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Case report

Solitary fibrous tumor of the abdominal wall re-surfacing as unilateral pleural effusion and mass: A case report and review of the literature

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ABSTRACT

Background: Solitary fibrous tumors (SFTs) are rare fibroblastic mesenchymal neoplasms that were initially described in the pleura, but have been increasingly recognized to occur in other parts of the body. They have been traditionally regarded as indolent tumors that are rare to metastasize after surgical resection. Here, we describe a case of a Filipino female who initially presented with unilateral pleural effusion and mass, and was ultimately diagnosed with recurrent solitary fibrous tumor that originated from the abdominal wall. Then, we reviewed existing literature on intra- and extrathoracic SFTs with focus on pathological characteristics, recommendations for treatment as well as post-treatment surveillance.

Case presentation: A 79-year-old Filipino female with a history of solitary fibrous tumor of the abdominal wall status post complete surgical resection 3 years ago presented with unilateral pleural effusion and mass, and was diagnosed with recurrent solitary fibrous tumor that metastasized to the lung. She was not a candidate for systemic chemotherapy and ultimately died 1 year later from progressive respiratory failure.

Conclusions: Solitary fibrous tumor are rare mesenchymal tumors that were initially described in the pleura, but have now been reported in many other sites. Complete surgical resection is the mainstay therapy for all cases; however, long-term monitoring and surveillance several years after initial presentation is crucial to prevent disease recurrence, and adjuvant treatment may be necessary for patients with high-risk features. Additional studies are needed to demonstrate the clinical utility of risk stratification models and to develop post-treatment surveillance guidelines for extrathoracic SFTs.

1. Background

Solitary fibrous tumors (SFTs) are traditionally thought of as "benign" tumors with a low risk of metastasizing after surgical resection. Here, we present a case of Filipino female who initially presented with unilateral pleural effusion and mass and was ultimately diagnosed with recurrent SFT that originated from the abdominal wall. Then, we performed a literature review of intra- and extrathoracic SFTs with focus on pathological characteristics, recommendations for treatment as well as post-treatment surveillance.

2. Case presentation

A 79-year-old Filipino female with a past medical history significant for asthma and allergic rhinitis initially presented to urgent care with shortness of breath and wheezing 1 day prior to admission. She was diagnosed with asthma exacerbation based on clinical presentation and given nebulized treatments, subsequently improved and discharged home. She returned to urgent care 1 day later with worsening dyspnea and a new onset of cough productive of yellow phlegm, no hemoptysis. She otherwise denied constitutional, neurologic, gastrointestinal, or genitourinary symptoms. No sick contacts and no recent travel history.

Her other medical history was notable for chronic atrial fibrillation on Xarelto with history of left atrial appendage thrombus, GERD, and abdominal wall SFT diagnosed 3 years ago status post surgical resection with negative margins and presumed cure. The tumor was 15 × 9.5 × 11.5 cm in size, and appeared to have originated from the fascia of internal oblique muscle that was not in association with the underlying peritoneum and did not have any evidence of intra-abdominal metastasis. A preemptive total abdominal hysterectomy with

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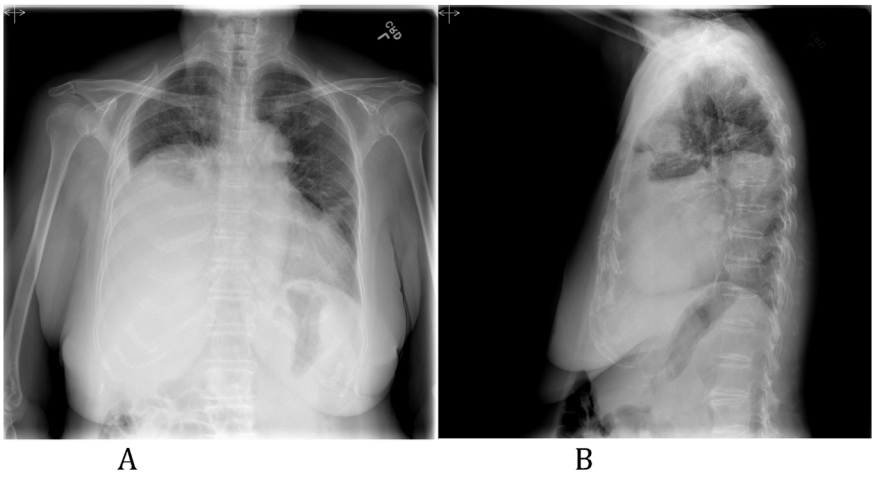

Fig. 1. Chest X-ray PA (1A) and Lateral (1B). Admit chest X-ray indicating moderate right-sided pleural effusion.

bilateral salpingo-oophorectomy was performed at the time even though the tumor was not of a gynecologic origin, with biopsy of the uterus devoid of hyperplasia, atypia or malignancy.

