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Case Report

A Small And a Large Solitary Fibrous Tumors of the Pleura

Blibech H[1]; Jebali MA[1]; Snene H[1]; Belkhir D[1]; Daoud S[1]; Mehiri N[1]; Ben Salah N[1]; Ben Farhat L[2]; Ayadi A[3]; Merghli A[4]; Louzir B[1]

[1]Department of Pulmonology, Mongi Slim Hospital University of Tunis El Manar Faculty of Medicine of Tunis, Tunis Tunisia

[2]Department of Radiology, Mongi Slim Hospital, Faculty of Medicine of Tunis, University of Tunis El Manar, Tunis Tunisia

[3]Department of Pathology, Abderrahmen Mami Hospital, Faculty of Medicine of Tunis, University of Tunis El Manar, Tunis Tunisia

[4]Department of Thoracic Surgery, Abderrahmen Mami Hospital, University of Tunis El Manar Faculty of medicine of Tunis, Tunis Tunisia

***Corresponding author: Blibech H**

Department of Pulmonology, Mongi Slim Hospital, 2046, Tunis, Tunisia,

Tel: +21698686608

Email: hana.blibech@gmail.com

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Abstract

Solitary Fibrous Tumor of the Pleura (SFTP) is an exceptionally rare mesenchymal tumor that typically originates from the visceral pleura and constitutes less than 5% of all pleural tumors. While it often displays benign histological features, the tumor's behavior remains enigmatic and less comprehensively understood, primarily owing to its limited incidence and case studies.

We present two cases of SFTP. The First one was a small pleural nodule a nodule in a 61 year old patien, confirmed as a SFTP through CT-guided biopsy. Successful tumor resection was performed, with histopathological confirmation, and the patient remained recurrence-free for two years. The second one was incidental finding in a 68-year-old patient of a huge pleural mass with necrotic regions and microcalcifications on imaging. Surgical removal revealed an encapsulated tumor with myxoid features, classified as an intermediate-risk SFTP. The patient experienced no complications and had no local recurrence 16 months post-surgery.

Solitary fibrous tumors within the pleura are uncommon. The preferred treatment approach is surgical resection, with diligent post-operative monitoring.

Keywords: Solitary fibrous tumor; Pleural tumors; Thoracic surgery

Case 1

A 62-year-old male with no prior medical history presented to our hospital with chest pain. Chest X-ray revealed an apical right opacity, and a subsequent chest CT demonstrated a pleural nodule in front of the lateral segment of the middle lobe measuring 11×20 mm (Figure 1). CT-guided transthoracic biopsy confirmed SFTP, and the tumor was successfully resected via mini thoracotomy in the 5th right intercostal space. The resected mass was histopathologically confirmed to be SFTP. The patient had no local recurrence after two years.

Case 2

A 68-year-old patient was admitted to the hospital with SARS-COV2 pneumonia. Chest X-ray showed an oval opacity occupying two-thirds of the right lung field. Chest CT demonstrated a well-limited right pleural mass with patches of necrosis and some microcalcifications, adjacent to the right pulmonary artery (Figure 2). CT-guided transthoracic biopsy confirmed SFTP, and the patient underwent successful surgery resulting in complete resection of the mass. The resected mass was found to be a well-circumscribed and encapsulated mass measuring 150×120×70 mm with myxoid reorganizations in section (Figure 3). The diagnosis of SFTP was confirmed by subsequent histopathologic and immunohistochemical examinations, including CD34, CD99, Bcl2, and Ki67, which showed marked 15% of tumor cells. Pathologists concluded that this was a SFTP of intermediate risk of recurrence, based on predictive factors of aggressiveness. The patient had no postoperative complications and no local recurrence after 16 months.

Discussion

Solitary Fibrous Tumors (SFTs) are rare spindle cell neoplasms, primarily originating from the pleura, as first described by Klemperer and Rabin in 1931 (republished in 1992) [1]. Most pleural SFTs are benign, and they typically occur in middle-aged patients.

