Int J Clin Exp Pathol 2020;13(9):2363-2368
www.ijcep.com /ISSN:1936-2625/IJCEP0115406

Case Report

Rapidly growing giant solitary fibrous tumor of the pleura: a case report and review of the literature

Yuanyuan Wang[1], Ying Xu[2], Ruiling Li[3], Ye Yuan[1]

[1]Department of Anesthesiology, First Hospital of Jilin University, People's Republic of China; [2]Department of Colorectal and Anal Surgery, First Hospital of Jilin University, People's Republic of China; [3]Department of Cardiology, Second Hospital of Tianjin Medical University, People's Republic of China

Received June 1, 2020; Accepted July 20, 2020; Epub September 1, 2020; Published September 15, 2020

Abstract: Solitary fibrous tumors of the pleura (SFTPs) are rare slowly-growing tumors that originate from mesenchymal cells of the submesothelial tissue of the pleura. A 57-year-old female presented to our institute, and complained of mild chest discomfort for one year. The computed tomography (CT) showed a thoracic mass measuring 3.4×1.5 cm. She refused surgical removal, and turned to the medical treatment with oral herbs with unknown specific ingredients; however, she had persistence of her symptoms and gradually developed general fatigue and severe dyspnea six months before presentation. The CT scan revealed a giant mass measuring 33×20×15 cm in the right hemithorax, leading to total atelectasis of the lung. After preoperative embolization of the tumor-supplying arteries, the tumor was completely removed. The diagnosis of SFTP was confirmed by subsequent histopathologic and immunohistochemical examinations. The postoperative course was uneventful with a 15-day length of stay, and the patient was in good health at the 15-month follow-up.

Keywords: Neoplasms, pleural effusion, solitary fibroma of the pleura

Introduction

Solitary fibrous tumors of the pleura (SFTPs) are rare neoplasms of mesenchymal origin [1], and have a benign biologic behavior in the majority of cases. SFTPs affect male and female individuals equally, and the highest incidence occurs between 60 and 70 years of age [2]. Most SFTPs grow slowly, so the majority of patients are asymptomatic, and clinical findings such as fever, weight loss, and fatigue are not characteristic [3]. Contrast-enhanced computed tomography (CT) and 3D technology aid in making the diagnosis of tumor and assessing the possibility of surgery, and the definite diagnosis requires histopathologic and immunohistochemical analysis.

Case report

A 57-year-old female complaining of general fatigue and severe dyspnea presented to our institute in March 2019. The patient reported a thoracic mass measuring 3.4×1.5 cm on her CT image with mild chest discomfort for one year. She refused surgical removal, but turned to medical treatment with oral herbs with unknown specific ingredients. Nevertheless, her symptoms continued to deteriorate; and she gradually experienced with dyspnoea, orthopnoea, and paroxysmal nocturnal dyspnoea during the following six months. The patient had a loss of body weight of 8 kg and the contrast-enhanced CT showed that the large heterogeneous tumor had increased significantly in size measuring 33×20×15 cm and completely filled the right hemithorax, leading to almost total atelectasis of the lung. There was obvious compression of the surrounding blood vessels and mediastinal and tracheal shift (**Figure 1A**, **1B**). She had no other chest complaints, such as cough, hemoptysis, or chest pain. Her past medical history and family history were unremarkable except that she underwent conservative medical management for the pleural effusion 26 years ago. She was a nonsmoker, and denied exposure to asbestos, radon gas, and other occupational exposures. Positive findings on physical examination included tracheal shift, enhanced jugu-

