



Solitary Fibrous Tumor of the Pleura: A Report 5 Cases

Plevral Soliter Fibröz Tümörlü 5 Olgunun Sunumu

Solitary Fibrous Tumor of the Pleura: A Report 5 Cases

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Öz

Bu çalışmanın amacı, ender görülen plevral soliter fibroz tümörlerin tedavi ve takiplerinde nelere dikkat edilmesi gerektiğini saptamaktır. 2010 ile 2015 yılları arasında, kliniğimizde plevral soliter fibröz tümör tanısı alan 5 hastanın kayıtları geriye dönük olarak incelendi. Olguların yaş ortalaması 56,2 (40-66) idi. 2 olgu semptomatikti. Diğer 3 olgu tesadüfen çekilen radyolojik tetkikler sonucunda tanı almıştı. 2 olguda malign plevral soliter fibröz tümör tanısı konuldu ve bu 2 olguda erkekti. Ortalama tümör çapı 8,6 cm (5-13) idi. Bütün hastalara komplet rezeksiyon yapıldı. Hastaların hepsi düzenli takip edildi (9-75 ay). Sadece 1 hastada rekürrens izlendi. Plevranın soliter fibroz tümörleri nadir görülen neoplazmalardır ve büyük boyutlara ulaşabilir. Komplet cerrahi rezeksiyon, bu tümörlerin en uygun tedavisidir. Cerrahi sonrası hastalar rekürrens açısından yakın takip edilmelidir.

Anahtar Kelimeler

Plevra; Rezeksiyon; Soliter Fibröz Tümör

Abstract

The aim of this study is to determine treatment and management challenges associated with the rarely seen solitary fibrous tumor of the pleura. The patient files of five cases diagnosed with a solitary fibrous tumor of the pleura in our clinic between 2010 and 2015 were retrospectively analyzed. Mean age was 56.2 (range 40-66). Two cases presented with symptoms. The other three cases were diagnosed by incidental findings in imaging studies. Two cases, both male, were diagnosed as malignant solitary fibrous tumor of the pleura. Mean tumor size was 8.6 cm (range 5-13 cm). All patients had complete resection of their lesions. All patients were followed up regularly (9-75 months). Only one patient had recurrence of disease. Solitary fibrous tumor of the pleura is a rare neoplasm that can reach enormous sizes. Complete surgical resection is the most appropriate treatment option. Patients must be closely followed up after surgery to detect recurrence.

Keywords

Plevra; Resection; Solitary Fibrous Tumor

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Introduction

Primary tumors of the pleura can be classified as localized or diffuse neoplasms. The localized form, solitary fibrous tumor of the pleura, is classified as either malignant or benign. Solitary fibrous tumor of the pleura, which had been presumed to be a variation of malignant pleural mesothelioma, was subsequently shown to be originating from the submesothelial mesenchymal layer [1]. Solitary fibrous tumors are quite rare, making up only 5% of all pleural tumors. Of these, only 10-20% are malignant [2,3]. The incidence of the tumor is reported to be 2.8/100.000 [4]. More than a third of patients have no symptoms. For patients who do have symptoms, the most frequent complaints are chest pain, cough, and shortness of breath. Diagnosis is usually made by incidental findings in imaging studies. For localized forms, optimal treatment consists of total excision of the mass and long term follow-up. For malignant forms, patients receive chemoradiotherapy after surgery [5]. In this study, we obtained the records of five patients who were operated on for, and diagnosed with, solitary fibrous tumor of the pleura, and noted clinical and radiological features and postoperative follow-up information. We reviewed these cases with regard to the current literature.

Material and Method

For the six years through 2010 and 2015, there were five patients diagnosed with solitary fibrous tumor of the pleura in our clinic. The records for these cases were retrospectively reviewed. We evaluated all cases with a history of complaints, preoperative chest x-rays and other imaging of the chest, blood chemistry, and complete blood count and pulmonary function tests. All cases had thoracotomies. None of the cases had additional diagnostic interventions. Excised surgical materials were analyzed by the Pathology Department.

