CASE REPORT

PLEURAL FIBROUS TUMOR: CASE REPORT

TUMOR FIBROSO PLEURAL: REPORTE DE CASOS

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RESUMEN:

El fibroma solitario de pleura poco frecuente el cual se ha estudiado para ahondar en el conocimiento de su patogenia, diagnóstico y tratamiento, logrando excelentes desenlaces para la mayoría de los pacientes afectados por esta patología. En los dos casos presentados se evidenció la presencia de marcadores inmunohistoquímicos como: CD34, STAT6, DESMINA, KI67 en el 5% y el tumor fibroso solitario fue benigno en el 78% de los casos como en el primer caso descrito en este artículo; los tumores con comportamiento maligno comprenden el 12 % de esta neoplasia, caracterizados por un comportamiento biológico indeterminado, que puede debutar con pérdida de peso, crecimiento rápido, invasión de estructuras adyacentes (pulmón, pared costal), derrame pleural, afectación ganglionar regional, recidiva local y traslado como característica, a pesar de la naturaleza distinta de este tipo de tumores, la resección quirúrgica supone el tratamiento de elección. El objetivo fue describir los hallazgos y desenlaces de 2 pacientes abordados quirúrgicamente para el manejo de TFS, para hacer de este un tema de alta relevancia en la comunidad académica y científica.

PALABRAS CLAVES: Primitivo, Crecimiento, Pleura, Extra Pleurales.

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SUMMARY:

The rare solitary fibroma of the pleura which has been studied to deepen the knowledge of its pathogenesis, diagnosis, and treatment, achieving excellent outcomes for the majority of patients affected by this pathology. In the two cases presented, the presence of immunohistochemical markers such as: CD34, STAT6, DESMINA, KI67 was evidenced in 5% and the solitary fibrous tumor was benign in 78% of the cases, as in the first case described in this article; Tumors with malignant behavior comprise 12% of this neoplasm, characterized by an indeterminate biological behavior, which can debut with weight loss, rapid growth, invasion of adjacent structures (lung, rib wall), pleural effusion, regional lymph node involvement, local recurrence and transfer as characteristic, despite the different nature of this type of tumors, surgical resection is the treatment of choice. Our objective is to describe the findings and outcomes of two patients surgically approached for the management of TFS, to make this a topic of high relevance in the academic and scientific community.

KEYWORDS: Primitive, Growth, Pleura, Extrapleural.

INTRODUCTION:

Solitary fibrous tumor (SFT) is a rare type of neoplasm that originates in the serous membranes such as: pleura, peritoneum, pericardium and mediastinum, in the pleura, this pattern tends to occur more frequently towards the outer lining around the lungs hence its name solitary fibrous tumors of the pleura [1].

The clinical manifestations are directly related to the location and size of the lesions; therefore, symptoms generally begin in advanced stages of the disease and clinical suspicion is unusual given that these tumors represent less than 5% of all pleural tumors [1]. In general, solitary fibrous tumors account for less than 2% of all soft tissue tumors [2]. It is not considered a sex-linked pathology since these tumors occur in equal

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proportions in both men and women, occurring to a greater extent between the sixth and seventh decade of life [3]. The pleura being the most frequent location of these lesions, with more than 80% arising from the visceral pleura and 20% arising from the parietal pleura [4].

The imaging diagnosis of solitary fibrous tumors can be difficult when these neoplasms are located in rare extra pleural regions, such as the mediastinum and lung parenchyma. This is important because it suggests that tumors originating at the mediastinal level usually follow a more aggressive pathologic course. In contrast, solitary intrapulmonary fibrous tumors are less common than their pleural counterparts and may be triggered from invagination of the visceral pleura and arise from interlobular septal connective tissue or progress from fibroblasts of the lung parenchyma. [4]

In this paper we will discuss the case of two female patients in their sixth decade of life who were admitted to the emergency department with respiratory symptoms, in whom the implementation of extension paraclinical plus intervention by the thoracic surgery service led to the diagnosis of a pleural fibrous tumor.