Patients may present respiratory symptoms due to tumor compression [2] or diagnosis can be discovered incidentally during imaging [3]. Chest radiographs reveal a distinct mass originating from the pleura. Chest contrast-enhanced Comput-

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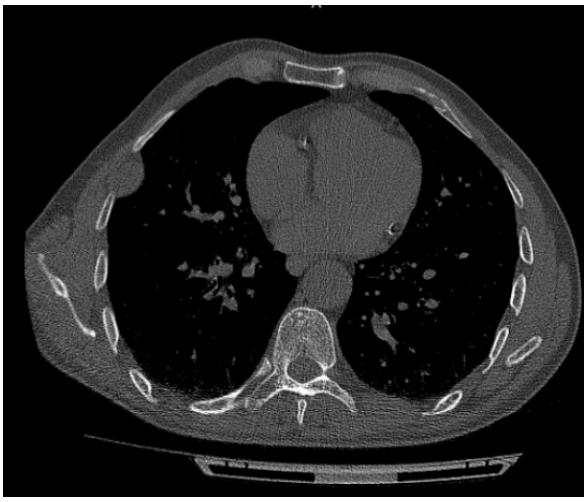

Figure 1: Chest CT without injection of contrast agent revealed a pleural nodule measuring 11×20 mm in the axial plane, located in front of the lateral segment of the middle lobe.

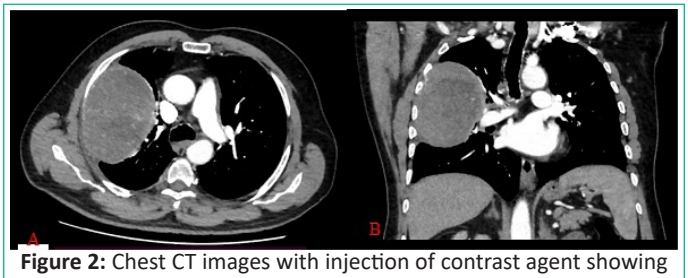

Figure 2: Chest CT images with injection of contrast agent showing a well-defined right pleural mass with patches of necrosis, measuring 130×86 mm in the axial plane and 110 mm in height. The mass was in contact with the right pulmonary artery but did not invade it. A: Axial view. B: Coronary view.

ed Tomography (CT) often shows a well-circumscribed, lobulated, hypervascular tumor with areas of necrosis, particularly in larger tumors [4]. T2-weighted Magnetic Resonance Imaging (MRI) typically displays a distinct, inhomogeneous mass with bright areas, indicating extensive necrosis [5].

The histological appearance of SFT may resemble and overlap with other benign and malignant entities, including hemangiopericytoma, leiomyoma, nodular fasciitis, inflammatory myofibroblastic tumor, fibromatosis, and benign peripheral nerve sheath tumors [4]. Tumors typically have small dimensions, limited cell density, originate from the visceral pleura, and often possess a stalk. Nevertheless, there have been documented cases of massive benign tumors that completely occupy one half of the thoracic cavity as was our second case [6,7].

SFT are typically well-defined, rubbery masses, often tethered by a pedicle and partially encapsulated. Microscopically, they exhibit a patternless proliferation of bland-looking spindly to oval epithelioid cells, forming short fascicles and clusters, mixed with thick or thin collagen bands and a prominent branching vasculature. Mature adipocytes and giant multinucleated stromal cells may also be present. Mitotic activity is generally low, with rarely more than 3 mitoses per 10 high-power fields [8,9].