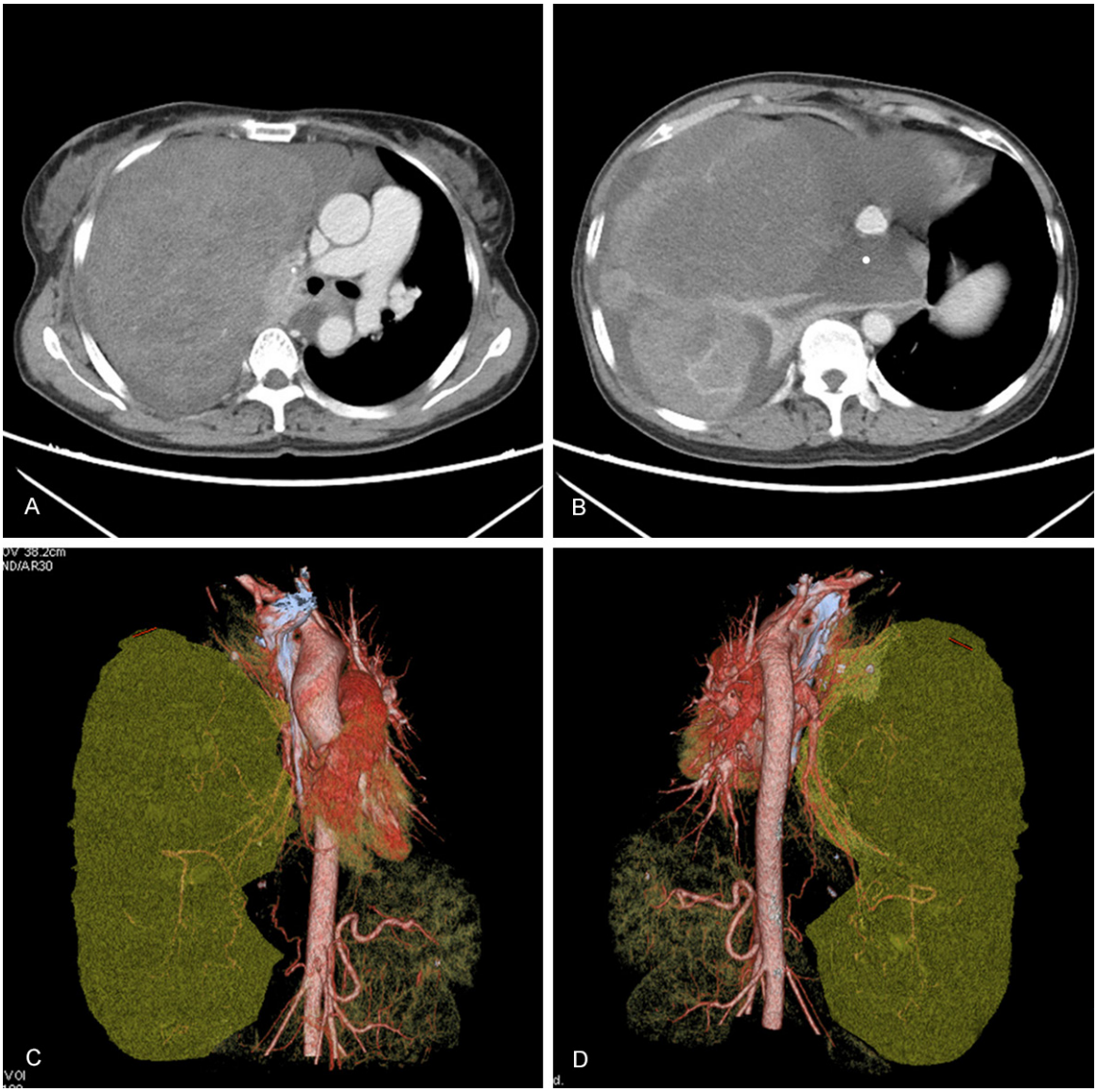

Figure 1. A, B. A contrast-enhanced CT scan demonstrated a giant mass in the right hemithorax and the shift of blood vessels, mediastinum, and trachea. C, D. 3D reconstruction of contrast-enhanced CT improved the visualization of the mass and the relationship with the surrounding tissues and organs.

lar pulsations, and significantly decreased right breath sounds. Electrocardiogram showed sinus tachycardia, and laboratory results were almost within normal limits. The patient had a CT guided transthoracic biopsy of the mass, and the histopathologic examination revealed the benign features of a solitary fibrous tumor. The tumor and its adjacent tissues and organs, especially the major blood vessels, were reconstructed by 3D technology to conduce to preoperative consultation (**Figure 1C**, **1D**).

