Turk Gogus Kalp Dama 2014;22(2):376-381
doi: 10.5606/tgkdc.dergisi.2014.8332

Pleural solitary fibrous tumors: an analysis of 11 cases

Plevranın soliter fibröz tümörleri: 11 olgunun incelemesi

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Background: In this study, we aimed to evaluate the patients operated in our clinic with the diagnosis of solitary fibrous tumor of the pleura (SFTP).

Methods: Between January 2000 and January 2011, medical data of 11 patients (6 females, 4 males; mean age 56.78 years; range 44 to 76 years) who were operated for SFTP in our clinic were retrospectively analyzed. Demographic characteristics of the patients, surgical approach applied, histology, morbidity, complementary treatment, and duration of follow-up were recorded.

Results: Two patients (18%) died during surgery. All patients underwent complete surgical excision. The surgical approach primarily consisted of thoracotomy and excision of the mass. One patient underwent left upper lobectomy, while another one underwent a wedge resection. The mass was removed with median sternotomy in one patient. The tissues of origin of the masses were the visceral pleura in two, mediastinal pleura in one, and parietal pleura in eight patients. The mean follow-up was 40.3 months (range, 30 to 89). One patient was admitted with recurrent endobronchial mass at 32 months postoperatively. The patient was treated with pneumonectomy.

Conclusion: Although SFTP has typically a benign nature, the clinical course of the disease still remains to be entirely elucidated. It should be kept in mind that these tumors have a potential of recurrence, even endobronchially, after complete surgical resection.

Keywords: Benign neoplasm; local neoplasm recurrence; pleural neoplasm; pneumonectomy; solitary fibrous tumor of the pleura.

Amaç: Bu çalışmada plevranın soliter fibröz tümörü (SFTP) tanısı ile kliniğimizde ameliyat edilen hastalar değerlendirildi.

Çalışma planı: Ocak 2000 - Ocak 2011 tarihleri arasında kliniğimizde SFTP nedeniyle ameliyat edilen 11 hastanın (6 kadın, 4 erkek; ort. yaş 56.8 yıl; dağılım 44-76 yıl) tıbbi verileri retrospektif olarak incelendi. Hastaların demografik özellikleri, uygulanan cerrahi yaklaşım, histoloji, morbidite, tamamlayıcı tedavi ve takip süreleri kaydedildi.

Bulgular: İki hasta (%18) cerrahi sırasında kaybedildi. Tüm hastalara total cerrahi eksizyon uygulandı. Cerrahi yaklaşım başlıca torakotomi ve kitlenin çıkarılması şeklinde gerçekleştirildi. Bir hastaya sol üst lobektomi, diğer bir hastaya ise kama rezeksiyon yapıldı. Bir hastada kitle median sternotomi ile çıkarıldı. Kitledeki doku orijini iki hastada viseral plevra, bir hastada mediastinal plevra, sekiz hastada ise parietal plevra idi. Ortalama takip süresi 40.3 ay (dağılım, 30-89) idi. Bir hasta ameliyat sonrası 32. ayda reküren endobronşiyal kitle ile başvurdu. Bu hasta pnömonektomi ile tedavi edildi.

Sonuç: Plevranın soliter fibröz tümörleri tipik olarak benign özellikte olmalarına karşın, hastalığın klinik seyri tam olarak aydınlatılamamıştır. Bu tümörlerin total cerrahi rezeksiyon sonrası, endobronşiyal rekürens dahil, nüks potansiyeline sahip olduğu akılda tutulmalıdır.

Anahtar sözcükler: Benign neoplazm; lokal neoplazm rekürensi; plevral neoplazm; pnömonektomi; plevranın soliter fibröz tümörü.

