



Solitary Fibrous Tumor of Rectus Sheath

Po-Hsien Wu¹, Huai-En Lu²

¹Department of Surgery, Division of Colon and Rectal Surgery, Tri-Service General Hospital, National Defense Medical Center, ²Department of Surgery, Song Shan Branch, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan

Solitary fibrous tumor (SFT) is a rare tumor with incidence <2% in all soft tissue tumors. About 30% of SFTs arise in thoracic cavity, 30% arise from the intra-abdominal cavity, and another 20% occur in head and neck. We presented a case with a painless, slow-growing intra-abdominal tumor for more than a decade. Physical examinations, blood test, and image studies provide little clues preoperatively. Definite diagnosis relies on pathological confirmation. Currently, there are numerous immunohistochemistry tools to facilitate making diagnosis correctly.

Key words: Solitary fibrous tumor, extrathoracic, clinicopathological features, surgery, tumor of rectus sheath

INTRODUCTION

Solitary fibrous tumors (SFTs) are uncommon mesenchymal neoplasms first described in 1931¹ and account <2% of all soft tissue tumors.² Initially, SFTs are thought to be intrathoracic tumors. Moreover, recent reports unveil that SFTs occur almost everywhere of human body including the intra-abdominal cavity, central nervous system, head and neck, bone, salivary gland, thyroid, liver, and prostate.³ SFTs of the abdominal wall are extremely uncommon and there are only 16 cases had been reported till date.⁴ SFTs are most commonly diagnosed from fifth to seventh decades, and the mean age is 58 years. There is no sex predilection.² In this report, we describe a new case of SFT that originates from rectus sheath and discuss the clinical symptoms, signs, image findings, and pathological characteristics.

CASE REPORT

A 42-year-old female presented to our surgery department because of a slow-growing abdominal mass for the past 10 years. Her family and medical history were unremarkable, and she had no history of previous abdominal surgery. There

was no nausea, vomiting, involuntary body weight loss, and pain. On physical examination, a firm, movable, painless mass in the left-sided abdomen on deep palpation. Laboratory tests including metabolic panels, tumor markers, and C-reactive protein were within normal range. Computed tomography (CT) with intravenous contrast showed a well-circumscribed mass, measuring 6 cm × 4.6 cm × 6.7 cm, with heterogeneous enhancement [Figure 1] attached to rectus sheath. Magnetic resonance (MR) imaging disclosed a bright high-signal tumor mass after gadolinium administration [Figure 2].

At laparotomy, a large, encapsulated tumor adhered to rectus sheath was found. The tumor did not invade to adjacent visceral organs. Grossly, the tumor was well capsulated, soft consistence, and covered with multiple vessels [Figure 3]. The size was about 7 cm × 5 cm × 4 cm. Histologically, the sections showed proliferative spindle cells separated by normal-looking endothelial cells lining small vessels and focally staghorn configuration. Furthermore, the neoplasm had uncommon mitotic activity (2/10 hpf), and there was no necrosis [Figure 4] in fields. Immunohistochemically, the tumor was positive for CD34 [Figure 5] and STAT6 [Figure 6] but negative for S100 protein, smooth muscle actin, and DOG1. Hence, the histological feature was compatible with benign SFT.

Received: November 04, 2018; Revised: January 24, 2019; Accepted: February 26, 2019

Corresponding Author: Dr. Po-Hsien Wu, Department of Surgery, Division of Colorectal Surgery, Tri-Service General Hospital, National Defense Medical Center, 325 Chenggong Road, Section 2, Taipei 114, Taiwan. Tel: +886-2-87923311; Fax: +886-2-87927245. E-mail: wupohsien@office365.ndmctsgh.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Wu PH, Lu HE. Solitary fibrous tumor of rectus sheath. J Med Sci 2019;39:193-6.