A complete surgical resection was planned after comprehensive consideration with multiple disciplines. The preoperative embolization of the tumor-supplying arteries was performed in order to reduce the risk of massive hemorrhage. The tumor was too large to be removed with the usual posterolateral incision. Hence, the combination of anterior approach with median sternotomy and right posterolateral thoracotomy through the fifth intercostals was performed after general anesthesia with a left double-lumen tube. After careful adhesiolysis, the main vascular pedicles of the tumor were ligated with nonabsorbable suture, and the mass was completely removed. After resection of the tumor, the patient's blood pressure

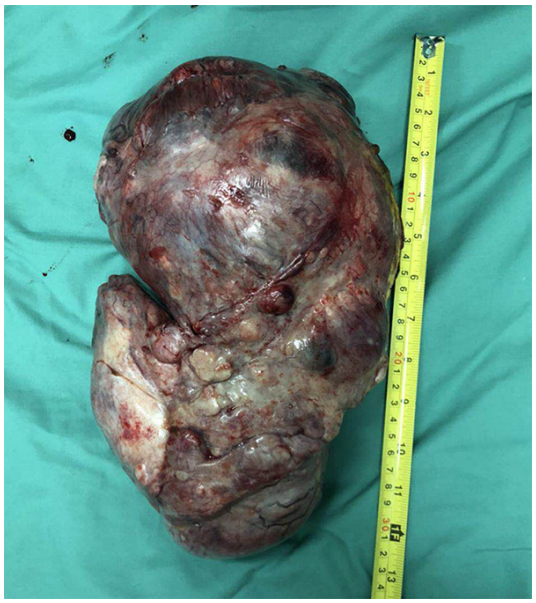

Figure 2. The surgical specimen measuring 33×20×15 cm in size was encapsulated with extensive vessels.

dropped immediately, and large doses of norepinephrine and fluid challenge were applied until the patient's hemodynamics were stable. The operation took 160 min and blood loss was 1000 ml. The patient with endotracheal tube was transferred to the intensive care unit, and she was successfully extubated 2 days later. Macroscopically, the well-circumscribed, encapsulated resected tumor was 33×20×15 cm with extensive vessels, and weighed 4500 g in the fresh state (**Figure 2**). Histopathologic examination showed spindle-shaped tumor cells mixed with bands of collagen, and was consistent with benign SFTP (**Figure 3**). Immunohistochemical stains demonstrated that tumor cells were strongly positive for STAT6, Vimentin, and Ki-67, negative for PCK (**Figure 4**). The final diagnosis was confirmed as a SFTP without malignant features. Afterward, the patient made an uneventful recovery, and was discharged on postoperative day 15. Her performance status improved from 3 to 1. At the 15-month follow-up, the patient remained in good health, and there was no evidence of recurrence or metastasis.

Discussion

SFTPs, first described by Klemperer and Rabin in 1931, are rare benign or low-grade malignant tumors, and account for < 5% of all pleural tumors. They derive from the CD34-positive dendritic mesenchymal cells layer and usually arise from visceral pleura [4]. Malignant SFTPs represent 10-20% of all SFTPs, with a 5-year survival of 81% [5]. The average tumor was 2.13 kg, while the present case reported that the giant SFTP with a maximal diameter of 33 cm occupied the entire right hemithorax and weighed 4500 g. SFTPs affect individuals of all ages, and the highest incidence occurs in the sixth or seventh decade of life. There is no relationship to asbestos, radon gas, and other occupational exposures [6, 7]. Most SFTPs grow slowly, so the majority of patients are often asymptomatic, and clinical findings such as fever, weight loss, and fatigue are not characteristic [3]. The tumor in this case grew rapidly in the short term, however, which might be related to taking the unknown herbs. SFTPs are usually diagnosed incidentally on physical or other examinations or in the later stages of the development, when compressing the adjacent trachea and lung tissue and causing chest tightness, dyspnea, and even respiratory insufficiency [8-10]. The compression of the lung tissue can result in total atelectasis of the lung. 10% of patients with SFTPs present with paraneoplastic syndromes, including hypertrophic osteoarthropathy, hypoglycemia, and clubbed nail-beds, and these symptoms always resolve after resection of the tumor [11, 12]. This patient did not demonstrate these signs and symptoms.