CASE REPORT

CASE # 1

A 73-year-old female patient who was admitted in November 2022 with a clinical picture of approximately 10 months of evolution characterized by chest pain, dyspnea and permanent oxygen requirement, with a history of hypertension on treatment with valsartan and bisoprolol without relevant surgical history, with no genetic information or family history. Physical examination revealed decreased vesicular murmur in the right lung, associated with signs of respiratory distress due to intercostal pulls, there was no evidence of central or peripheral cyanosis.

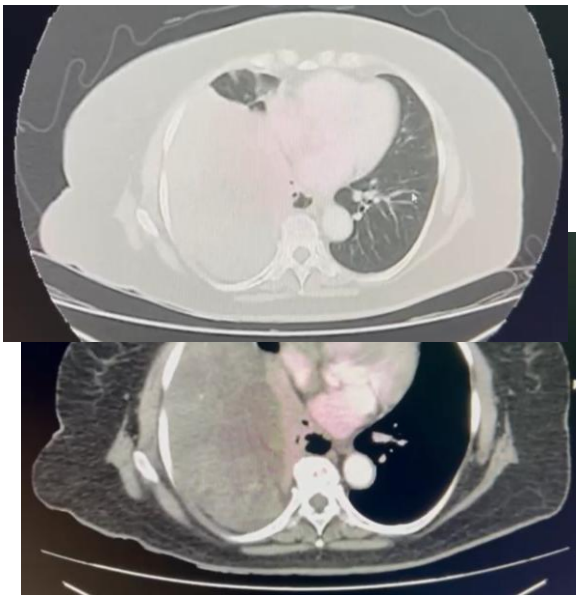
DIAGNOSIS

The institutional report of the chest X-ray indicates the presence of cardiomegaly, atelectasis in the right lung with no radiological signs of pulmonary infiltrates. **Fig. 1.** After these findings, a CT scan of the thorax is indicated showing a solid mass in the

pleural cavity involving the lower 2/3 of the right hemithorax measuring 164*170*98mm. It shows a heterogeneous density with hypervascular areas and 2 punctate nodular calcifications in its posterior aspect, causing an important compressive effect on the underlying parenchyma Fig2-3. Percutaneous biopsy is performed by the interventional radiology service in which two cylinders of tumor tissue of approximately 1.4*0.1cm are taken, whose anatomopathological analysis is described as a tumor lesion characterized by elongated cells with atypical hyperchromic nuclei, obtaining a mesenchymal fuso-cellular tumor as a diagnosis. The use of immunohistochemical markers is implemented which show strong and diffuse expression for VIMENTA, STAT-6, CD99 and CD-34 with a proliferation index measured with Ki-67 of 2%, all these results after analysis without findings compatible with a solitary fibrous tumor.

AP and lateral radiography with evidence of right hemithorax involvement.

Figure 2. Lung parenchymal window with evidence of tumor mass described in



tomography.

Figure 3. mediastinal window tumor lesion in right hemithorax.

Given the findings, the patient was evaluated by cardiology, who performed a TT echocardiogram and concluded that the patient had dilated right chambers and severe pulmonary hypertension, LVEF of 55%, a patient at high surgical risk, for which they suggested postoperative care in the ICU and did not recommend the use of antihypertensive drugs for the

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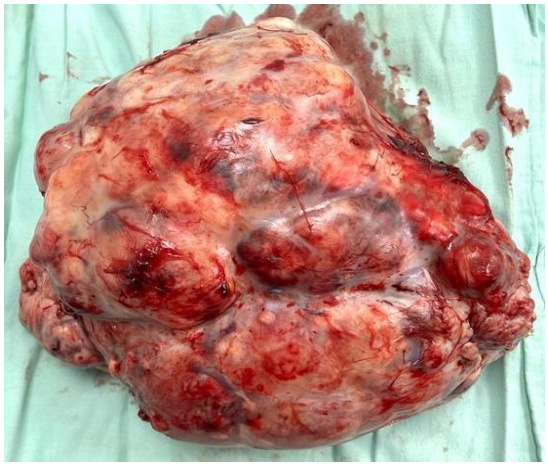
management of pulmonary hypertension.

After the findings previously described, the thoracic surgery team assessed the patient and together they indicated surgical management of the lesion with previous fibrobronchoscopy, pleurectomy and decortication by thoracoscopy plus tumor resection by thoracostomy were performed on July 14, 2022 with the following intraoperative findings: loculated right pleural effusion with presence of parietal pleural thickening, fibrinoid visceral pleura with total pulmonary entrapment secondary to large giant mass in right hemithorax which had lobulated and capsulated characteristics with presence of multiple adhesions to the pulmonary parenchyma with vascularization by bundle of diaphragmatic origin.