Malignant counterparts typically display hypercellularity, variable cytologic atypia (occasionally moderate to marked), tumor necrosis, infiltrative margins, and a higher frequency of mitoses. Immunohistochemical staining for vimentin, Bcl2, CD 99, and CD34 is performed to distinguish SFT from other neoplasms. SFTs are usually completely negative for S-100 protein, cytokeratin AE1/AE3, and neurofilaments. In some cases, immunohistochemical negativity has been reported for c-kit, CAM

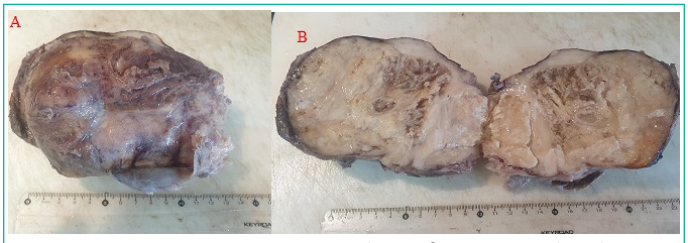

Figure 3: Macroscopic image revealing a firm encapsulated mass, measuring 150×120×70 mm (A). It presents a fasciculated and whitish appearance on section, with with myxoid reorganizations (B).

5.2, factor XIIIa, HMB-45, AE-1, SMA, CD31, and Fli-1 [9,10].

Surgical management has been the gold standard treatment for SFTs [7]. Given the variable location of the primary tumor, surgical planning and intervention is highly variable based upon location and involved structures. Obtaining adequate negative margins has been shown to decrease the rate local disease recurrence and improve survival. Video-assisted thoracoscopic surgery has clear advantages for removal of small tumors (<5 cm) that do not involve the chest wall, including shorter operative duration, less bleeding during surgery, less chest tube drainage, and faster recovery.

The use of radiation therapy in treatment of SFTs is unfortunately limited by lack of data given the rarity of this tumor. It should be considered in a case-by-case basis likely as part of a multidisciplinary plan.

The relatively low incidence of SFTs has made it difficult to identify the most effective chemotherapy for advanced SFT, Results have been variable, but most have shown low or questionable response rates [11-13]. Because of these low response rates, more research has started looking into targeted therapies. In a retrospective analysis of 11 patients, combination therapy with temozolomide and bevacizumab was shown to have a partial response in 79%, stable disease in 14%, and progressive disease in 7% [13].

Overall the prognosis for SFT is better than many adenocarcinomas with 5-year survival rates between 59–100% and 10-year survival rates between 40% to 89% [14,15]. Multiple factors have been associated with survival. Gold *et al.* [16] demonstrated recurrent tumors, macro- or microscopically positive resection margins, tumor size >10 cm, >4 mitoses/10 HPF, increased nuclear pleomorphism, increased cellularity and presence of malignant component all to be poor prognostic factors. In another large study by England *et al.* [11] they identified size >10 cm, tumor necrosis or hemorrhage, increased cellularity, pleomorphism, and >4 mitoses/10 HPF to be consistent with malignant behavior and worse prognosis. Demicco *et al.* developed and proposed a risk stratification model to classify patients into low, intermediate and high risk based on patient age, tumor size and mitotic figures/10 HPF [17].

Routine follow up is essential for SFT after surgical resection. Oncologic surveillance provides opportunity for early treatment of recurrent or metastatic disease. Given the low incidence of SFT, there are not currently agreed upon guidelines for surveillance. Many research groups have attempted to create risk models in order to guide post-operative surveillance with varying results. As stated above, the review from MD Anderson displayed that tumors greater than 15 cm in older patients (greater than 55) with mitotic figures greater than or equal to 4/10 high power fields were at high risk of metastasis deserving close surveillance [18].

Conclusion

SFTP is rare and the clinical symptoms are not obvious. Imaging, pathology, and immunohistochemistry are valuable tools for diagnosis and differential diagnosis. While surgical resection is the gold standard of treatment and provides favorable outcomes for resectable disease, optimal treatment of advanced and metastatic disease is still unclear, but currently involves a combination of surgical, radiation, and chemotherapeutic strategies under the supervision of a multidisciplinary team.

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