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Case Report

Primary pleural myxoid liposarcoma: case report and literature review

Doris M. Palacios R.[1]*, Daniel B. Castillo M.[1], Jose L. Ruiz P.[2], Rafael Silva F.[3]

[1]Department of Surgery, [2]Department of Thoracic Surgery, [3]Department of Pathology, The North Central Hospital of PEMEX, Mexico City, Mexico

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***Correspondence:**
Dr. Doris M. Palacios R.,
E-mail: michpalaciosk2@gmail.com

ABSTRACT

Myxoid liposarcoma is a histological subtype of malignant tumors within the group of sarcomas. It is more common in men between the ages of 40 and 50 years. Diagnosis is difficult because they are usually asymptomatic lesions, computed tomography (CT) scan and magnetic resonance are the studies of choice. The gold of treatment is surgical resection with free margins. Chemotherapy and radiotherapy have shown a good response. A 46-year-old male was detected incidental mediastinal lesion by radiography, CT scan showed a hypodense lesion in the right hemithorax that extended to the left hemithorax, infiltrating the diaphragm and large vessels. The patient underwent an exploratory thoracotomy, finding a multilobulated tumor and mucous content approximately 600 ml, adjacent structures were infiltrated, so complete resection was not possible. Subsequently, adjuvant chemotherapy given. The histopathological diagnosis was myxoid liposarcoma. Myxoid liposarcoma is a malignant lesion. The primary pleural origin is rare. Surgical resection with free margins has a good prognosis. Due to advanced disease, a complete resection in this case was not possible, that compromised the patient prognosis.

Keywords: Pleural liposarcoma, Myxoid liposarcoma, Pleural tumor

INTRODUCTION

Liposarcoma is a malignant tumor of mesenchymal origin derived from adipose tissue. It has an incidence of 9.8 to 18% and represents the second most common histological variant of the group of sarcoma tumors (15-20%).[1,2] Myxoid liposarcoma is a histological subtype considered a low-grade tumor, usually affects adults in their fourth or fifth decades of life.

Some recent series report a peak incidence between 75 and 85 years, with a prevalence of up to 2 times more frequent in men than in women, but the clinicopathological features are not complete elucidated.[3-6] These tumors are painless, progressively growing and sometimes palpable.[3,7] A diagnosis of liposarcoma requires histological evidence of lipoblastic differentiation. Only 30 cases have been previously reported. The present case report is to expand the current knowledge of pleural myxoid liposarcoma.[6]

CASE REPORT

A 46 years old male with one year of evolution with gastric fullness, abdominal distension, oppressive chest pain, and cough. Has intermittent use of 100 mg acetylsalicylic acid for a recommendation of a private physician, and positive alcoholism, there was no significant findings in his medical history. Physical examination revealed pulmonary consolidation syndrome in the right hemithorax and a mediastinal lesion in the chest X-ray (Figure 1).

Contrast-enhanced tomography reported hypodense lesion 10-13 HU, without contrast enhancement, and poorly defined borders, localized in the posterior mediastinum surrounding the vena cava and compresses both atria, infiltrating the right hemidiaphragm and the left hemithorax, displacing lung parenchyma without involvement (Figure 2). Computed tomography (CT)-guided percutaneous biopsy tumor was performed, with an

insufficient sample for histopathological diagnosis, subsequently, a right anterior lateral thoracotomy was made, finding a 20×25 cm multilobulated tumor, that infiltrates the diaphragm, pericardium, and inferior vena cava, with mucous content, approximately 600 ml, surrounded by a pleural sac, subtotal resection was possible, the right lung expanded completely after removal of the tumor (Figure 3).

The postoperative course was uneventful. Microscopic examination showed a low-grade myxoid liposarcoma, nuclear inmunoreactivity for Ki 67 was observed in 30% of the tumor (Figure 4).

Adjuvant chemotherapy was received, actually in the fourth cycle.

