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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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
summarized in Table 1, all of which underwent surgical resection only with no tumor recurrence at the specified follow-up periods.

SFTs may occur at any age, but most commonly in the elderly population. Men and women are equally affected. Interestingly, a female predominance has been noted in SFTs of the abdominal wall. It has not been found to be associated with any environmental exposure or have any genetic predisposition. Refractory hypoglycemia (Doege-Potter syndrome) have been reported as a paraneoplastic syndrome in association with SFTs [19], which is caused by tumor secretion of high-molecular-weight insulin-like growth factor II (IGF2), a mediator of non-islet cell tumor-induced hypoglycemia [20].

Upon reviewing the literature, SFTs are historically considered indolent tumors that are rare to metastasize. Unfortunately, the behavior is unpredictable with a wide spectrum of biologic behavior. Most SFTs behave in a benign fashion after complete surgical resection; however, some have been reported to behave aggressively either via local recurrence or distant metastasis. Thus, long-term surveillance is mandatory. In a long-term follow-up study of 24 patients who were diagnosed with extrathoracic SFTs, 2 patients developed local recurrences at 6 months and 5 years, and 1 patient developed pulmonary metastases that were diagnosed after 7 years [21]. In addition, there is no strict correlation between morphology and clinical course. Most histologically benign SFTs prove to be non-recurring and non-metastasizing lesions. However, lesions located in the mediastinum, abdomen, pelvic or retroperitoneum tend to behave more aggressively than pleuropulmonary SFTs [22,23].

In the first large series that analyze clinicopathologic correlates of tumor behavior in 79 patients with SFTs published in 2002, Gold et al. note an overall low rate of local recurrence and metastasis after surgical treatment, with a small but statistically significant increased risk of local recurrence in extrathoracic SFTs. There was no difference in metastasis-free survival between thoracic and extrathoracic SFTs [1].

More recent studies have identified certain risk factors that may be used to predict local recurrence and distant metastases. A retrospective analysis of 81 patients treated for SFTs between 1995 and 2012 in a multicenter study showed positive resection margin was significantly correlated with local recurrence, while tumor size > 10 cm, and high mitosis rate were significantly correlated with higher incidence of metastases and showed a trend toward worse overall survival [24].

A risk assessment model for SFTs of all sites except meninges used patient age at presentation, tumor size, and mitotic index was proposed by Demicco et al., in 2012 [25] based on clinicopathological study of 110 cases that delineated patients at high risk for poor outcomes. Based on this risk assessment model, patients with SFTs in the high-risk group have a 5- and 10-year disease-specific survival rate of 60 and 0%, respectively, while those in the moderate-risk group have a survival rate of 93 and 93%, respectively [25]. However, this model has not been validated in large-scale studies and has not been validated in the clinical setting.

Above studies indicate the importance of long-term follow up for patients with SFTs, particularly those with high-risk features given the indolent nature of disease and possibility of late-recurrence. However, there exist no guidelines to follow for modality and frequency of post-treatment surveillance. De perrot et al. [26] proposed an algorithm for the treatment of pleural SFTs after complete resection based on the tumor classified either as benign or malignant, pedunculated or sessile, and recommended adjuvant treatment only for malignant sessile tumors, with CT imaging every 6 months for the first 2 years and then yearly for those with high-risk features. The classification of benign or malignant is based on a large clinicopathologic review of 223 localized fibrous tumors of the pleura by England et al. that defined malignancy as high cellularity, > 4 mitotic figures per 10 high-power fields, pleomorphism, hemorrhage, and necrosis [27].

In a paper published 10 years later, Abu Arab proposed a modified algorithm that stressed adjuvant treatment such as radiotherapy and chemotherapy also for malignant pedunculated tumors following radical resection of pleural SFTs, with CT imaging thereafter for surveillance [28]. However, it is difficult to extrapolate this approach to extrapleural SFTs, as risk assessment is more focused on pathologic characteristics rather than shape of tumor. There exist no studies that address monitoring for disease recurrence for extrapleural SFTs, and the modality and frequency of monitoring are left to the discretion of the clinician. Additional studies are needed to address the clinical utility of high-risk features to devise post-treatment surveillance for extrapleural SFTs.

In conclusion, SFTs 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.

Author contribution

XB collected the data and wrote the manuscript. JZ and CDC edited the manuscript. XB and CDC designed the study. All authors reviewed the manuscript.

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