Available online at
www.tgkdc.dergisi.org
doi: 10.5606/tgkdc.dergisi.2014.8332
QR (Quick Response) Code

Received: February 08, 2013 *Accepted*: September 06, 2013

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A solitary fibrous tumor of the pleura (SFTP) is a very rare, slow-growing neoplasia. Approximately 800 cases have been reported in the literature,[1,2] which amounts to about 5% of all pleural tumors.[3] A pathological description was provided by Klemperer and Coleman[4] in 1931, and they proposed that the tumor originated in the mesothelial region. Recent electron microscopic and immunohistochemical studies indicate, however, that the origin may be more mesenchymal than mesothelial.[5,6] As a result, the nomenclature was changed from "localized mesothelioma" to "solitary fibrous tumor of the pleura".[7]

A majority of SFTPs with a benign histology have a pedunculated structure,[1,8,9] but approximately 12% are malignant and cause death by metastasis or local recurrence.[10] In some cases, the tumors that are benign in structure remain stable for many years before experiencing a malignant transformation.[11] These tumors are most frequently seen in patients in their sixties and seventies, with an average age of 50-57 at first diagnosis.[9,12] A possible pathogenesis related to asbestos or radiation therapy has been suggested in only two published case reports.[13,14] In addition, while 75% of symptomatic tumors are malignant, 54-67% of all patients are benign.

The most frequent symptoms are a cough, chest pain, and dyspnea in a study by Shields.[15] Furthermore, Chaugle et al.[16] found that the level of insulin-like growth factor 2 (IGF-2) was elevated in less than 5% of patients with SFTPs and the level returned to normal within three-four days after the surgery. Radical surgical excision is usually the primary treatment for SFTPs,[17] but a thoracoscopic approach can be used in cases that involve small, pedunculated lesions of the visceral pleura.[9] As for adjuvant radiotherapy or chemotherapy, these can be helpful in certain patient subsets.[11,18] Most SFTPs (88%) are generally benign; however, in 12% of the cases, advanced or extensive intrathoracic growth or unresectable recurrences may be present.[10] Moreover, recurrence and metastasis are more common in patients with malignant lesions than in those whose lesions are benign.[19]

PATIENTS AND METHODS

In this retrospective study, 11 patients (6 females, 5 males; mean age 56.8 years; range 44-76 years) who underwent surgery for SFTPs in our clinic between 2000 and 2012 were evaluated according to the following characteristics: age, gender, signs and symptoms, X-rays, tumor histology and localization, type of surgical resection performed, clinical results, recurrence, and survival. All patients underwent a preoperative examination that included routine blood tests, respiratory function tests, a standard chest X-ray, electrocardiography, and chest computed tomography (CT). In addition, fluorine 18-fluorodeoxyglucose-positron emission tomography (FDG-PET) images were available for four of the patients (36%) who had their surgery after 2007. We also recorded the perioperative and postoperative 30-day hospital mortality rates, whether in our clinic or elsewhere, and SFTP malignancy was evaluated by histological examination based on the presence of hemorrhage, necrosis, more than four mitoses for 10 fields, vascular invasion, or cellular pleomorphisms.[9]

The histological examinations revealed that the patients' tumors tested positive for CD34 and vimentin. Furthermore, an endobronchial recurrence that developed 32 months after the primary surgery in one patient (9%) was also diagnosed as an SFTP. All of the patients underwent a complete surgical resection. The mediastinal lymph nodes were not sampled, and none received neoadjuvant or adjuvant chemotherapy or radiation therapy. The long-term postoperative follow-up included a physical examination and a chest X-ray at one, three, and six months for the first postoperative year. Thereafter, these were performed on a yearly basis along with chest CT. The average follow-up duration was 40.3 months (range 30-89).

RESULTS

Only four patients (36%) had a history of tobacco smoking, and none had a history of asbestos exposure. The initial symptoms were cough in four patients (36%), dyspnea in three (27%), chest pain in two (18%), and asthenia in one (9%) while another patient was entirely asymptomatic. Furthermore, no extrathoracic symptoms, such as hyperglycemia or hypertrophic pulmonary osteoarthropathy, were seen. A bronchoscopy was performed on 10 patients (91%) without eliciting any pathological condition (Table 1), and the final pathology reports for all 11 patients indicated the presence of an SFTP.

