case Report

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Thoracic pleomorphic liposarcomas: a prognostic variability depending on location

Abstract

Liposarcomas are rare tumors representing 1% of malignant tumors. These are high grade malignant mesenchymal tumors related to adipocyte tissue differentiation. Pleomorphic forms are very aggressive with a high likelihood of systemic spread and poor prognosis. Through two cases of chest pleomorphic liposarcoma, our goal is to look on the prognostic variability of the same pathological entity depending on its location.

Keywords: liposarcoma, mediastinal neoplasms, prognosis, thoracic surgery

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Abbreviations: WHO, Word health organization; CT, chest tomography; FNCLCC, fédérationnationale des centres de luttecontre le cancer (National federation of centers for the fight against cancer); AFP, alpha-fetoprotein; β-HCG, beta choriogonadotropin; MRI, magnetic resonance imaging ; PPAR-γ, peroxisome proliferator-activated receptor gamma; BCL2, B-cell lymphoma 2; VEGF, vascular endothelial growth factor; MMP2, matrix metalloproteinase-2

Introduction

Liposarcomas are rare tumors representing 1% of malignant tumors.[1] These are high grade malignant mesenchymal tumors related to adipocyte tissue differentiation.[2] According to the new WHO classification, there are 4 types: differentiated liposarcoma, pleomorphic, myxoid high grade and undifferentiated.[3] Their chest locations are rare. Pleomorphic forms are very aggressive with a high likelihood of systemic spread and poor prognosis.[4] Through two cases of chest Pleomorphic Liposarcomas, our goal is to look on the prognostic variability of the same pathological entity depending on its location.

Case report 1

This is Mrs. BK, 62years old with hypercholesterolemia on treatment who consulted in our department for a back intervertebral left shoulder swelling which has been gradually increasing in volume for 7months. Clinical examination found her in good condition with a back painless mobile mass 12cm long, without signs of skin involvement (Figure 1). This mass had both soft and hard parts with a scar from a biopsy a month ago. Thoracic CT scan (Figure 2) showed a dorsal mass with fat and solid components of 86x73x30mm extending from the posterior arches of the 4th to the 7th ribs without their invasion with an extension to the latissimus dorsi. The staging investigations based on brain, abdominal and pelvic CT scan did not show secondary location. The pathological and immunohistochemical results of the biopsy was in favor of an invasive Pleomorphic Liposarcomas grade II extending its pseudo-capsule as classified by FNCLCC. The medical file was discussed in a multidisciplinary meeting for Thoracic Oncology Group and the indication of surgical resection was made. At surgery the patient was put in a right lateral position. A circumferential peri-tumoral incision respecting a margin of 2cm was performed. A one-piece resection carrying the tumor and part of the latissimus dorsi muscle was performed with a 2cm normal tissue margin. Deep cutting of para spinal and upper margins was also performed. Skin approximation with adjacent muscles mobilization reduced the surface of the wound by 2/3 (Figure 3). Pathological findings confirmed healthy resection margins and deep tissue cuttings that were free of any tumor. A skin graft was scheduled one month later.

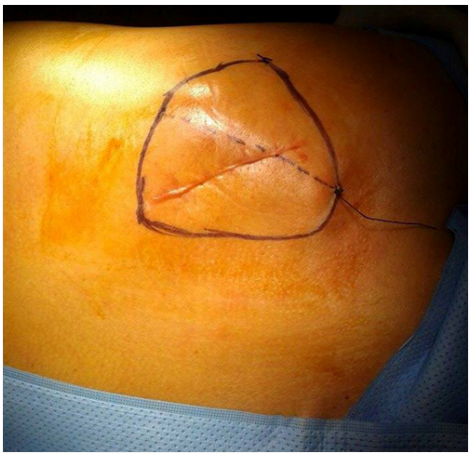

Figure 1 Back painless mobile mass 12 cm long without signs of skin involvement.

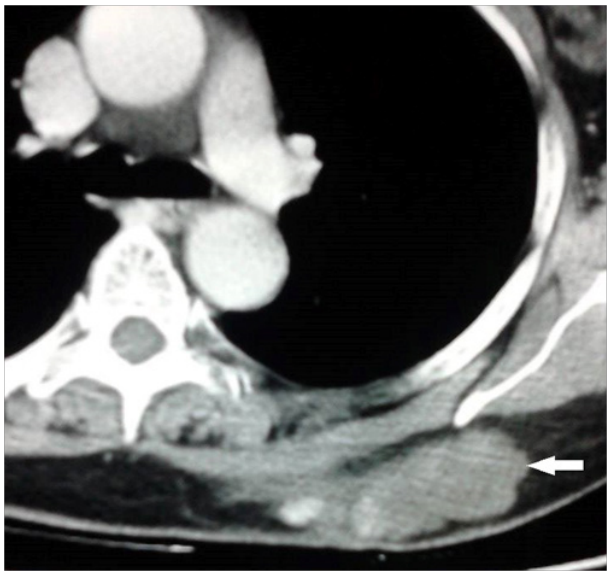

Figure 2 Dorsal mass (white arrow) with fat and solid components of 86x73x30mm with an extension to the latissimus dorsi.