Image 4. Tumor surgically removed.

after removal of the tumor it was sent to pathology who described it as a neoplasm composed of elongated spindle cells of mesenchymal aspect with necrotic areas (pattern which was not evident in the first biopsy) with a diagnosis of malignant tumor of mesenchymal origin, immunohistochemistry was performed and the result was compatible with strong reactivity with cd34 membrane



pattern and nuclear pattern for STAT6, with a cell proliferation index KI67 of 4%, diagnostic indicators of solitary fibrous tumor, confirming the diagnostic impression of the first immunohistochemistry performed.

After surgery, the patient was transferred to the intermediate care unit where she stayed for 5 days and then transferred to the hospitalization service. During her stay in the intermediate care unit, postoperative follow-up was performed with control chest X-ray, which showed great improvement in lung expansion, and in which there was no evidence of tumor recurrence or pleural effusion **Fig. 5.**

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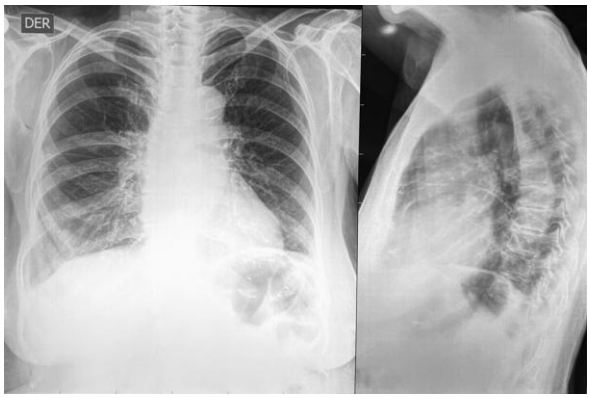

Postoperative control X-ray with lung expansion.

Additionally, during her stay in the intermediate care unit, the patient was evaluated by the sports medicine service, which indicated early mobilization and physical conditioning therapy. After 10 days of in-hospital management, there was evidence of improvement in the respiratory pattern, mobility and pain modulation. 6 months after her intervention, the patient attended postoperative control by outpatient clinic where total symptomatic recovery of the patient was evidenced since she did not present signs of respiratory distress, the scar of a not very unusual approach such as the anteromedial lateral thoracoscopy was in perfect condition, the patient has a good physical capacity and pain modulation.

CASE # 2

A 58-year-old female patient, who was admitted to the emergency department in February 2022 with clinical symptoms of 1 month of evolution characterized by dyspnea on small efforts, without pathological history, without important surgical history. No genetic information or family history was provided. Physical examination revealed decreased vesicular murmur and fine apical cramp in the right lobe. There were signs of respiratory distress and intercostal retractions.

DIAGNOSIS

On admission he underwent multiple imaging studies and pathological analysis which reported:

Chest X-ray: radio opacity in the left hemithorax and tracheal deviation to the right,

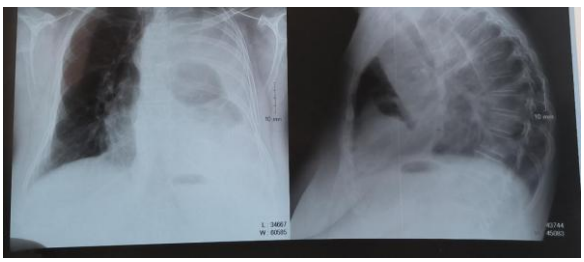

Image 6. X-ray taken on admission to the emergency room.

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As evidenced in the admission radiography and tomography images which reported mass occupying more than 50% of the left hemithorax with greater involvement of the apicoposterior segment of the upper lobe of the lung field, generating displacement of cardiomediastinum to the right, compatible with neoplastic process. The thoracic surgery team requested a biopsy by interventional radiology, which was taken and sent to studies which described the presence of cells with elongated nuclei normochromatic with slight atypia no mitosis or necrosis was observed, with this described give the diagnosis of solitary fibrous tumor and indicate the need to perform complementary immunohistochemical markers, which is identified as strong reactivity with membrane pattern for CD34 and nuclear pattern for stat 6 with a Ki67 cell proliferation index of 1%.