The standard uptake values for FDG were under the cut-off value of 2.5 (range 1.0 to 2.4), and the largest tumor diameter varied from 5-16 cm. Of these tumors, there were six (54.5%) located in the left hemithorax and five (45.5%) in the right hemithorax. A left thoracotomy was performed on six (54.5%) of the patients, one underwent a median sternotomy with collar excision (9%), and the four remaining patients (36%) underwent a right thoracotomy. The mass was excised in nine patients (82%), while one (9%) had a wedge resection, and another (9%) underwent a left

Table 1. Patient characteristics (n=11)

Characteristic	Number of patients			
	n	%	Mean	Range
Age in years			56.8	44-76
Gender				
Male	5	45		
Female	6	54		
Smoking history	4	36		
Presenting symptoms	10	91		
Dyspnea	3	–		
Chest pain	2	–		
Cough	4	–		
Fatigue	1	–		
Asymptomatic patients	1	9		
Imaging				
Computed tomography only	11	–		
Computed tomography plus PET	4	–		
Preoperative procedures				
Fiberoptic bronchoscopy	10	–		
Fine-needle aspiration biopsy	7	–		
Preoperative histopathological diagnosis				
Malignant mesenchymal tumor	2	18		
Solitary fibrous tumor	2	18		
Inflammation	1	9		
Squamous metaplasia	1	9		
Hemangiopericytoma	1	9		
Postoperative histopathological diagnosis				
Benign SFPT	11	–		

PET: Positron emission tomography; SFPT: Solitary fibrous tumor of the pleura.

lower lobectomy. The tissues of origin of these SFTP masses were the visceral pleura in two cases, the mediastinal pleura in one, and the parietal pleura in the other eight. In addition, two patients (18%) were victims of operative mortality. The patient who presented with an endobronchial recurrence at 32 months underwent a left pneumonectomy and died because of respiratory failure on the ninth postoperative day. Another patient, who had undergone the removal of a giant mass in the right hemithorax, died as a result of hypotension, hypercalcemia, and cardiac arrest on postoperative day six.

Very rarely have SFTPs been reported in conjunction with endobronchial localization. In fact, to our knowledge, only one such case has been published in the English literature. Our patient with the endobronchial recurrence had a primary tumor in the left hemithoracic fissure and had undergone a resection of the mass via a left thoracotomy. A recurrence of the tumor was later developed at the left upper lobe ostium. This 66-year-old male patient had no history of either smoking or asbestos exposure, but he had undergone an appendectomy and a prostatectomy for benign prostatic hyperplasia. This patient had also received drug treatment for hypertension and type 2 diabetes mellitus (DM) over the previous 10 years. After determining the presence of the mass in the chest X-ray of this patient during his first consultation for dyspnea, chest CT was performed, which showed a well-delimited, solid mass measuring 5x7 cm localized in the left hemothoracic fissure. The mass had a benign maximum standard uptake value (SUVmax) of 2.4 on the FDG-PET (Figure 1), but no pathological condition was revealed when the preoperative fiberoptic bronchoscopy was performed. The patient had a forced expiratory volume in one second (FEV1) of 1.76 (67%) and a forced expiratory vital capacity (FVC) of 2.21 (66%).

The mass in the fissure was excised via a left thoracotomy (Figure 2). The patient was discharged on the second postoperative day due to the absence of any complications. The mass was then histologically diagnosed as a CD34 and vimentin-positive SFTP. After an uneventful follow-up period of approximately

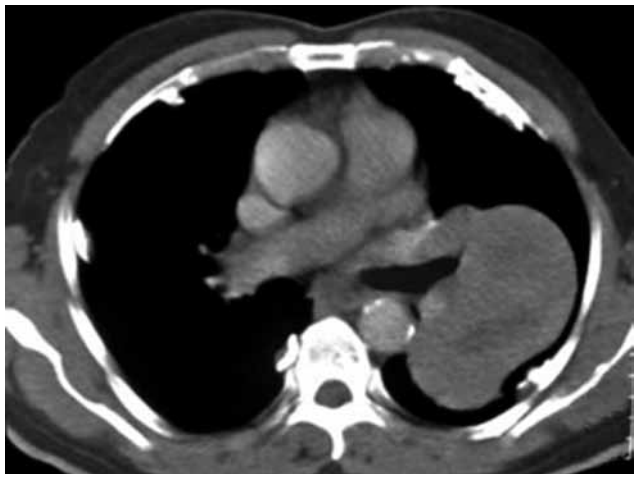

Figure 1. Mass shown on chest computed tomography before the primary operation.