The thoracic CT scan is a very useful diagnostic method. It can identify the location and size of the tumors clearly and help surgeons to assess the possibility of surgery. In addition, malignancy is suggested by the presence of a maximum diameter >10 cm, calcification, inhomogeneous density, rich blood supply, obscure boundaries, and pleural effusion on CT image [13, 14]. Other imaging examinations such as ultrasound, positron emission tomography scan, and magnetic resonance imaging are of little value in identification of the malignant transformation. 3D technology helps further to clarify the relationship of the tumor and its adjacent organs and important blood vessels. It is difficult to make an accurate diagnosis based on clinical presentation and imaging. The definite pre-operative diagnosis requires histopathologic and immunohistochemical analysis of tis-

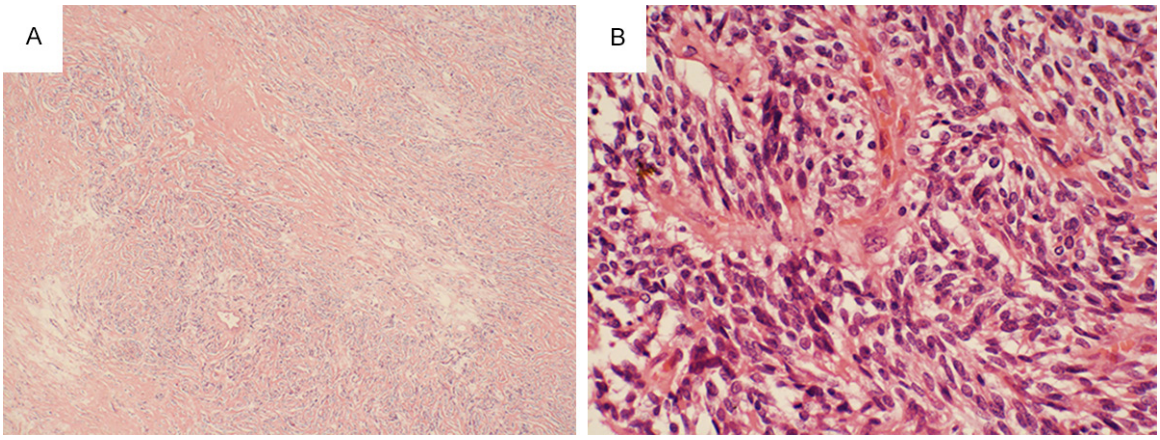

Figure 3. Histopathologic examination revealed spindle-shaped tumor cells mixed with varying amounts of collagen, and hyalinization of fibrous tissue.