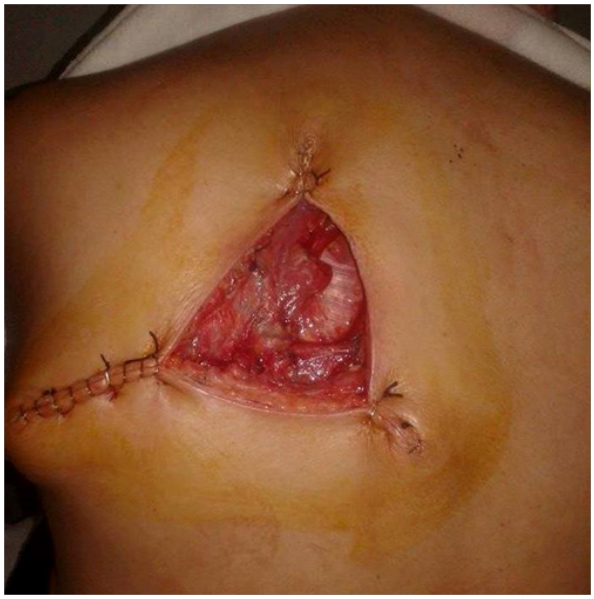

Figure 3 Skin approximation with adjacent muscles mobilization reducing the surface of the wound by 2/3.

Case report 2

Mr BS, aged 52, had as a complaint of left para-sternal chest pain of moderate intensity without radiation. The clinical examination revealed a patient in good general condition without associated comorbidity. Chest CT scan showed an anterior mediastinal mass (Figure 4) with both tissue and fatty components of 81x75x16 mm not enhanced after contrast injection invading the pericardium and heart muscle with pericardial and bilateral pleural effusion more marked on left. CT brain, abdominal and pelvis showed no distant metastasis. Echocardiography showed a normal heart compressed anteriorly by a tissue mass. Assays of AFP and β-HCG were normal. Exploration by left parasternal mediastinoscopy was decided at first. After a 3 cm incision, exploration by mediastinoscopy showed an anterior mediastinal hard mass fixed to the pericardium. Several biopsies were performed. The pathological and immune his to chemical results were in favor of pleomorphic liposarcoma grade II according to the classification of the FNCLCC. After discussion of the Thoracic Oncology Group on medical file, indication for radiotherapy was made.

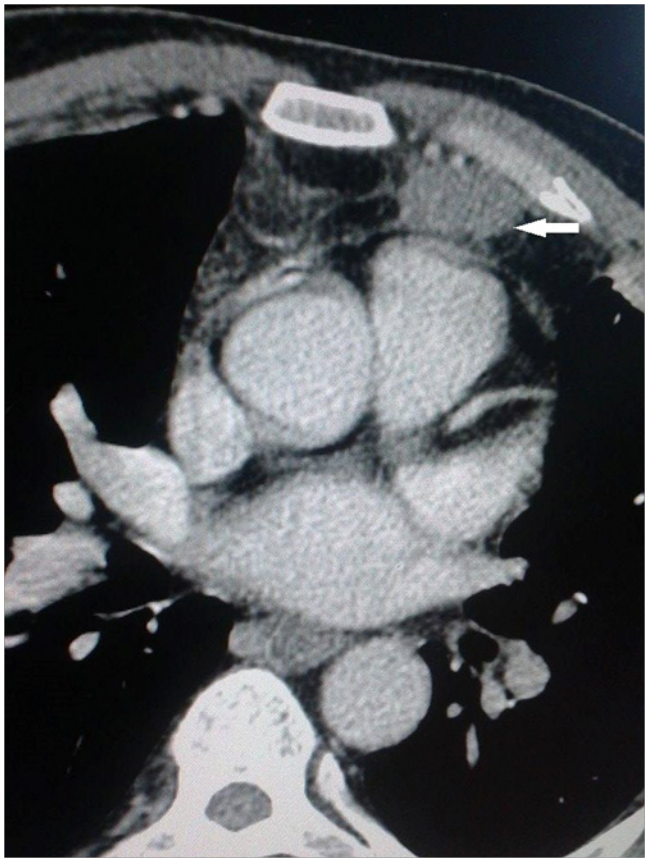

Figure 4 Anterior mediastinal mass (white arrow) with both tissue and fatty components of 81x75x16mm not enhanced after contrast injection invading the pericardium and heart muscle.