Results

Our cases have a mean age of 56.2 (range 40-66). Three of them were female and two of them were male. Only two of the patients were symptomatic. For the other patients, diagnoses were made incidentally during investigation of other diseases or routine follow-up of unrelated chronic conditions (IMAGE 1). All of the patients had thoracotomy; none had additional interventions for preoperative diagnosis. Tumors were totally excised in each patient (IMAGE 2). Mean tumor size was 8.6 cm (range 5-13 cm) (IMAGE 3). Only one patient required lobectomy for total excision of the tumor due to extensive adhesions to the left lower lobe. Three of the cases had their tumors in the right lung, two of them in the left. All cases of solitary fibrous tumor of the pleura originated from the visceral pleura. Histopathologic examination showed that two of the tumors were malignant solitary fibrous tumors of the pleura. Neither of these cases had any evidence of tumor extending beyond the surgical margins, i.e. the surgical margins were clear for both these cases. It's noteworthy that both cases of malignant solitary fibrous tumor of the pleura were male patients. There were no postoperative complications and there was no operative mortality. The mean length of hospital stay was 4.8 days (range 3-6) (TABLE 1). Cases were followed up on the postoperative first, third, sixth,



Image 1. Solitary fibrous tumor of the pleura in the paravertebral space next to the right lower lobe.

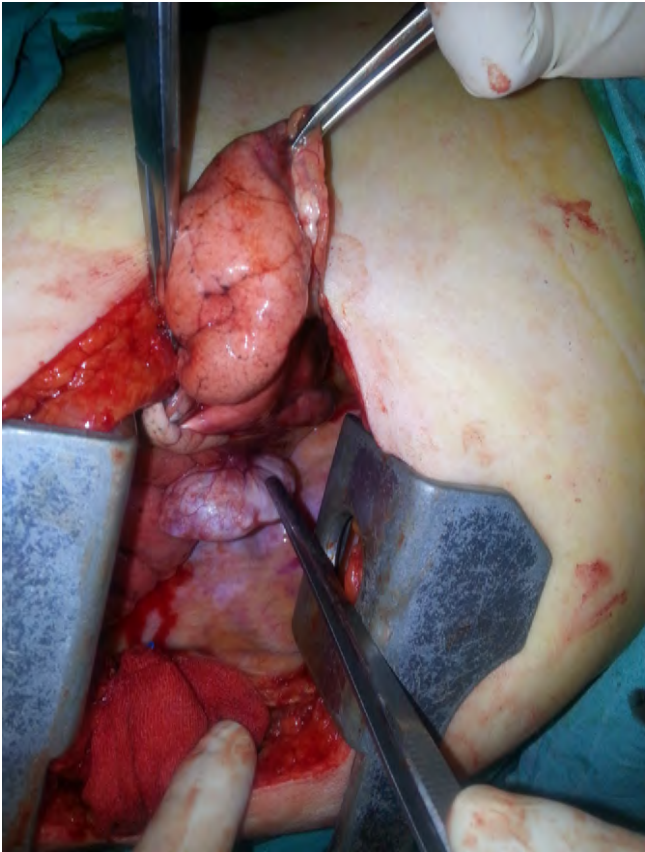


Image 2. Solitary fibrous tumor of the pleura attached to the right lower lobe with a pedicle.



Image 3. Macroscopic image of the lesion excised from right lower lobe. A well-circumscribed mass lesion, which was attached to the right lower lobe with a pedicle, was reported to be a solitary fibrous tumor of the pleura based on its histopathology.

Table 1. Demographic and clinical features of patients.