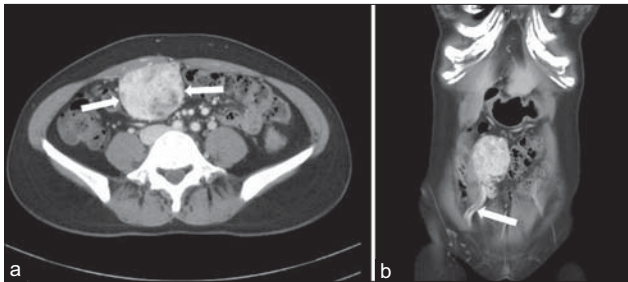


Figure 1: Abdominal computed tomography showing a well-circumscribed tumor mass attached to the rectus muscle and sheath. Axial (a) and coronal (b) contrast-enhanced computed tomography scans showing a heterogeneous enhancing mass (arrows in a) that displaces small bowel and also serpentine vessels near the mass is seen (arrow in b)



Figure 3: Intraoperative findings: A 7 cm × 5 cm × 4 cm encapsulated mass with multiple feeding vessels on tumor surface

The postoperative course was uneventful, and she was discharged 5 days after the surgery.

DISCUSSION

Clinical symptoms and signs of SFTs vary based on the location and size of the tumor. A palpable, nontender, slow-growing mass is the most common presentation. Large tumor may cause gastrointestinal symptoms such as nausea, vomiting, and constipation.

Imaging modalities such as CT and MR could help to diagnose SFT. On CT, SFT shows a well-circumscribed and hypervascular mass with external compression of adjacent organs. Calcification is rare in SFT. Central hypodense indicates tumor necrosis or cystic change.⁵ In our case, the CT images are compatible with these image features. On MR, SFT usually presents with intermediate intensity on T1-weighted image and heterogenous hypointense on T2-weighted image, which is related to hypercellularity or collagen. Intense

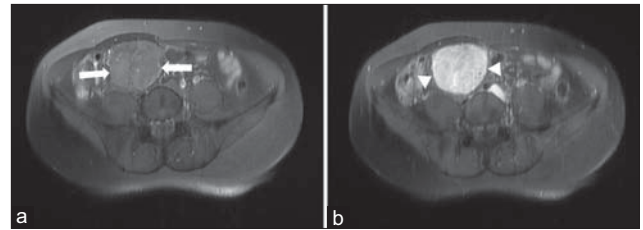


Figure 2: Axial fat-saturated T1-weighted magnetic resonance image revealed a mildly hyperintense lobulated mass (arrows in a) and heterogeneous high signal intensity after gadolinium administration (arrowheads in b)

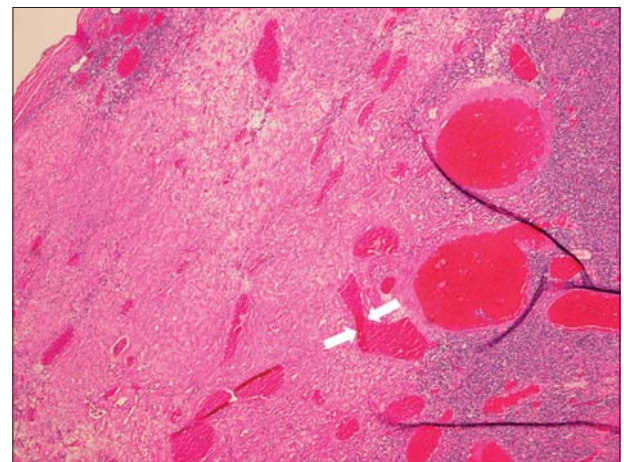


Figure 4: Histological findings (H and E) showing alternating hypocellular and hypercellular areas and several prominent staghorn capillaries

enhancement is seen after the administration of gadolinium contrast material.^{5,6} However, this case demonstrates a high signal intensity on T1-weighted image before administration of gadolinium and is less common.

