

Chapter 25

SOLITARY FIBROUS TUMOR

Fatma BUĞDAYCI BAŞAL¹

INTRODUCTION

Solitary fibrous tumor (SFT) is a rare malignancy originating body cavity sites includes pleura, peritoneum, and meninges. It arises from extrathoracic fibroblastic mesenchymal structures unlike known it arises inside of thorax (1,2). It was first described in pleura in 1931 (3). Solitary fibrous tumor's epidemiology, clinic presentation, diagnosis, differential diagnosis, and treatment are reviewed here, especially pleural SFT.

EPIDEMIOLOGY AND RISK FACTORS

The solitary fibrous tumor is a rare tumor seen less than %2 among soft tissue tumors and also pleural SFTs account for less than 5 percent of all tumors arising from the pleura (4-7). The most common seen age is fifth and seventh decades but it may arise at any age (8-10). The solitary fibrous tumor occurs with equal frequency for men and women and most often occur during middle age, although it is occasionally seen in younger people. The etiology of SFTs remains unknown, and no association has been demonstrated with risk factors predisposing SFT, as tobacco, radiation, asbestos and the other environmental toxicants (5,11,12).

CLINIC PRESENTATION

The solitary fibrous tumor is a slow-growing and long-standing tumor that likes to be seen in serosal membranes, dura of meninges, cavity sites and deep soft tissues. Most tumors are slow growing and are diagnosed incidentally as painless masses (12). For intrathoracic SFT, the originating tissues are pleura, lung parenchyma and mediastinum. Another area that extrathoracic SFT arises from peritoneum, retroperitoneum and pelvic site (13). The liver is the most common involvement visceral organ with the urogenital tract. When pelvic involvement is large, it may not be possible to assign the primary organ. The solitary fibrous tu-

¹ MD,Health Science University Dr. A.Y. Ankara Oncology Research and Training Hospital, Department of Medical Oncology , dr.fatmabb@gmail.com

areas have very wide range from pleura to meninges moreover breast, tyroid, retroperitoneum. The majority of SFTs behave in an indolent fashion and do not recur locally or distantly. The patients have malign histological criterians may recur who are in %10-25 percent part. The main treatment is complete resection. Adjuvant radiotherapy or chemotherapy do not require any benefit in complete resected patients. Radiotherapy may be considered for incomplete resected cases. Cytotoxic chemotherapy can provide stabil responses generally in advanced SFT. Adding of the molecular targeting therapies, encouraging results are expected but some new trials are needed to understand the best model of therapy.

REFERENCES

1. Travis WD, Churg A, Aubry MC, et al. Mesenchymal tumours. In: Tumors of the Lung, Pleura, Thymus and Heart, Travis WD, Brambilla E, Muller-Hermelink HK, and Harris CC (Eds), IARC Press, Lyon 2004. p.142.
2. Fletcher CD, Bridge JA, Lee JC. Extrapleural solitary fibrous tumor. In: WHO Classification of Tumours of Soft Tissue and Bone, Fletcher CD, Bridge JA, Hogendoorn PC, Mertens F (Eds), IARC Press, Lyon 2013. p.80.
3. Klemperer P, Rabin CB. Primary neoplasms of the pleura: a report of five cases. Arch Pathol. 1931;11:385.
4. Gold JS, Antonescu CR, Hajdu C, et al. Clinicopathologic correlates of solitary fibrous tumors. Cancer 2002; 94:1057.
5. Chick JF, Chauhan NR, 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.
6. Sung SH, Chang JW, Kim J, et al. Solitary fibrous tumors of the pleura: surgical outcome and clinical course. Ann Thorac Surg 2005; 79:303.
7. Cardillo G, Lococo F, Carleo F, Martelli M. Solitary fibrous tumors of the pleura. Curr Opin Pulm Med 2012; 18:339.
8. Fletcher CDM, Bridge JA, Lee, J-C. Extrapleural solitary fibrous tumor. In: WHO Classification of Tumours of Soft Tissue and Bone, 4th, Fletcher CDM, Bridge, JA, Hogendoorn CW, Mertens F (Eds), International Agency for Research on Cancer, Lyon 2013.
9. Travis WD, Churg A, Aubry MC, Ordonez NG. Mesenchymal Tumours. In: Pathology and Genetics of Tumours of the Lung, Pleura, Thymus, and Heart, 1st, Travis WD, Brambilla E, Muller-Hermelink HK, Harris CC (Eds), International Agency for Research on Cancer, Lyon 2004. p.141.
10. Tapias LF, Mercier O, Ghigna MR, et al. Validation of a scoring system to predict recurrence of resected solitary fibrous tumors of the pleura. Chest 2015; 147:216.
11. Elizabeth G Demicco, Christian Meyer, Solitary fibrous tumor (<http://www.uptodate.com>) Nisan 2018, erişim tarihi Ocak 2019.
12. Mert Saynak, Nirmal K. Veeramachaneni, Jessica L. Hubbs et al. Solitary Fibrous Tumors of Chest: Another Look with the Oncologic Perspective. Balkan Med J. 2017 May; 34(3): 188–199.
13. Meittinen Markku. Chapter 12: Solitary fibrous tumor, hemaangiopericytoma, and related tumors.. In: Modern Soft Tissue Pathology: Tumours and Non-Neoplastic