two and a half years, the patient presented with a cough, hemoptysis, and a fever in the 32nd postoperative month. A vegetating mass was seen on fiberoptic bronchoscopy that was obstructing the entrance to the left upper lobe, but a histological examination was inconclusive. However, when the biopsy sample taken via the rigid bronchoscopy was histologically analyzed, a diagnosis of an SFTP was then made. After the patient's refused to undergo the proposed surgery, the upper lobe ostium was made permeable via a rigid bronchoscopy. Approximately four months later, another fiberoptic bronchoscopy performed due to the recurrence of symptoms showed a regrowth of the mass (Figure 3). An FDG-PET scan showed a mass with an SUVmax of 2.4 occupying both the upper lobe and mediastinal space, and surgery was proposed. His preoperative respiratory function tests showed an FEV_1 of 2.55 (67%) and an FVC of 3.28 (85%). Exploration identified a 3x4 cm mass in the left upper lobe and a 5x5 cm mediastinal mass; therefore, a left pneumonectomy was performed. The patient was diagnosed with pneumonia postoperatively when a chest X-ray showed infiltration on the right lung, and antibiotherapy was then initiated. Furthermore, the fiberoptic bronchoscopy was normal except for increased secretion. The patient was intubated on postoperative day seven for respiratory failure and died on day nine in the intensive care unit (ICU) due to cardiopulmonary arrest.

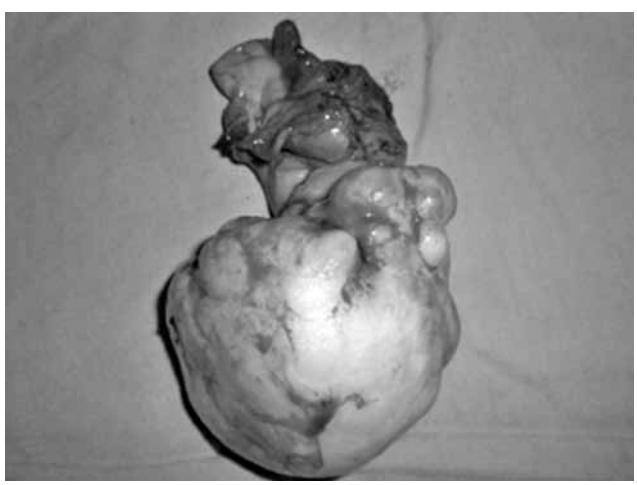

Figure 2. Pathology specimen after the excision of the mass.

DISCUSSION

Solitary fibrous tumors of the pleura are most frequently characterized as being pleural in origin. Extrathoracic locations may include the meninges, nasal and oral cavities, epiglottis, salivary glands, thyroid parenchyma, breasts, kidneys, bladder, and the spinal chord.[10,12] These tumors can remain asymptomatic for an extended period of time, with some being diagnosed by chance. The incidence rate in hospitalized patients is around 2.8 per 100,000, and these tumors make up 5% of all pleural malignancies.[20]

The frequency does not differ according to gender,[9,10,12] but some authors have noted a higher frequency in women.[8,10] In addition, the peak age for diagnosis is between 50 and 57 years old.[9,12] In our 11 patients, there were more women than men, and

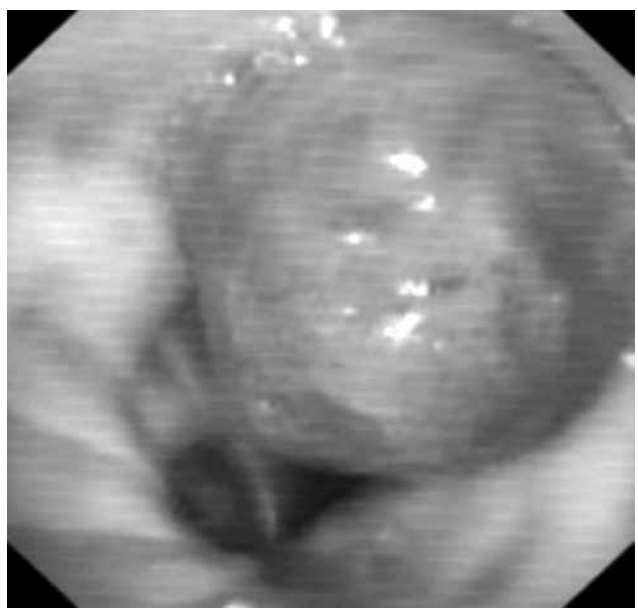

Figure 3. Bronchoscopic view of the regrowth of a recurring endobronchial mass four months after debulking by rigid bronchoscopy.