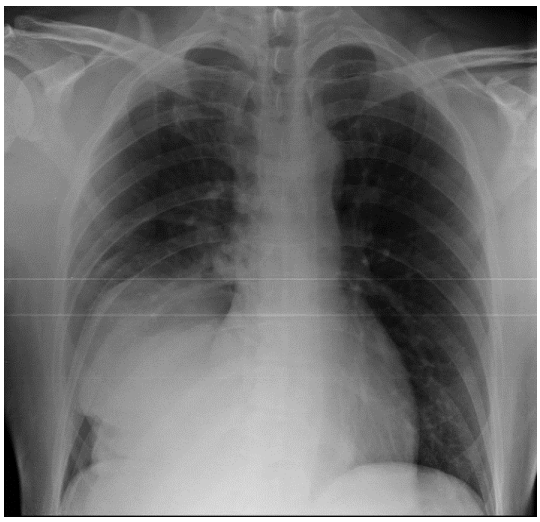

Figure 1: AP chest X-ray showing a radiopaque lesion in the right hemithorax.

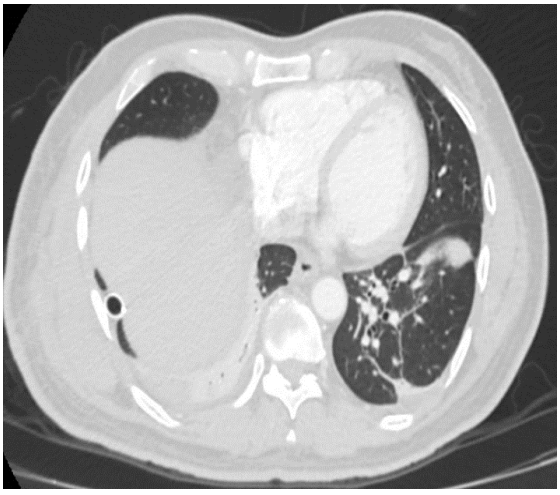

Figure 2: Axial view of contrast tórax tomography, showing a hypodense lesion 10-13 HU, without contrast enhancement, poorly defined borders, localized in the posterior mediastinum, infiltrating the right hemidiaphragm and displacing lung parenchyma without involvement.

Figure 3: Surgical piece of liposarcoma.

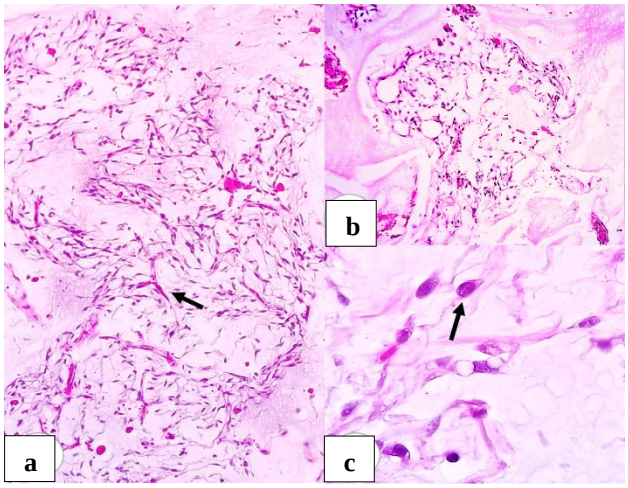

Figure 4: (a) Liposarcoma micrograph 10X. Hematoxylin-eosin (H and E) staining myxoid stroma, cells with a fusiform appearance, with small caliber vessels in the shape of a bird's foot; (b) H and E 10X microcystic pattern; and (c) H and E 40X vacuolated fusiform and stellate stromal cells (lipoblasts).

DISCUSSION

The first case of pleural liposarcoma was reported in 1942 by Ackerman and Wheeler.[17] Primary malignant pleural neoplasms are rare and constitute only 10% of pleural tumors, mesothelioma is the most frequent (90%). Primary pleural liposarcoma is extremely rare, less than 30 cases have been described in the literature, but the exact incidence is unknown.[8]

The pathological classification of liposarcomas has undergone some changes, currently, four histological variants are recognized: well-differentiated tumor (40-50%), dedifferentiated (15-20%), myxoid liposarcoma (20-30%), and pleomorphic (5-10%).[1-4]

Myxoid liposarcoma shows a specific and reciprocal chromosomal translocation that occurs in more than 95%

of cases t (12;16), (q13: p1), fusing the CHOP and FUS genes, causing deregulation in the differentiation of adipocytes and allows the proliferation of immature forms, the round cell variant is characteristic and known for its worse prognosis. [1,2,5,7,9]

