Journal of
Case Reports: Open Access

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Case Report Open Access

Not so solitary! A case of a Solitary Fibrous Tumors of the Pleura

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Received Date: November 26, 2021 **Accepted Date:** December 26, 2021 **Published Date:** December 28, 2021

Citation: Jonathan Torrens Olán (2021) Not so solitary! A case of a Solitary Fibrous Tumors of the Pleura. Case Reports: Open Access 6: 1-6

Abstract

Primary neoplasms of the pleura are rare and generally divided into two major categories: diffuse and localized tumors. The most frequently encountered pleural tumor is mesothelioma, which arises from mesothelial tissue. It is more commonly associated with asbestos and has poor prognosis. Conversely, localized pleural tumors are rare and immunohistochemical studies have demonstrated that these tumors have a mesenchymal rather than a mesothelial origin. For this reason, the term "localized mesothelioma" was abandoned, and these tumors are now called solitary fibrous tumors of the pleura (SFTP). SFTP generally display benign features yet malignant potential has been described in 10-20% of cases. Only about 5% of all malignant pleural tumors are comprised of SFTP. SFTP are typically asymptomatic and discovered incidentally during chest x-ray examination showing a well-defined, lobular, solitary nodule or mass located in the periphery and typically adjacent to the pleural surface. This is important to have in the differential diagnosis, because when symptoms occur, they are usually secondary to the mass effect on adjacent structures and typically include cough, chest pain and dyspnea, which are very common respiratory symptoms for many conditions. It is also associated with paraneoplastic syndromes such as hypertrophic pulmonary osteoarthropathy and refractory hypoglycemia. The most important and valuable positive markers in SFTP are CD34, CD99, Bcl-2 and STAT-6. In most cases complete surgical resection is the only necessary treatment, and recurrence is rare.

Keywords: Pleural Tumor; Pleural Disease; Solitary Fibrous Tumor

Case Report

A 74-year-old woman with a past medical history significant for essential hypertension, former smoker, and goiter presented to the urgency room with dyspnea on exertion of one month of evolution. The patient has no known asbestos exposure. Physical examination was remarkable for decreased breath sounds on the left hemithorax. Chest Xray was initially evaluated and showed evidence of near complete opacification of the left lung. Given this finding, a chest CT scan was performed which revealed a 15.1 cm x 11 cm x 19.3 cm extrapulmonary and intrapulmonary mass in the left hemithorax with compression and subtotal collapse of the left lower lung. A PET CT scan was also performed and showed a 13 cm FDG avid mass in the left chest involving nearly all the left lower lung, and the posterior segment of the left upper lung. This is a tool which helps to categorize the tumor between malignant and benign, depending on the of its metabolic activity.

A CT guided core biopsy of the lesion was done, and pathology was consistent with a solitary pleural fibrous tumor. Immunohistochemistry was strongly positive for CD34, STAT6, and negative for pankeratin. The patient was then referred to a thoracic surgery specialist for evaluation. She was taken to the operating room and underwent a tumor embolization, to reduce blood loss and then a complete tumor resection was perfromed. The surgery was successful with an encouraging prognosis.

Discussion

Solitary fibrous tumor of the pleura is a rare neoplasm, and it accounts for almost 5% of all pleural tumors. However, it can also present as an intrapulmonary mass. Although most are benign, around 12% are found to be malignant. It usually occurs in the sixth and seventh decades of life with no sex predominance. Incidence is less than 3 per 100000 patients and less than 1000 cases have been described in the literature [1-5]. Histologically it exhibits a proliferation of uniform elongated spindle cells alternating with areas of connective tissue arranged in a haphazard distribution or 'pattern less pattern'. Immunohistochemistry plays an important role for the histopathologic diagnosis of this tumor, being the CD34, CD99, Bcl-2 and STAT-6 the most valuable positive markers [6-8] (Figures 1,2,3 and 4).

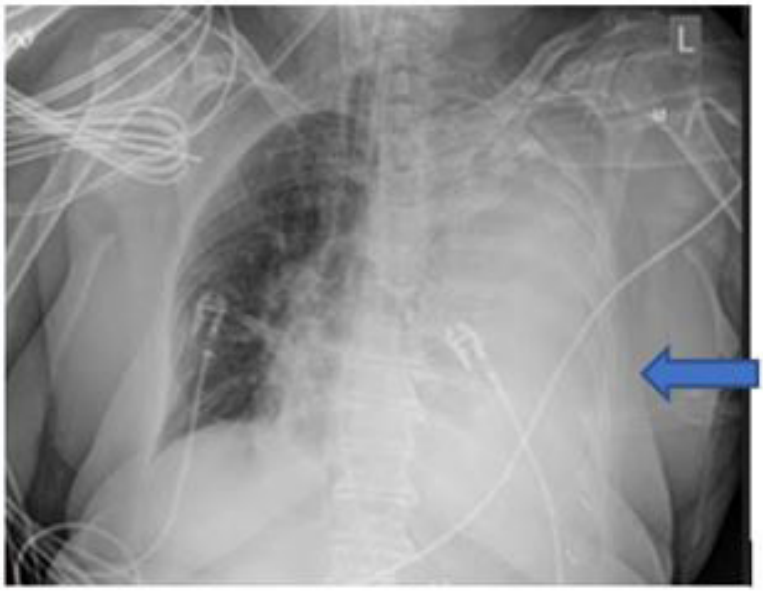

Figure 1: 1: Anteroposterior chest x ray showing near complete opacification of left lung field (blue arrow)