Recognizing typical morphology and specific immunohistochemical findings are keys to diagnose SFT on histological examination. Grossly, the tumor usually presents a sharply circumscribed mass with smooth, bosselated, or lobulated external surface.⁷ Microscopically, classical morphological appearance including “patternless pattern” caused by variable spindle cells arrayed in the stroma. Furthermore, prominent and anastomosing “staghorn” capillaries are also observed.^{8,9} Enzinger and Smith published criteria to identify malignant SFT including ≥4 mitotic figures/10 hpf, larger size, high cellularity, nuclear pleomorphism, and presence of hemorrhage or necrosis.¹⁰ Since the histologic characteristics are relatively nonspecific to diagnose SFT, immunohistochemistry plays a vital role to help diagnose and differentiate from other spindle cell tumors. Conventional markers of SFT including diffuse strong positive of CD34,¹¹ moderately positive of Bcl-2 and CD99, and absence of S100, actin, and DOG1.^{4,8}

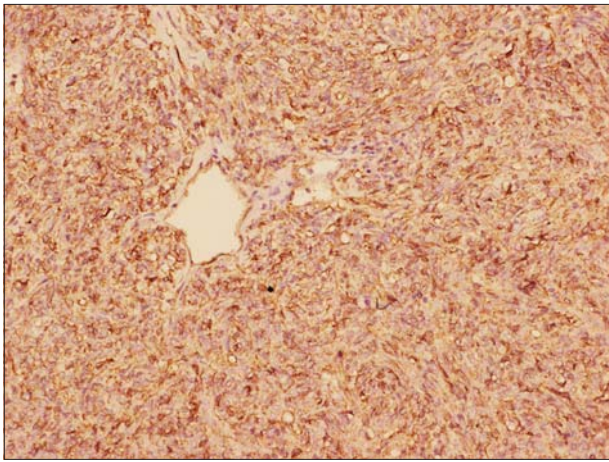


Figure 5: Immunohistochemical features of the neoplasm: Diffusely immunoreactive to CD34

Furthermore, newer immunohistochemistry STAT6 test to detect NAB2-STAT6 fusion gene had been used to differentiate SFT from other mimics and reached significant high sensitivity and specificity.^{12,13} In our case, both histologic characteristics and immunohistochemistry tests showed classic SFT appearance.

Differential diagnosis of tumor arising from rectus sheath including desmoid tumor, hematoma, and infection.¹⁴ The most common tumor arising from rectus sheath is desmoid tumor. Other etiologies including hematoma and infection processes such as scar abscess or diverticulitis. Clinical presentations and image characteristics of desmoid tumor on CT and MR mimic SFTs.⁵ Therefore, surgical pathology is the cornerstone of making diagnosis. Unlike SFTs, desmoid tumor shows reactive actin and nonreactive CD34 on immunohistochemistry tests.¹⁵ Hematoma of rectus sheath cannot be accurately diagnosed solely based on symptoms and signs (such as anorexia, nausea, fever, and tachycardia) due to lack of specificity. Hence, ultrasound or CT plays a role of making diagnosis and furthermore preventing unnecessary surgery. Most hematomas could be managed conservatively. The third common mass of rectus sheath is infection. When a mass accompanied with fever, tenderness and leukocytosis raise the suspicion of infection. Numerous infectious processes such as scar abscess, diverticulitis, and Crohn's disease had been reported to cause rectus sheath mass. Some will be resolved with antibiotics alone and others may need surgical or percutaneous drainage.¹⁴

Surgical resection with curative intent is the mainstay therapy for SFT.¹⁶ Adjuvant radiation therapy and chemotherapy are not recommended contemporarily because of the lack of validating data to support these treatments. Newer molecularly target agent therapy is still under-investigated.

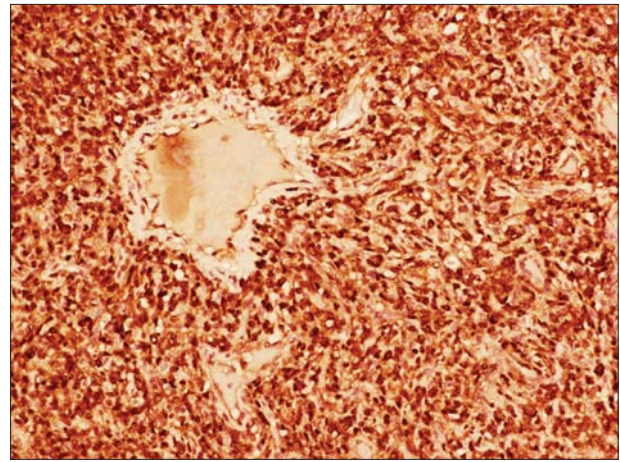


Figure 6: Immunohistochemical features of the neoplasm: Strong nuclear STAT6 antibody reactivity whereas vascular endothelial cells are negative