It is usually located deep in the soft tissues, commonly in the extremities, popliteal region, and buttocks, in the retroperitoneal, mesenteric and pleural areas it is rare.[3,4,7,10] In the case of intrathoracic tumor it produces non-specific symptoms due to a process slow-growing that causes displacement and compression of mediastinal structures, chest pain, cough, and dyspnea are the most common symptoms.[3]

Macroscopically, they are lesions between 9-14 cm on average, well-circumscribed multinodular, with a gelatinous surface and areas of greater consistency that correspond to the areas with round cells.[3,9,11] Microscopically, it is a neoplasm of small, round, non-adipose mesenchymal cells, with variable immature forms of lipoblasts, with nuclear atypia, in a prominent myxoid stroma.[5]

Immunohistochemical studies are of little value, only the S100 protein can be useful for the diagnosis of indeterminate forms such as the round cell variety.[1] Overexpression of MDM2 and CDK4 proteins can be used to confirm a diagnosis, an expression of these markers is not seen in benign adipocyte tumors.[11]

The biological behavior of liposarcoma depends on several factors such as age, tumor size, grade, depth, and surgical margins. They are usually lesions that extend locally into the soft tissues up to 33% (contralateral limb, axilla, retroperitoneum, and bone), regardless of tumor grade. Its distant dissemination is low due to poor vascularization, ranging between 20-27% in an interval of 2.2 years from diagnosis.[1,12] dissemination occurs in 45% of cases and extrapulmonary in 55%.[2] Although it is true that the vast majority of liposarcomas originate de novo, those of primary pulmonary location it is an exception.[1]

The recurrence ranges from 57 to 78% depending on the location and accessibility for surgical resection, it usually occurs in the first 2 years after primary treatment with a mean time of 8 years. However, there is no significant relationship between the tumor size or location and clinical outcome since local and systemic recurrence can be high.[2,3,11,13]

The preoperative diagnosis of liposarcoma is often difficult, even determining the origin, the initial radiological evaluation should include magnetic resonance imaging or computed contrast tomography, which can be useful in differentiating between benign lesions and malignant tumors.[13] The characteristics of a heterogeneous mass with minimal fat (<10-25%)are suggestive of myxoid liposarcoma, it generally has a slight enhancement, a myxoid matrix is predominant, causing diffuse hypoattenuation.[8,14]

Surgery is the base of treatment for disease control.[3,7] Local resection with clean margins is the treatment of choice in localized liposarcomas, a margin of 2-5 cm is accepted, however the optimal resection in other areas such as the retroperitoneum is still controversial.[5,11,13] Occasionally, adjuvant chemotherapy with a response rate of 10-66% or external radiotherapy with a response rate of up to 59% is useful in reducing the size of the tumor and allowing a better resection margin; some results suggest a good response with combined therapy.[7,12,13] Standard chemotherapy is based on anthracyclines as first-line treatment.[15] Radiotherapy is indicated in tumors larger than 5 cm or when positive margins and a high-grade tumor are obtained.[2] Surgery with adjuvant radiotherapy has been shown to improve local control and survival rate.[16] Unresectable tumors are generally treated with palliative chemoradiotherapy.[8] Myxoid liposarcoma has better chemo and radiosensitivity than other histological subtypes.[5]

Pure myxoid lesions usually have a 5-year survival rate of 70-91%, patients with metastasis disease have a worse prognosis, the degree of malignancy depends on the predominant component, areas of gelatinous material are low grade, while round cell tumors are high-grade, with a 5-year survival prognosis of 18-21%. Necrosis and p53 overexpression are independent predictors of poor prognosis.[3,13]

CONCLUSION

Primary pleural myxoid liposarcoma is a rare entity. These patients usually have a good prognosis, but survival rates in advanced stages are highly variable. There are many factors involved in the evolution of the disease, none of which accurately predicts a good or bad prognosis. In the case presented, it was not possible to perform a complete resection of the lesion due to the locoregional infiltration of the tumor, so adjuvant chemotherapy was currently administered. That compromised the patient's prognosis, which at the moment is uncertain.

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