Family history was notable for hypertension in mother, gastric cancer in father, asthma in sister and brother. She was a life-long non-smoker, with no alcohol or illicit drug use. Her medications included Flovent 110mcg 2 puffs BID, Omnaris 50mcg 2 sprays daily, Proventil 108mcg PRN wheezing, Xarelto 20mg PO daily, Atenolol 100mg PO daily, Protonix 40mg PO daily.

Her initial vital signs were notable for pulse oximetry of 92% on room air otherwise within normal limits. She was found to have a moderate right-sided pleural effusion on chest x-ray (Fig. 1) and was subsequently admitted to the hospital. A non-contrast CT of chest was performed that revealed a large right lower lobe mass with multiple additional bilateral round lung masses and mediastinal lymphadenopathy (Fig. 2).

Transthoracic needle aspiration biopsy of one of her lung masses was planned for hospital day (HD) 2, but overnight, she had respiratory decompensation requiring ICU transfer and intubation. Bronchoscopy was unremarkable and did not show any intraluminal obstruction. On HD3, a diagnostic thoracentesis was performed however cytology from thoracentesis as well as from the bronchoalveolar lavage (BAL) was nondiagnostic. On HD5, BAL culture returned positive for pseudomonas, and she was started on Cefepime.

On HD10, CT-guided core needle biopsy of a right lung mass was performed; cytology revealed malignant cells consistent with solitary fibrous tumor with extensive necrosis and high mitotic activity (Fig. 3). The tumor was strongly and diffusely positive for CD34, bcl2, and vimentin. The histology and immunohistochemical profile were compared between the current specimen and the original specimen from 3 years ago, and was consistent with an interpretation for the current specimen as a metastatic SFT rather than a primary tumor. Hematology-oncology was consulted but thought the patient was not an appropriate candidate for cytotoxic chemotherapy given extensive tumor burden and patient's previously stated wishes regarding quality of life and goals of care. She was subsequently extubated on HD11 and transitioned to room air, and discharged to home with hospice on HD13. She had multiple re-admissions for complications related to her metastatic disease, and eventually died from progressive respiratory failure 7 months later, which was 4 years after initial diagnosis of abdominal wall SFT.

3. Discussion

Solitary fibrous tumors are rare spindle cell neoplasms that are commonly thought of as intrathoracic tumors, however up to 50–70% can occur outside of the thorax. It comprises less than 2 percent of all soft tissue tumors [1]. These tumors arise in a wide range of anatomic sites. Approximately 30% from thoracic cavity, most commonly from pleura [2,3], and 30% from peritoneal cavity, most commonly from retroperitoneum and pelvic soft tissues [4–6]. About 20% occur in head and neck [7,8], the meninges [9], or extracranial sites such as the sinonasal tract [10], oral cavity [11], deep tissue of the neck, and the orbit [12]. The remaining cases can arise from the extremities [13], the bone [14], and from the abdominal wall [15–18]. SFTs that originate from the abdominal wall is extremely rare. There are only a few case reports of SFTs that originate from the abdominal wall in the current literature. Those that provided treatment and disease outcome were

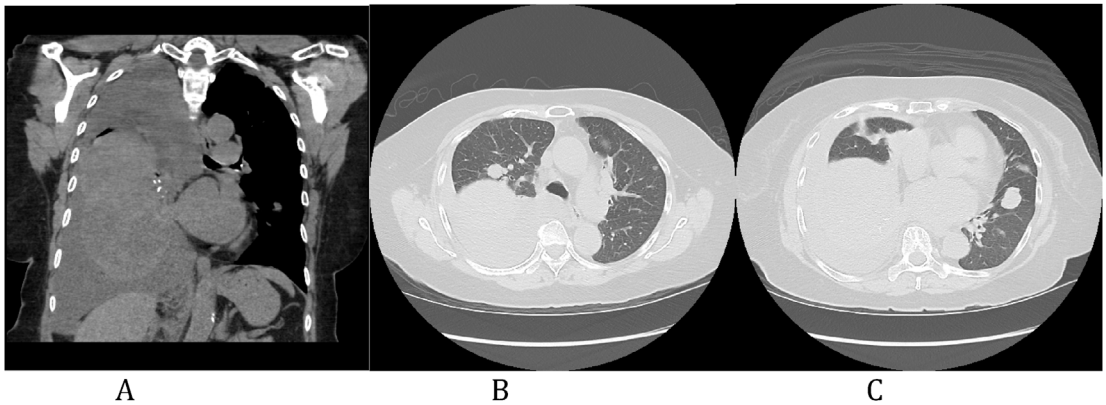

Fig. 2. CT chest notable for large poorly visualized right lower lobe mass 9.5 cm in maximal diameter (2A), with multiple additional bilateral round lung masses (2B & 2C) and mediastinal lymphadenopathy.