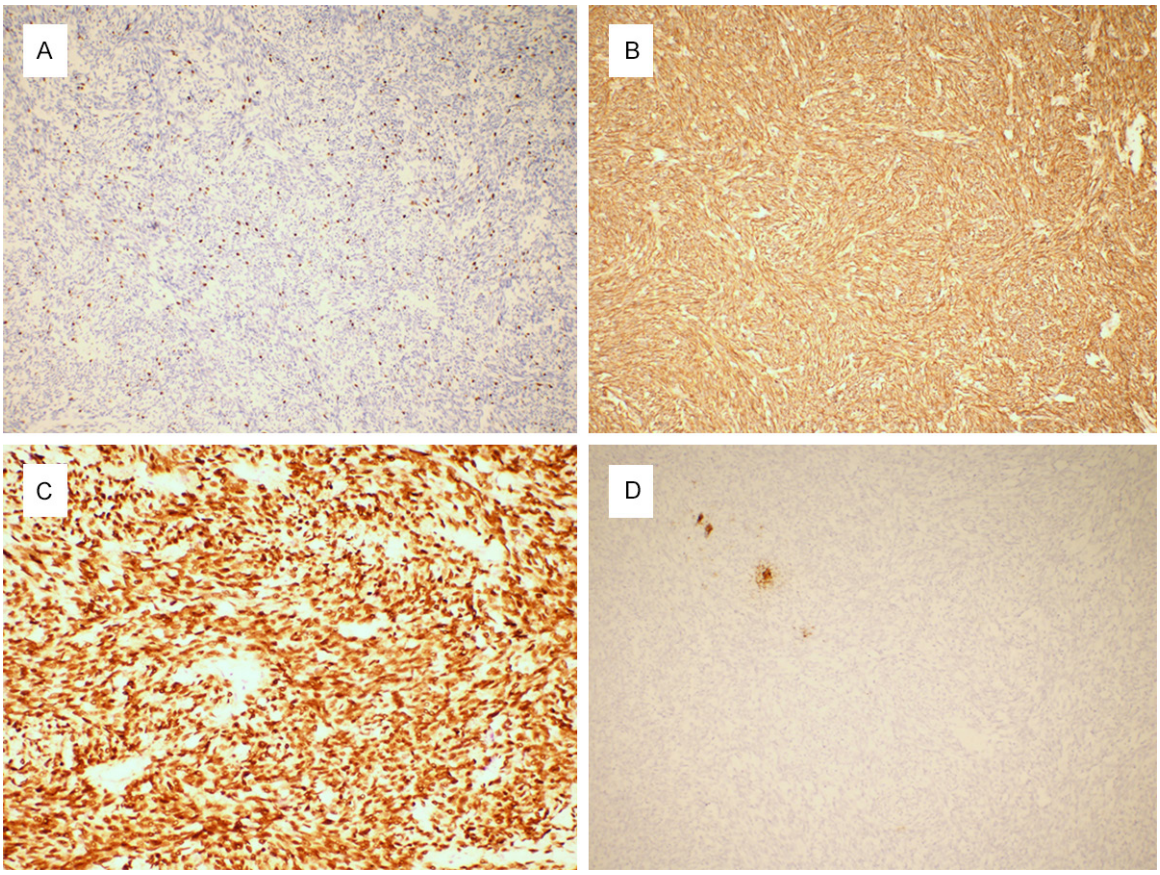

Figure 4. Immunohistochemical stains demonstrated that the tumor cells were positive for Ki-67 (A), Vimentin (B), and STAT6 (C), and negative for PCK (D).

sues obtained by transthoracic needle biopsy, which can also be used to differentiate benign and malignant features and other soft tissue tumors [12]. Because this tumor is often located close to the chest wall, biopsy is relatively simple and safe with few complications [15]. Histopathologically, benign SFTPs are hypocellular in character, and the presence of hypercellular, pleomorphism, mitotic activity, and necrosis may indicate aggressive behavior.

Immunohistochemical examinations play a significant role in the diagnosis of SFTPs and exclusion of sarcomatoid carcinoma of the lung, mesothelioma, synovial sarcoma, and other rare sarcomas [16]: CD34, CD99, Vimentin and BCL-2 are positively expressed, and expression of strong nuclear STAT6 is largely specific for SFTP; whereas S-100 is negative [12, 17].

Given their rarity, limited data exist on the treatment of SFTPs, and the management is based on expert opinions. The primary treatment for SFTPs is complete surgical resection including lobectomy, which has a good prognosis, and a second surgical resection should be performed for recurrence. The difficulty of surgery depends on the size of the tumor, poor exposure, and pleural adhesions. The preoperative embolization of the abundant blood supply and feeding arteries of the tumors should be performed in giant SFTPs in order to reduce the risk of massive hemorrhage [18]. Chemotherapy and radiotherapy may have potential roles in the treatment algorithm; however, no current standard protocols exist. Long-term survival after complete resection of benign SFTPs is excellent. Benign lesions may also recur and metastasize, mainly depending on the histologic characteristics of SFTPs and radical nature of the surgery, so long-term surveillance of patients after surgery is recommended [19].

Conclusion

The present case described a rare giant SFTP that grew very rapidly, which might be related to taking of unknown herbs. The contrast-enhanced CT and 3D technology helped to make the final surgical plan by clarifying the tumor and its adjacent organs and important blood vessels. The preoperative embolization of the tumor-supplying arteries was effective to reduce the risk of massive hemorrhage. The confirmed diagnosis depended on post-operative histologic and immunohistochemical examinations. Surgical resection with wide negative margins constitutes the predominant mode of therapy at present.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Ye Yuan, Department of Anesthesiology, The First Hospital of Jilin University, 71 Xinmin Street, Changchun, Jilin, People's Republic of China. Tel: 86-13604430557; Fax: 86-10-88782955; E-mail: yeyuan1225@sina.com

References

[1] DeVito N, Henderson E, Han G, Reed D, Bui MM, Lavey R, Robinson L, Zager JS, Gonzalez RJ, Sondak VK, Letson GD and Conley A. Clinical characteristics and outcomes for solitary fibrous tumor (SFT): a single center experience. PLoS One 2015; 10: e0140362.

[2] Kim HJ, Lee HK, Seo JJ, Kim HJ, Shin JH, Jeong AK, Lee JH and Cho KJ. MR imaging of solitary fibrous tumors in the head and neck. Korean J Radiol 2005; 6: 136-142.