Discussion

Pleomorphic Liposarcomas is the rarest of Liposarcomas with a frequency of less than 5%.[4] There are only few studies with large series of patients with long-term monitoring. Evans in his series of 66 Liposarcomas found only 3 cases of Pleomorphic form.[5] It usually comes from the mesenchymal tissue and is localized mainly in the shoulder region, the lower limb, in the retroperitoneal and mesenteric region.[6] It grows locally and invades the neighboring structures as was the case in of our second patient where the invasion was pericardial and myocardial.[7] At mediastinal level, liposarcoma would represent only 1% of all mediastinal tumors.[8] They occur at a mean age of 45 with a slight male predominance.[1,9] Mediastinal location remains special because of the proximity to vital organs. Therefore, the clinical signs will be dominated by signs of compression on adjacent organs. This will mainly be dyspnea, cough or just isolated chest pain as the case of our patient. In case of a large compressive tumor, a superior vena cava syndrome may occur. Even so, in 12% of cases the discovery is on radiology.[1] Chest X-ray reveals a widening of

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the upper mediastinum. On CT and MRI, liposarcoma appears as a heterogeneous fat whose appearance and enhancement after contrast injection varies with different fat or tissue components.[10] In our patient, there was a mass to fat tissue component and not enhanced after contrast injection, indicating a preponderance of fat structures. Thirty-three percent of these Liposarcomas are located on the trunk but on the chest wall it represents only 15% of primary sarcoma.[11] This is usually a palpable mass rarely painful, with a soft consistency (5%) measuring an average of 10cm.[6,12] In our first patient, there was a dorsal mass measuring 86mm long. MRI remains the investigation of choice in the parietal locations. It allows better differentiation of the various components of the tumor and guides a diagnosis. It specifies the topography of the tumor and its relationship to adjacent structures especially with the medullary canal in the posterior locations.[11] The appearance is that of a heterogeneous mass with low fatty component and sometimes areas of necrosis.[1] In our patient it was not necessary. Indeed chest CT was the first exam requested and it allowed us to assess tumor respectability specifying on one hand the presence of an interface separating the tumor from adjacent structures and on the other hand it stated a partial invasion of latissimus dorsi muscle. The classification of the national federation centers of the fight against cancer (FNCLCC) has served as a reference. An overall score reflecting the aggressiveness of the tumor is obtained by the combination of various factors such as tumor differentiation, mitotic index and extent of tumor necrosis.[12] On clinical pathology, Pleomorphic liposarcoma differs from other high grade sarcomas by the presence of Pleomorphic adipocytes which is difficult to demonstrate, but whose presence is indispensable for its diagnosis.[4] Immunohistochemistry plays a small role in its diagnosis. It reveals an increased expression of the PPAR-γ (marker of adipogenesis), of BCL2, VEGF (marker of angiogenesis), metalloprotease MMP2 and other biomarkers.[4] In all cases the wide surgical resection when feasible is the best treatment for sarcomas in general and pleomorphic liposarcoma in particular. A first biopsy is offered if the tumor is not respectable at the outset; otherwise en bloc resection is performed. Adjuvant radiation therapy can be offered in case of incomplete tumor resection despite low sensitivity of liposarcoma to both radiotherapy and chemotherapy.[8] On the chest wall there is a fairly common problem of wound closure. In our patient mobilization of surrounding muscles reduced by 2/3 the area of the wound before he benefited from a further skin graft. The prognosis of these tumors is dependent of the entire surgical resection keeping in mind good margins of resection.[13] Indeed, one of the features of pleomorphic liposarcoma is the loco-regional invasion because their pseudo capsules are quickly exceeded thus being responsible for a poor prognosis. It is even worse when it is close to vital organs like in its mediastinal location. This reflects the difficulty of complete surgical resection and increased risk of recurrence. Chen in a series of 9 mediastinal liposarcoma, reported 4 deaths including 3 patients who had undergone a partial tumor resection.[14] Okurnori in his literature review of 14 cases of liposarcoma, reported 10 deaths after a median survival of 2.7 years. On the other hand, in chest parietal locations, the possibility of a radical tumor resection and better local control, offers better prognosis.[15] Tsukushi in a series of 44 operated chest wall sarcoma, including 9 liposarcoma, had an overall 5-year survival of 88.5% and a disease-free survival at 5 years at the same rate.[15] Gross meanwhile, found in a series of 55 sarcoma of the chest wall that underwent surgical resection, overall disease-free survival at 5years of 75.3% and an overall 5-year survival estimated at 87.3%. Nonetheless the risk of recurrence remains. Indeed even when resection margins appear macroscopically healthy, the risk of recurrence can be up to 10% whereas if the resection area enters tumor, the recurrence is almost inevitable with a risk of between 40 and 100%. An alternative would be the perioperative frozen section microscopic study of resection margins as in the principles of micrographic surgery of Mohs to further reduce the recurrence rate.

Thus, pleomorphic liposarcomas are rare malignancies that are very aggressive due to the invasion of adjacent structures. It emerges from these observations that the pleomorphic liposarcoma, depending of its location, has a fairly strong prognostic variability. There is particularly good prognosis for the parietal lesions unlike those of mediastinal locations. This is related to the possibility of an extensive surgical resection and the possibility of local control by adjuvant therapies. However this surgery must ensure healthy resection margins that provide a good long-term survival without recurrence.

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Conflict of interest

The author declares no conflict of interest.

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