- Conditions, Miettinen, Markku (Eds), Cambridge University Press, New York 2010. p.335.
14. Demicco EG, Park MS, Araujo DM, et al. Solitary fibrous tumor: a clinicopathological study of 110 cases and proposed risk assessment model. *Mod Pathol* 2012; 25:1298.
 15. Espot NJ, Lewis JJ, Leung D, et al. Conventional hemangiopericytoma: modern analysis of outcome. *Cancer* 2002; 95:1746.
 16. Hasegawa T, Matsuno Y, Shimoda T, et al. Extrathoracic solitary fibrous tumors: their histological variability and potentially aggressive behavior. *Hum Pathol* 1999; 30:1464.
 17. van Houdt WJ, Westerveld CM, Vrijenhoek JE, et al. Prognosis of solitary fibrous tumors: a multicenter study. *Ann Surg Oncol* 2013; 20:4090.
 18. Brunnemann RB, Ro JY, Ordonez NG, et al. Extrapleural solitary fibrous tumor: a clinicopathologic study of 24 cases. *Mod Pathol* 1999; 12:1034.
 19. Daigeler A, Lehnhardt M, Langer S, et al. Clinicopathological findings in a case series of extrathoracic solitary fibrous tumors of soft tissues. *BMC Surg* 2006; 6:10.
 20. Erdag G, Qureshi HS, Patterson JW, Wick MR. Solitary fibrous tumors of the skin: a clinicopathologic study of 10 cases and review of the literature. *J Cutan Pathol* 2007; 34:844.
 21. Lococo F, Cesario A, Cardillo G, et al. Malignant solitary fibrous tumors of the pleura: retrospective review of a multicenter series. *J Thorac Oncol* 2012; 7:1698.
 22. Lahon B, Mercier O, Fadel E, et al. Solitary fibrous tumor of the pleura: outcomes of 157 complete resections in a single center. *Ann Thorac Surg* 2012; 94:394.
 23. Tapias LF, Mino-Kenudson M, Lee H, et al. Risk factor analysis for the recurrence of resected solitary fibrous tumours of the pleura: a 33-year experience and proposal for a scoring system. *Eur J Cardiothorac Surg* 2013; 44:111.
 24. de Perrot M, Kurt AM, Robert JH, et al. Clinical behavior of solitary fibrous tumors of the pleura. *Ann Thorac Surg* 1999; 67:1456.
 25. Briselli M, Mark EJ, Dickersin GR. Solitary fibrous tumors of the pleura: eight new cases and review of 360 cases in the literature. *Cancer*. 1981;47:2678–89.
 26. Fung EC, Crook MA. Doege-Potter syndrome and ‘big-IGF2’: a rare cause of hypoglycaemia. *Ann Clin Biochem* 2011; 48:95.
 27. Herrmann BL, Saller B, Kiess W, et al. Primary malignant fibrous histiocytoma of the lung: IGF-II producing tumor induces fasting hypoglycemia. *Exp Clin Endocrinol Diabetes* 2000; 108:515.
 28. Tominaga N, Kawarasaki C, Kanemoto K, et al. Recurrent solitary fibrous tumor of the pleura with malignant transformation and non-islet cell tumor-induced hypoglycemia due to paraneoplastic overexpression and secretion of high-molecular-weight insulin-like growth factor II. *Intern Med* 2012; 51:3267.
 29. Cardillo G, Facciolo F, Cavazzana AO, et al. Localized (solitary) fibrous tumors of the pleura: an analysis of 55 patients. *Ann Thorac Surg* 2000; 70:1808.
 30. Rena O, Filosso PL, Papalia E, et al. Solitary fibrous tumour of the pleura: surgical treatment. *Eur J Cardiothorac Surg* 2001; 19:185.
 31. Magdeleinat P, Alifano M, Petino A, et al. Solitary fibrous tumors of the pleura: clinical characteristics, surgical treatment and outcome. *Eur J Cardiothorac Surg* 2002; 21:1087.
 32. Shanbhogue AK, Prasad SR, Takahashi N, et al. Somatic and visceral solitary fibrous tumors in the abdomen and pelvis: cross-sectional imaging spectrum. *Radiographics* 2011; 31:393.