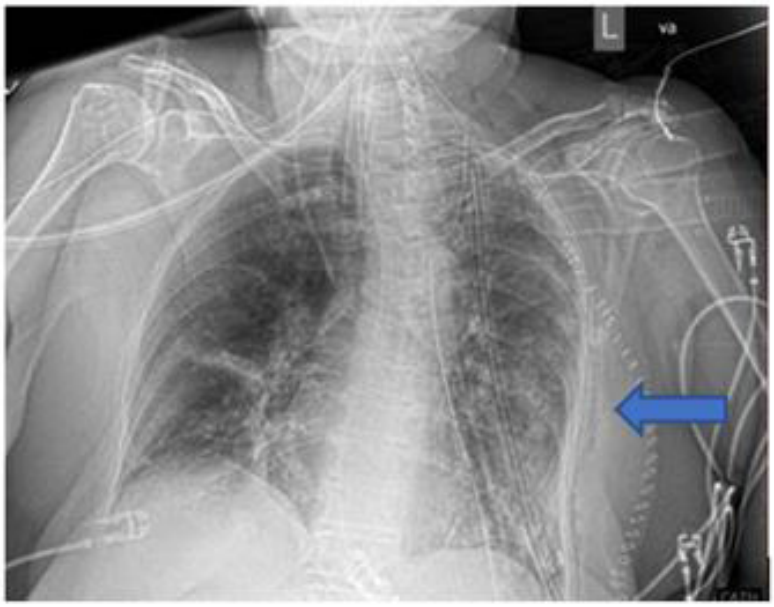

Figure 2: Anteroposterior chest x ray taken after SFTP removal. Without presence of left lung field opacification (Blue arrow)

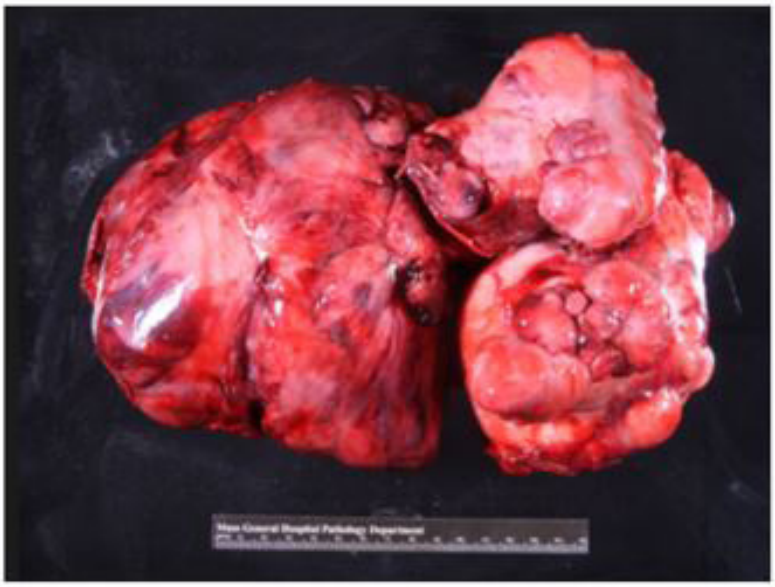

Figure 3: Gross pathological specimen showing large solitary pleural fibrous tumor

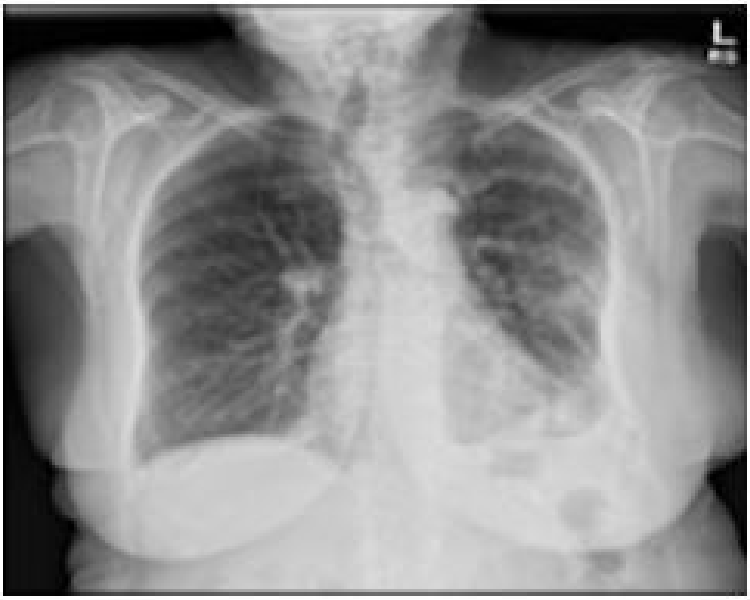

Figure 4: Posteroanterior chest x ray taken at clinic follow up 2 weeks after surgery

Conclusion

In patients with a solitary fibrous tumor of the pleura, a complete surgical resection with negative margin remains the treatment of choice in all cases, as performed in our patient. At this moment is controversial the role for conventional chemotherapy or radiotherapy as adjuvant therapy in these cases. Vascular adhesions to adjacent visceral or parietal pleura are frequent leading to bleeding, however, formal lobectomy is rarely required. Benign SFTP has a high cure rate. Around 8% of patients develop local recurrence, which is usually amenable to curative re-excision, with an overall long-term cure rate of 88% to 92%. Patient tolerated the procedure and remained stable without recurrence.

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