	Age	Sex	Symptoms	Tumor size (cm)	Localization of tumor	Malign/Benign	Length of stay (days)	Length of follow-up (months)
1	58	Female	None	12	Right lower lobe	Benign	6	75
2	65	Female	None	5	Left upper lobe	Benign	3	60
3	52	Male	Chest pain and dyspnea	13	Left lower lobe	Malign	6	43
4	40	Male	Chest pain	8	Right lower lobe	Malign	3	29
5	66	Female	None	5	Right lower lobe	Benign	6	9

and twelfth months, and every six months thereafter. At follow-up, the patients were checked for any new complaints and abnormal findings in the physical examination. Complete blood count and blood chemistry was analyzed and radiologic tests were performed.

Only one patient, the one who had had a left lower lobectomy, went through a rethoracotomy for local recurrence. The tumor was excised totally in the reoperation.

Comments

Solitary fibrous tumor of the pleura is an extremely rare variety of neoplasm. In 1931, Klamperer and Rabin categorized pleural tumors as either diffuse or localized and stated that the diffuse type arises from multipotent mesothelial cells and are true mesotheliomas, whereas the localized type arises from subpleural areolar tissue [6][7]. There have been approximately 900 cases reported in the English literature [8].

Although the disease can be seen at any age, incidence peaks in the fifth and sixth decades [8]. In our series, 80% of the patients were in this range. Previous studies could not find a difference in the predisposition of this disease between sexes. In our study the female to male ratio was 3:2. Since the current literature states that malign solitary fibrous tumor of the pleura has similar incidence in both sexes [10], it is coincidental that both cases with the malignant form of the disease in our series were male patients.

More than a third of cases are asymptomatic. Symptomatic cases might have cough, shortness of breath, chest pain, hemoptysis, fever, chills, night sweats, fatigue, loss of appetite, sensitivity in the chest wall, pleural effusion, superior vena cava syndrome, and changes in electrocardiography [4] [12-14]. The rest of the patients were diagnosed incidentally during investigation of other diseases or during routine follow-up of unrelated chronic conditions.

Only 10-20% of solitary fibrous tumors of the pleura are malignant. In a multicenter retrospective case series of 50 patients, the recurrence rate of malignant solitary fibrous tumors of the pleura was 15% [5]. The aforementioned study recommended performing a chest wall resection, lobectomy, and even pneumonectomy if required for total excision of the tumor. For patients who have local recurrence, the recommended treatment is total excision of the recurrent tumor, if resectable. Patients with unresectable tumors should receive chemoradiotherapy. Resection with video-assisted thoracoscopic surgery is possible in selected cases [9][15]. Whatever the choice of surgical technique, for optimal treatment total excision of the tumor is vital. It is the most important determinant of long term survival. To be specific, after total excision of benign solitary fibrous tumors of the pleura, the 5-year survival rate can be as high as 100%.

The 5-year survival rate for patients with malignant solitary fibrous tumors is around 81% [5]. If total resection is not possible, another treatment modality is chemoradiotherapy. Survival rates for these patients are lower.

Recurrence of solitary fibrous tumors of the pleura is more common in the malignant form and is reported to be 15% [5]. Recurrence in the benign form is rare, but not unheard of. Local recurrence even after 15 years has been reported. Survival in cases with recurrence is most influenced by whether or not the recurrent tumor can be totally excised. For this reason, it's imperative that patients diagnosed with solitary fibrous tumor of the pleura are followed up at regular intervals. Radiotherapy is a treatment option for recurrence of malignant solitary fibrous tumors of the pleura. It has been reported that malignant solitary fibrous tumor of the pleura responds very well to radiotherapy [16].

In conclusion, the most important treatment element for both benign and malignant forms of solitary fibrous tumors of the pleura is total surgical excision. One should not refrain from aggressive surgical procedures for total excision of the tumor, particularly in the malignant form. Patients should be followed up at regular intervals postoperatively. If there is recurrence, eligible patients should have a repeat surgery. Outcomes for solitary fibrous tumors of the pleura are favorable with appropriate surgery and follow-up.

Competing interests

The authors declare that they have no competing interests.

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