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Case report

Recurrence of a malignant solitary fibrous tumor of the pleura 17 years after primary tumor resection – A case report

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ABSTRACT

Most recurrences of solitary fibrous tumor of the pleura (SFTP) occur within 2 years. Here we report a rare case of bilateral recurrence in a 61 year old female, 17 years after the original surgery for a right sided malignant SFTP. On repeat CT scan a 10 cm right mass and two small left lower lobe nodules were found. Patient underwent staged reoperations. She was also found to have a secondary smaller right tumor intraoperatively. All four tumors were confirmed to be recurrent SFTP on pathologic examination with identical immunohistochemistry to the original tumor.

1. Introduction

Solitary fibrous tumors of the pleura are rare entities occurring with a frequency of 2.8/100.000 individuals [7]. Although the majority of these tumors are benign and rarely recur, malignant tumors have high risk for recurrence and scrutinous surveillance is warranted [2,7]. In this case report we present a rare recurrence of a malignant SFTP, 17 years after the resection of the original tumor. First a large, 10 cm right lower lobe recurrent tumor as well as a secondary smaller tumor in the inferior pulmonary ligament were resected. Both were proved to be recurrent SFTP on pathologic examination. At the same time patient also developed enlarging left lower lobe pulmonary nodules measuring 1.0 cm and 1.3 cm respectively. These were wedge resected via robotic VATS, 2 month after the first surgery. Pathology also confirmed the same type of solitary fibrous tumor.

2. Case presentation

The patient is a 68 year old non-smoker female with history of hyperlipidemia and hypothyroidism who at the age of 51 underwent right posterolateral thoracotomy and wedge resection of a 15 cm pedunculated tumor arising from the right lower lobe of the lung. Pathology confirmed solitary fibrous tumor of the pleura. The tumor was classified as malignant due to its large size (> 10 cm) and marked cellularity, presence of more than occasional mitotic figures and foci of coagulative tumor cell necrosis on microscopic examination. The tumor was positive for CD34, negative for desmin, keratin, actin, EMA, CD68, CD31 with negative surgical margin. No cytogenetic examination was performed. She did not receive adjuvant therapy and was undergoing surveillance for 5 years postoperatively with repeated imaging without signs of recurrence. One year ago, however she developed persistent dry cough which led to a PET-CT which showed right sided pleural effusion and an associated round mass like atelectasis in the right lower lobe without significant metabolic activity. It also demonstrated two left lower lobe nodules without suspicious uptake. Subsequent CT chest was performed one year later which showed a persistent right sided pleural effusion and a 10 cm masslike ovoid lesion in the right posterior hemithorax and increased size of the previously seen left lower lobe nodules (Fig. 1). She was hospitalized and underwent drainage of the effusion, as well as, CT guided biopsy of the mass which confirmed the recurrence of the solitary fibrous tumor.

Staged resection of the large right sided tumor and subsequent resection of the contralateral lung nodules were planned. During the first surgery a redo right thoracotomy was performed. Initial thoracoscopic examination of the pleural space revealed the presence of a large pedunculated tumor originating from the right lower lobe visceral pleura with dense adhesions to the parietal pleura close where the prior thoracotomy incision was made. A smaller secondary tumor was also discovered at the level of right pulmonary ligament. 2100 mL of serous fluid was drained. The old posterolateral incision was reopened and the tumor was freed up from its parietal attachments. There was no evidence of tumor extension into the chest wall. Wedge resection was performed. The secondary pedunculated pulmonary ligament tumor measuring 1.8 cm was also removed with wedge resection (Figs. 2 and 3.).

Pathologic examination of the main tumor revealed solitary fibrous tumor of the pleura measuring 15 cm with negative margins, with strong positivity for CD34 and Stat-6 and negative immunohistochemistry for desmin, actin, CD117, S-100 with a Ki-67 proliferation index of 3–5%. 0 mitoses per 10 HPF and no necrosis were

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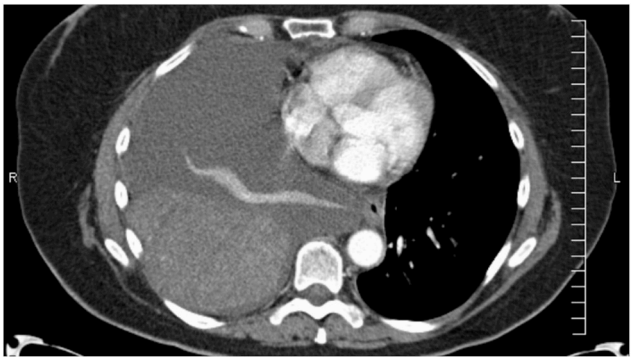

Fig. 1. Large round mass in the right hemithorax causing atelectasis and pleural effusion.