33. Rosenkrantz AB, Hindman N, Melamed J. Imaging appearance of solitary fibrous tumor of the abdominopelvic cavity. *J Comput Assist Tomogr* 2010; 34:201.
34. Kruse M, Sherry SJ, Paidpally V, Mercier G, Subramaniam RM. FDG PET/CT in the management of primary pleural tumors and pleural metastases. *AJR Am J Roentgenol.* 2013;201:W215–26.
35. Khanchel F, Driss M, Mrad K, Romdhane KB. Malignant solitary fibrous tumor in the extremity: Cytopathologic findings. *J Cytol.* 2012;29:139–41.
36. Dey P. Diagnostic dilemma: diagnostic algorithm in fine needle aspiration cytology of mediastinal tumors. *Indian J Pathol Microbiol.* 2010;53:395–402.
37. Dey P. Diagnostic dilemma: diagnostic algorithm in fine needle aspiration cytology of mediastinal tumors. *Indian J Pathol Microbiol.* 2010;53:395–402.
38. Weynand B, Noël H, Goncette L, Noirhomme P, Collard P. Solitary fibrous tumor of the pleura: a report of five cases diagnosed by transthoracic cutting needle biopsy. *Chest.* 1997;112:1424–8.
39. Scarsbrook AF, Evans AL, Slade M, Gleeson FV. Recurrent solitary fibrous tumour of the pleura due to tumour seeding following ultrasound-guided transthoracic biopsy. *Clin Radiol.* 2005;60:130–2.
40. Robinson LA. Solitary fibrous tumor of the pleura. *Cancer Control.* 2006;13:264–9.
41. England DM, Hochholzer L, McCarthy MJ. Localized benign and malignant fibrous tumors of the pleura. A clinicopathologic review of 223 cases. *Am J Surg Pathol* 1989; 13:640.
42. Cranshaw IM, Gikas PD, Fisher C, et al. Clinical outcomes of extra-thoracic solitary fibrous tumours. *Eur J Surg Oncol* 2009; 35:994.
43. Vallat-Decouvelaere AV, Dry SM, Fletcher CD. Atypical and malignant solitary fibrous tumors in extrathoracic locations: evidence of their comparability to intra-thoracic tumors. *Am J Surg Pathol* 1998; 22:1501.
44. Chang YL, Lee YC, Wu CT. Thoracic solitary fibrous tumor: clinical and pathological diversity. *Lung Cancer* 1999; 23:53.
45. Yokoi T, Tsuzuki T, Yatabe Y, et al. Solitary fibrous tumour: significance of p53 and CD34 immunoreactivity in its malignant transformation. *Histopathology* 1998; 32:423
46. Herawi M, Epstein JI. Solitary fibrous tumor on needle biopsy and transurethral resection of the prostate: a clinicopathologic study of 13 cases. *Am J Surg Pathol* 2007; 31:870.
47. Nielsen GP, O’Connell JX, Dickersin GR, Rosenberg AE. Solitary fibrous tumor of soft tissue: a report of 15 cases, including 5 malignant examples with light microscopic, immunohistochemical, and ultrastructural data. *Mod Pathol* 1997; 10:1028.
48. de Perrot M, Fischer S, Bründler MA, et al. Solitary fibrous tumors of the pleura. *Ann Thorac Surg* 2002; 74:285.
49. Brozzetti S, D’Andrea N, Limiti MR, et al. Clinical behavior of solitary fibrous tumors of the pleura. An immunohistochemical study. *Anticancer Res* 2000; 20:4701.
50. Hasegawa T, Matsuno Y, Shimoda T, et al. Frequent expression of bcl-2 protein in solitary fibrous tumors. *Jpn J Clin Oncol* 1998; 28:86.
51. Suter M, Gebhard S, Boumghar M, et al. Localized fibrous tumours of the pleura: 15 new cases and review of the literature. *Eur J Cardiothorac Surg* 1998; 14:453.
52. Lee JC, Fletcher CD. Malignant fat-forming solitary fibrous tumor (so-called “lipomatous hemangiopericytoma”): clinicopathologic analysis of 14 cases. *Am J Surg Pathol* 2011; 35:1177.