the average age was 56.8. No correlations were found related to the appearance of the tumor and genetic predisposition or exposure to asbestos, tobacco, or other environmental elements.[13] None of our patients had a history of exposure to asbestos, but four had a history of smoking.

Hemoptysis, obstructive pneumonia, and paraneoplastic syndromes have been observed in 10% of patients with clubbing and hypertrophic osteoarthropathy (Pierre Marie-Bamberger syndrome).[9] Our patient had an endobronchial recurrence at two and a half years and suffered from hemoptysis, a fever, and a cough, but none of the patients in our study had extrathoracic manifestations.

The use of FDG-PET has recently become more popular, and it has been particularly useful for following up postoperative patients. A malignant SFTP should be kept in mind when there is a high FDG uptake. All of the tumors in our patients had originally been characterized as being histologically benign, and Kohler et al.[21] reported a high FDG uptake in three cases that involved histologically malignant SFTPs. Additionally, the FDG uptake varied from 1.0-2.4 in our patients which was normal as all of our patients had benign histology.

The spindle-cell appearance of SFTPs somewhat resembles hemangiopericytoma,[22] and this had been the characterization of the preoperative fine-needle aspiration biopsy (FNAB) material in one of our cases. However, the histological examination of the resected product revealed a diagnosis of a benign SFTP. Histologically, SFPTs test positive for CD34, CD99, Bd2, and vimentin but are negative for keratin. In our study, the tumors in our patients were positive for CD34 and vimentin but negative for cytokeratin.

The diagnostic value of preoperative FNAB under CT control is relatively low.[18,23] In our series, the FNAB was performed on only seven patients, and it correctly diagnosed two of them (30%). Cardillo et al.[18] reported a success rate of 39% for this type of biopsy while Magdeleinat et al.[23] noted a rate of 45%, and Kohler et al.[21] reported a rate of 40%. Weynand et al.[24] determined that tru-cut biopsies can be used for a confident preoperative diagnosis of fibrous tumor of the pleura.

The long-term disease-free survival rate for patients with SFTPs has been reported as 90% in several large series composed of patients with benign tumors.[1,9,10] In our patients, who had average follow-up period of 40.3 months (range 30-89), only one patient had a recurrence. The natural course of an SFTP is generally benign. However, 10-30% show more aggressive development with local recurrences, malignant transformation, and distant metastasis. In our patient with the recurring tumor, a complete resection was first performed, and no satellite nodules were found at that time. The recurrence only became apparent at 32 months. Patients who have undergone up to six reoperations because of recurrences have been reported in the literature,[25] but we found only one case report with that involved an endobronchial tumor.[26]

A radical surgical excision with clean surgical margins is essentially the primary treatment for SFTPs, even in recurrent cases. We do not recommend the endobronchial approach since tumor recurrence may occur even with this preferred technique. However, after the first recurrence, our patient refused the proposed surgery, so we performed a rigid bronchoscopy to make the upper lobe ostium permeable. There is little information in the literature regarding the surgical management of endobronchial tumors, but Pak et al.[26] did report that a sleeve resection of the left mainstem bronchus with lung preservation was performed on a patient with an endobronchial SFTP.[26] In our patient, the tumor was not suitable for a sleeve resection; therefore, a pneumonectomy was performed instead. The effects of postoperative chemotherapy or radiation treatment has not been clearly established for this kind of tumor, and we did not propose any adjuvant treatment for our patients since they had all undergone a complete resection.

In conclusion, the prognosis for long-term disease-free survival after a complete surgical resection is good. Even when an SFTP is known to be benign, the clinical course of the tumor is not entirely clear. In addition, an endobronchial recurrence represents an extremely rare situation, but this should still be kept in mind in the evaluation of these patients.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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