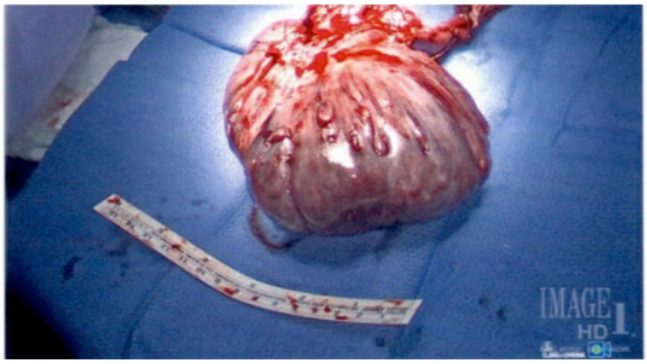

Fig. 2. Main tumor.

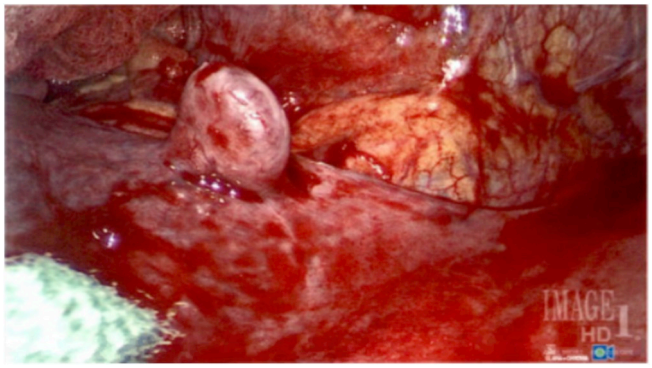

Fig. 3. Secondary tumor at the pulmonary ligament.

seen. Due to its size, however, this tumor was considered to be malignant. The secondary, smaller tumor pathology also revealed SFTP. Pleural effusion fluid cytology showed no malignant cells. Of note, the gross examination of the larger tumor revealed presence of old staple lines within corresponding to the original location of the lung wedge in 2001 and therefore likely corresponding to a local recurrence instead of a new primary. Although no genomic testing was performed the immunohistochemical examination was identical with the original tumor.

Two months after the first surgery after full recovery she was readmitted for the planned staged resection of the left sided lung nodules. She underwent a robotic assisted thoracoscopic wedge resection of a 1 cm left lower lobe superior segment and a 1.3 cm anteromedial segment lung nodules. Final pathology confirmed the diagnosis of recurrent solitary fibrous tumor with negative margins, and positive immunohistochemistry for STAT6 and CD34 and negativity for S100, identical to the right mass analysis. No increased mitotic activity was seen and the lesions measured less than 10 cm but in the light of previous tumors these were considered malignant metastases. The patient had a short, uneventful postoperative course and was discharged on POD 2.

3. Discussion

Solitary fibrous tumor of the pleura (SFTP) are rare mesenchymal soft tissue tumors of pluripotent fibroblastic or myofibroblastic origin that account for approximately 5% of all pleural neoplasms [1,2]. Several features causing them to harbor malignant potential including large size, certain immunohistochemical properties, increased cellularity, nuclear atypia and extensive necrosis. While complete surgical resection is curative in benign SFTP (bSFTP) with 5 year disease free survival over 96%, about 12% of SFTP are malignant and recur in 16–60% of the cases [2–4]. More specifically a recent French study stratified the local versus metastatic recurrence incidence (LRI vs MRI) in all SFT with the LRI being 19.2% and 38.6% at 10 and 20 years, while MRI occurring 31.4% and 49.8% at 10 and 20 years, respectively. There is no significant association between positive margins and eventual metastasis or local recurrence [9]. A comparative table of study results regarding time to recurrence, percentage of recurrence and treatment option with outcomes are summarized in Table 1 [2,4,9,13–17].