[3] Lahon B, Mercier O, Fadel E, Ghigna MR, Petkova B, Mussot S, Fabre D, Le Chevalier T and Dartevelle P. Solitary fibrous tumor of the pleura: outcomes of 157 complete resections in a single center. Ann Thorac Surg 2012; 94: 394-400.

[4] Sung SH, Chang JW, Kim J, Lee KS, Han J and Park SI. Solitary fibrous tumors of the pleura: surgical outcome and clinical course. Ann Thorac Surg 2005; 79: 303-307.

[5] Pusiol T, Piscioli I, Scialpi M and Hanspeter E. Giant benign solitary fibrous tumour of the pleura (> 15 cm): role of radiological pathological correlations in management. Report of 3 cases and review of the literature. Pathologica 2013; 105: 77-82.

[6] Chick JF, Chauhan NR and Madan R. Solitary fibrous tumors of the thorax: nomenclature, epidemiology, radiologic and pathologic findings, differential diagnoses, and management. AJR Am J Roentgenol 2013; 200: W238-248.

[7] Harrison-Phipps KM, Nichols FC, Schleck CD, Deschamps C, Cassivi SD, Schipper PH, Allen MS, Wigle DA and Pairolero PC. Solitary fibrous tumors of the pleura: results of surgical treatment and long-term prognosis. J Thorac Cardiovasc Surg 2009; 138: 19-25.

[8] Furukawa N, Hansky B, Niedermeyer J, Gummert J and Renner A. A silent gigantic solitary fibrous tumor of the pleura: case report. J Cardiothorac Surg 2011; 6: 122.

[9] Jadczak P, Guz W, Kaznowska E, Ramotowski R, Szalacha-Tarala E, Gorecki A and Samojedny A. Solitary fibrous tumour of the pleura - cases analysis. Pol J Radiol 2014; 79: 368-373.

[10] Abe M, Nomori H, Fukazawa M, Sugimura H, Narita M and Takeshi A. Giant solitary fibrous tumor of the pleura causing respiratory insufficiency: report of 3 cases. Ann Thorac Cardiovasc Surg 2014; 20 Suppl: 441-444.

[11] de Perrot M, Kurt AM, Robert JH, Borisch B and Spiliopoulos A. Clinical behavior of solitary fi-

brous tumors of the pleura. Ann Thorac Surg 1999; 67: 1456-1459.
[12] de Perrot M, Fischer S, Brundler MA, Sekine Y and Keshavjee S. Solitary fibrous tumors of the pleura. Ann Thorac Surg 2002; 74: 285-293.
[13] Song SW, Jung JI, Lee KY, Kim MY and Park SH. Malignant solitary fibrous tumor of the pleura: computed tomography-pathological correlation and comparison with computed tomography of benign solitary fibrous tumor of the pleura. Jpn J Radiol 2010; 28: 602-608.
[14] Cardillo G, Lococo F, Carleo F and Martelli M. Solitary fibrous tumors of the pleura. Curr Opin Pulm Med 2012; 18: 339-346.
[15] Ali SZ, Hoon V, Hoda S, Heelan R and Zakowski MF. Solitary fibrous tumor. A cytologic-histologic study with clinical, radiologic, and immunohistochemical correlations. Cancer 1997; 81: 116-121.
[16] Abu Arab W. Solitary fibrous tumours of the pleura. Eur J Cardiothorac Surg 2012; 41: 587-597.
[17] Demicco EG, Harms PW, Patel RM, Smith SC, Ingram D, Torres K, Carskadon SL, Camelo-Piragua S, McHugh JB, Siddiqui J, Palanisamy N, Lucas DR, Lazar AJ and Wang WL. Extensive survey of STAT6 expression in a large series of mesenchymal tumors. Am J Clin Pathol 2015; 143: 672-682.
[18] Perrotta F, Cerqua FS, Cammarata A, Izzo A, Bergaminelli C, Curcio C, Guarino C, Grella E, Forzano I, Cennamo A, Tafuri D, Rocca A, Bianco A and Mazzarella G. Integrated therapeutic approach to giant solitary fibrous tumor of the pleura: report of a case and review of the literature. Open Med (Wars) 2016; 11: 220-225.
[19] Cardillo G, Carbone L, Carleo F, Masala N, Graziano P, Bray A and Martelli M. Solitary fibrous tumors of the pleura: an analysis of 110 patients treated in a single institution. Ann Thorac Surg 2009; 88: 1632-1637.