Complementary studies were requested such as echocardiogram which was within normal with LVEF of 60%, normal diastolic filling indicators, along with a bone scan which reports images of anterior and posterior projection normal and symmetrical biodistribution in all osteoarticular structures in which no foci of hypercaptation suggesting the presence of osteoblastic lesions were appreciated.

With all the imaging findings described the thoracic surgery team decided to take the patient to the tumor board where the decision was made to be taken to surgery for open extraction of the neoplasm, on December 15, 2022 the patient was taken to the operating room where the approach was performed starting with fibrobronchoscopy with intraoperative findings such as the presence of extrinsic compression of the left main bronchus with a decrease of more than 50% of the lumen, Subsequent interventionism is performed with the presence of left hemothorax of 1500 cc with left pleuropulmonary mass encapsulated fibrous and lobulated and hypervascularized 25x20 cms fibrous adhesions to lung parenchyma and with intercostal vascular pedicle of the 3 left bundle, parietal pleura of inflammatory appearance is evidenced.

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Image 8. Extracted left tumor sample

After removal of the lung neoplasm it was labeled and sent for pathology studies which describes a mass with a weight of 2.200 grams with measures of 22x17x14 cm with irregular nodular surface with characteristics in more than 50% of a desmoplastic pattern which is constituted by spindle cells with atypical nuclei with a mitotic count of 15 x high power field which gives the diagnosis of desmoplastic malignant mesothelioma and suggest the realization of immunohistochemical markers which reported reactivity for CD34, STAT6, DESMINA, KI67 5% and negative for CKAE1AE3, CALRETININ, CK5/6, CK7, CK20 giving as pathological diagnosis a solitary fibrous tumor with malignant characteristics, reports which are totally opposite to those reported by the biopsy taken by interventional radiology.

After leaving the surgical procedure the patient is transferred to the surgical NICU of the hospital, during the 7 days of stay in the intermediate care unit the patient is evaluated by specialties such as pulmonology which indicates to continue management with inhalers refers that does not warrant other management by that specialty, additionally during her stay is performed by radiographic control of the pulmonary status of the patient and the presence of new effusions, or recurrences.

One month after the intervention, the patient came for outpatient control where her evolution was satisfactory, her respiratory capacity was evaluated, which has improved compared to the days prior to surgery, she denied respiratory symptoms and only manifested slight pain in the surgical region, analgesic management was indicated and control with a new chest X-ray.

DISCUSSION

Solitary fibrous tumor of the pleura is a rare neoplasm, in which numerous advances have been made in the

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understanding of pathogenesis, diagnosis and treatment that allow a favorable outcome for most patients affected by this pathology. The use of diagnostic techniques, such as immunohistochemical studies, allow determining the origin in submesothelial cells, with fibroblastic and mesenchymal characteristics. [5]

The immunohistochemical analysis helps in its diagnosis, originating positivity for CD34 (hematopoietic progenitor cell antigen), for CD99 and vimentin, with negative reaction for the markers: cytokeratin, EMA, S100 protein and desmin, the latter being crucial to differentiate between whether the tumor has pleural or pulmonary involvement. [6]

The two cases previously presented evidence the presence of immunohistochemical markers such as: CD34, STAT6, DESMINA , KI67 5%, solitary fibrous tumors in 78% of cases are of benign behavior as is the first case reported in this article, but 12% represent those malignant tumors[7], which have uncertain biological behavior, often characterized by weight loss, rapid growth, invasion to adjacent structures (lung, costal wall), pleural effusion, regional lymph node involvement, local recurrence and metastasis. [8] The result of the immunohistochemistry of case number 2 classifies it as a solitary fibrous tumor with malignant characteristics, despite the fact that the imaging studies did not reflect the presence of metastatic foci or during the intervention there was no evidence of involvement of adjacent organs. Surgical resection is the treatment of choice, despite which up to 20% develop local recurrence or metastasis, with a 5-year survival rate of 68%[9,10], therefore, the follow-up of each case will be performed quarterly or semiannually in order to be attentive to its evolution.

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