The majority of SFTP recurrences occur within 2 years of resection of a primary tumor [4]. The presence of an ipsilateral secondary local recurrence and co-existing contralateral metastases are extremely unusual. To our knowledge this is the first report in the literature with concomitant ipsi- and contralateral metastatic recurrence of a pleural SFT after 17 years [2,5,6,12]. It is unclear, however whether the right sided recurrent tumor represent a local recurrence or a metastatic recurrence on the same side as the primary tumor. Since the pathologic examination revealed identical biomarkers between the old specimen and the new one and the gross examination of the new right lower lobe mass demonstrated the tumor originating in the old right lower lobe wedge staple line, despite the lack of cytogenetic examination it is reasonable to contemplate that the second tumor represents a local recurrence. Alternatively though less likely since the pathologic examination revealed identical biomarkers, however histologic examination revealed no necrosis and 0 mitoses in contrast to the original tumor's more than occasional mitosis and coagulative necrosis, the second tumor could also represents a metastatic recurrence with a de novo tumor. The left sided nodules and small right secondary pedunculated tumor are considered metastatic recurrence based on their locations and their identical immunohistochemistry to the original and the main right sided tumors.

Factors associated with a high risk for local or metastatic recurrence are sessile morphology, resection larger than a wedge, age > 60 and high mitotic count (> 4/10 HPF), tumor localization other than limb and CD34 negativity. None of these applied to our case. The association between positive margins and eventual metastasis or local recurrence

Table 1
Comparative table of study results regarding recurrence, treatment and outcome of mSFTP.

Author	Calculated time to recurrence	Number of recurrences (% of all examined/mSFTP cases)	Site of recurrence		Treatment of mSFTP recurrences		Outcome (long term survival of all mSFTP/recurrent)
Subtype			Local	Distant	Local	Distant	
Lahon[2]	Median: 29 mo	9.5/19%	61%	39%	Resection: 90%;	XRT: 20% CHT:20% XRT + CHT: 60%	68% at 5y/-
Cardillo[13]	–	1.8/25%	–	–	Resection: 100%	–	–
De Perot[4]	Majority occurs: < 24 mo	10%/mSFTP, sessile:63% mSFTP, pedunculated: 14%					mSFTP, sessile: 70%/-mSTFP, pedunculated: 93%/-
Lococo[14]	Mean: 34 months, range: 2–128	-/30%	60%	40%	Resection: 67%	Chemotherapy:100%	81.1% at 5y/70%
Demicco[15]	Mean LR: 141 mo (15–195) Mean: MR: 30 mo (4–119)	LR:10% MR:31%	–	–	–	–	–
Harrison–Phipps[16]	Median: 34 mo	9.9%/54%	37.5%	62.5%	Resection: 100%	–	45.5% at 5y/-
Schirosi[17]	–	LR: 18.2%	–	–	–	–	5-year OS of 82.5%
[a]Salas[9]	LR: 4.3 and MR:3.6 years	LR:12.3%/LRI at 10y: 19.2% MR:16.7%/MRI at 10y:31.4%	12.3%	16.7%	–	–	OS at 10y: 76.8% OS at 20y:51.7%

mSFTP: malignant solitary fibrous tumor of the pleura; XRT: radiation; CHT: chemotherapy; LR: local recurrence, MR: metastatic recurrence; LRI: local recurrence incidence; MRI: metastatic recurrence incidence; OS: overall survival; 5y: 5 year.

[a] The study examined all site SFT, not just pleural.

are controversial [2,9–11]. Complete, repeated resection of the recurrent tumor is considered to be the gold standard therapy and adjuvant treatment is provided to improve outcome. Adjuvant therapy options include radiotherapy and chemotherapy, however, no current standard protocol or regimen exists due to the rarity of the disease. Adjuvant radiotherapy can be used to achieve local control, particularly for margin positive or recurrent tumors, however there are no randomized data suggesting its routine use for all malignant SFTP. Adjuvant chemotherapy (e.g. ifosfamide and doxorubicin) for patients with margin-positive or recurrent SFTs can be considered on a case-to-case basis and after multimodality assessment [8].

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