53. Robinson DR, Wu YM, Kalyana-Sundaram S, et al. Identification of recurrent NAB2-STAT6 gene fusions in solitary fibrous tumor by integrative sequencing. *Nat Genet* 2013; 45:180.
54. Chmielecki J, Crago AM, Rosenberg M, et al. Whole-exome sequencing identifies a recurrent NAB2-STAT6 fusion in solitary fibrous tumors. *Nat Genet* 2013; 45:131.
55. Mohajeri A, Tayebwa J, Collin A, et al. Comprehensive genetic analysis identifies a pathognomonic NAB2/STAT6 fusion gene, nonrandom secondary genomic imbalances, and a characteristic gene expression profile in solitary fibrous tumor. *Genes Chromosomes Cancer* 2013; 52:873.
56. Barthelmeß S, Geddert H, Boltze C, et al. Solitary fibrous tumors/hemangiopericytomas with different variants of the NAB2-STAT6 gene fusion are characterized by specific histomorphology and distinct clinicopathological features. *Am J Pathol* 2014; 184:1209.
57. Fabio Perrotta,¹ Francesco Saverio Cerqua,² Antonino Cammarata et al. 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(1): 220–225.
58. Walker CM, Takasugi JE, Chung JH, et al. Tumorlike conditions of the pleura. *Radiographics* 2012; 32:971.
59. Galateau-Salle F, Churg A, Roggli V, Travis WD World Health Organization Committee for Tumors of the Pleura. The 2015 World Health Organization Classification of Tumors of the Pleura: Advances since the 2004 Classification. *J Thorac Oncol*. 2016;11:142–54.
60. Chen N, Slater K. Solitary fibrous tumour of the liver-report on metastasis and local recurrence of a malignant case and review literature. *World J Surg Oncol*. 2017 Jan 18;15(1):27.
61. Ghasemi-Rad M, Wang KY, Jain S et al. Solitary fibrous tumor of thyroid: a case report with review of literature. *Clin Imaging*. 2019 Jan - Feb;53:105-107.
62. Zhou Y, Chu X, Yi Y et al. Malignant solitary fibrous tumor in retroperitoneum: A case report and literature review. *Medicine (Baltimore)*. 2017 Mar;96(11):e6373
63. Tsai SY1, Hsu CY2, Chou YH et al. Solitary fibrous tumor of the breast: A case report and review of the literature. *J Clin Ultrasound*. 2017 Jul 8;45(6):350-354.
64. Cheng K, Gu S, Gao B et al. Analysis of misdiagnosis of 38 cases of solitary fibrous tumor of the pleura. *Zhonghua Jie He He Hu Xi Za Zhi*. 2015 Jul;38(7):520-3.
65. Constantinidou A, Jones RL, Olmos D, et al. Conventional anthracycline-based chemotherapy has limited efficacy in solitary fibrous tumour. *Acta Oncol* 2012; 51:550.
66. Wilky BA, Montgomery EA, Guzzetta AA, et al. Extrathoracic location and “borderline” histology are associated with recurrence of solitary fibrous tumors after surgical resection. *Ann Surg Oncol* 2013; 20:4080.
67. Spitz FR, Bouvet M, Pisters PW, et al. Hemangiopericytoma: a 20-year single-institution experience. *Ann Surg Oncol* 1998; 5:350.
68. Stacchiotti S, Libertini M, Negri T, et al. Response to chemotherapy of solitary fibrous tumour: a retrospective study. *Eur J Cancer* 2013; 49:2376.
69. Park MS, Ravi V, Conley A, et al. The role of chemotherapy in advanced solitary fibrous tumors: a retrospective analysis. *Clin Sarcoma Res* 2013; 3:7
70. Stacchiotti S, Saponara M, Frapolli R, et al. Patient-derived solitary fibrous tumour xenografts predict high sensitivity to doxorubicin/dacarbazine combination confirmed in the clinic and highlight the potential effectiveness of trabectedin or eribulin against this tumour. *Eur J Cancer* 2017; 76:84.

71. Park MS, Patel SR, Ludwig JA, et al. Activity of temozolomide and bevacizumab in the treatment of locally advanced, recurrent, and metastatic hemangiopericytoma and malignant solitary fibrous tumor. *Cancer* 2011; 117:4939.
72. Choi H, Charnsangavej C, Faria SC, et al. Correlation of computed tomography and positron emission tomography in patients with metastatic gastrointestinal stromal tumor treated at a single institution with imatinib mesylate: proposal of new computed tomography response criteria. *J Clin Oncol* 2007; 25:1753.
73. George S, Merriam P, Maki RG, et al. Multicenter phase II trial of sunitinib in the treatment of nongastrointestinal stromal tumor sarcomas. *J Clin Oncol* 2009; 27:3154.
74. Stacchiotti S, Tortoreto M, Baldi GG, et al. Preclinical and clinical evidence of activity of pazopanib in solitary fibrous tumour. *Eur J Cancer* 2014; 50:3021.
75. Valentin T, Fournier C, Penel N, et al. Sorafenib in patients with progressive malignant solitary fibrous tumors: a subgroup analysis from a phase II study of the French Sarcoma Group (GSF/GETO). *Invest New Drugs* 2013; 31:1626.