CONCLUSION

We present a rare SFT of rectus sheath in a middle-aged woman. Preoperative diagnosis of the disease is difficult because of the nonspecific findings on physical examinations and imaging studies. Correct diagnosis of the tumor needs surgical *en bloc* resection and histopathological confirmation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Klemperer P, Coleman BR. Primary neoplasms of the pleura. A report of five cases. *Arch Pathol* 1931;11:385-412.
2. Gold JS, Antonescu CR, Hajdu C, Ferrone CR, Hussain M, Lewis JJ, *et al*. Clinicopathologic correlates of solitary fibrous tumors. *Cancer* 2002;94:1057-68.
3. Guillou L, Fletcher JA, Fletcher CD, Mandahl N. World Health Organization Classification of

- Tumours -Pathology and Genetics of Tumours of Soft Tissue and Bone. France: IARCPress; 2002.
4. Sahu A, Nahak SR, Nayak SK, Agrawal KC. Solitary fibrous tumor of rectus sheath: A rare case report. *Ann Int Med Dent Res* 2017;3:25-9.
 5. Shanbhogue AK, Prasad SR, Takahashi N, Vikram R, Zaheer A, Sandrasegaran K, *et al.* Somatic and visceral solitary fibrous tumors in the abdomen and pelvis: Cross-sectional imaging spectrum. *Radiographics* 2011;31:393-408.
 6. Zhanlong M, Haibin S, Xiangshan F, Jiacheng S, Yicheng N. Variable solitary fibrous tumor locations: CT and MR imaging features. *Medicine (Baltimore)* 2016;95:e3031.
 7. 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;13:640-58.
 8. Baliga M, Flowers R, Heard K, Siddiqi A, Akhtar I. Solitary fibrous tumor of the lung: A case report with a study of the aspiration biopsy, histopathology, immunohistochemistry, and autopsy findings. *Diagn Cytopathol* 2007;35:239-44.
 9. Hasegawa T, Matsuno Y, Shimoda T, Hasegawa F, Sano T, Hirohashi S, *et al.* Extrathoracic solitary fibrous tumors: Their histological variability and potentially aggressive behavior. *Hum Pathol* 1999;30:1464-73.
 10. Enzinger FM, Smith BH. Hemangiopericytoma. An analysis of 106 cases. *Hum Pathol* 1976;7:61-82.
 11. van de Rijn M, Lombard CM, Rouse RV. Expression of CD34 by solitary fibrous tumors of the pleura, mediastinum, and lung. *Am J Surg Pathol* 1994;18:814-20.
 12. Doyle LA, Vivero M, Fletcher CD, Mertens F, Hornick JL. Nuclear expression of STAT6 distinguishes solitary fibrous tumor from histologic mimics. *Mod Pathol* 2014;27:390-5.
 13. Yoshida A, Tsuta K, Ohno M, Yoshida M, Narita Y, Kawai A, *et al.* STAT6 immunohistochemistry is helpful in the diagnosis of solitary fibrous tumors. *Am J Surg Pathol* 2014;38:552-9.
 14. Lambroza A, Tighe MK, DeCosse JJ, Dannenberg AJ. Disorders of the rectus abdominis muscle and sheath: A 22-year experience. *Am J Gastroenterol* 1995;90:1313-7.
 15. Geramizadeh B, Marzban M, Churg A. Role of immunohistochemistry in the diagnosis of solitary fibrous tumor, a review. *Iran J Pathol* 2016;11:195-203.
 16. Espot NJ, Lewis JJ, Leung D, Woodruff JM, Antonescu CR, Shia J, *et al.* Conventional hemangiopericytoma: Modern analysis of outcome. *Cancer* 2002;95:1746-51.