

Laparoscopic Resection of a Solitary Fibrous Tumor of the Retroperitoneum, Report of a Case and Literature Review

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Introduction: Solitary fibrous tumor (SFT) is a rare mesenchymal tumor that is usually located in the pleural cavity, but it can arise in any part of the body. SFTs can develop in the extremities, in the head and neck region, meninges, orbits, thoracic wall, mediastinum, pericardium, peritoneum, retroperitoneum, pelvis, adrenal glands, liver and urogenital system.

Case Presentation: A 29-year-old female, was admitted in the gynaecology and obstetrics unit, for suspected pelvic tumor. CT scan showed a mass with hypervascular behavior and adherent to the uterus. The patient underwent laparoscopic resection of the mass. Abdominal exploration revealed a 5 cm mass, rising from the right iliac retroperitoneum above the pelvic space. The tumor was resected totally laparoscopically. The final histological findings were consistent with benign SFT. There were few mitotic cells (2×10 HPF) and a positivity for CD34 BCL2 and actin 1A4 and negative for CD117, desmin and CD99; Mib1 was $< 5\%$.

Conclusions: This is the first case of SFT of the retroperitoneum removed laparoscopically. The laparoscopic approach is useful for the advantages of minimally invasive surgery especially in case of pedunculate tumor as the case reported.

Keywords: Solitary Fibrous Tumor; Retroperitoneal Space; Laparoscopy

1. Introduction

Solitary Fibrous Tumor (SFT) is a rare mesenchymal neoplasm, described in 1931, for the first time, by Klemperer and Rabin as a pleura-based lesion (1). Despite this first description, it is now commonly accepted that SFTs is derived from mesenchymal rather than mesothelial cell, explaining numerous other extraserosal sites reported in literature (2). For this reason, it is now distinguished into two entities: pleural and extrapleural. Extrapleural SFTs can be found in any location: the most part is found in subcutaneous tissues, the remnants in the deep tissue of the extremities, in the head and neck region (including larynx, hypopharynx, parapharyngeal space, tongue, orbit, paranasal sinuses and nasal cavity), meninges, orbits, thoracic wall, mediastinum, pericardium, peritoneum, retroperitoneum, pelvis, adrenal glands, liver and urogenital system. Abdominal localizations are quite rare with 160 cases appearing in the literature (3), we report the first case of SFT of the retroperitoneum in the right iliac region, preoperative misdiagnosed with a pelvic tumor, treated by laparoscopic surgical resection.

2. Case Presentation

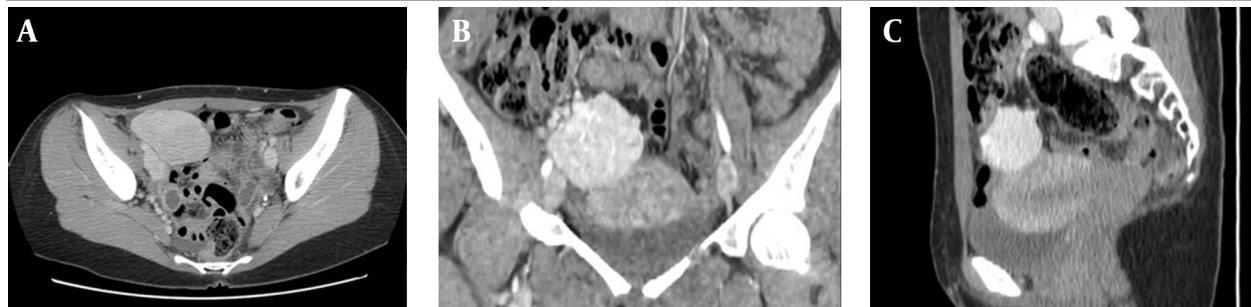
A 29-year-old female was admitted in the gynaecology

and obstetrics unit, for suspected pelvic tumor. During a follow-up control the transvaginal ultrasound showed a pelvic mass, suggesting an ovarian tumor. Past medical history revealed a laparoscopic resection for ovarian cyst 6 years before and diathermocoagulation of the portio 2 years before. Serum tumor marker levels, including CA125 and beta-HCG, were normal. Contrast enhanced CT scan of the abdomen and pelvis was performed confirming that mass with hypervascular behaviour and strictly adherent to the uterus (Figure 1). Thereby the patient underwent surgical resection of the mass by laparoscopic approach. Abdominal exploration revealed a 5 cm in diameter peduncolated mass, rising from the right iliac retroperitoneum above the pelvic space (Figure 2) and located between cecal appendix, external right iliac vessels and the ileopsoas muscle. The vascularisation of that mass was provided by new vessels from the infundibulo-pelvic ligament and external right vessels. After the opening of the peritoneum covering the mass, this was resected totally laparoscopically by using the vessel sealing system device (Ligasure TM). At the end of the procedure a closed drain was left in situ and the mass extracted by morcellation inside a plastic bag, without any spillage. The final histological findings were consistent with benign SFT. The tumor was composed of fibroblastic appearing cells, showing a characteristic "pat-

ternless" growth pattern with small fascicular areas. The vessels adopted a hemangiopericytoma-like growth pattern consisting of elongated and dilated vessels with thin walls (Figure 3). There were few mitotic cells (2×10 HPF) and no nuclear atypia observed in 10 high power fields, nor any tumor necrosis. Immunohistochemistry showed that the neoplastic cells were strongly and diffusely posi-

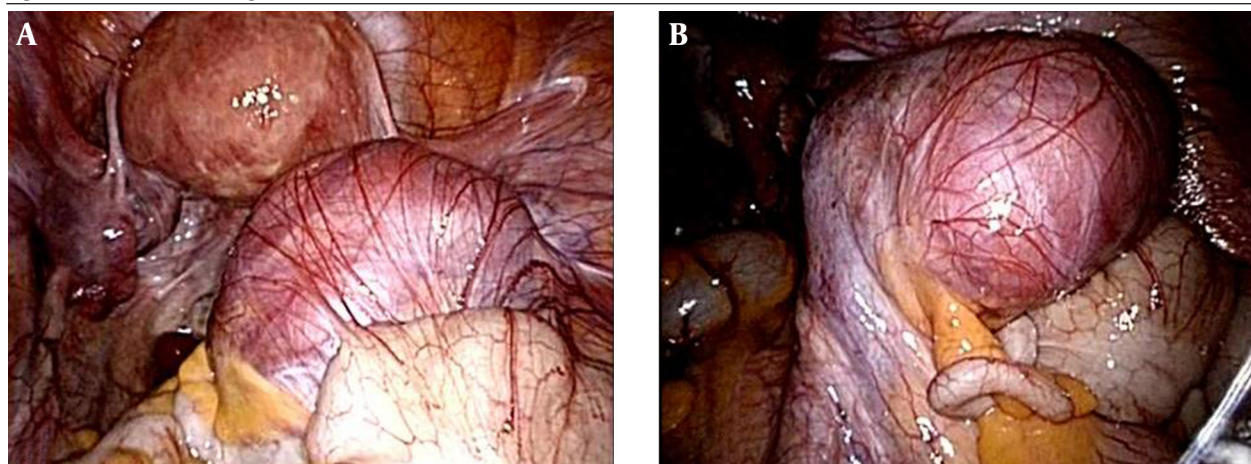
tive for CD34 and negative for c-kit (CD117) (Figure 4), and they were also positive for BCL2 and actin 1A4 and negative for desmin and CD99; Mib1 was $< 5\%$. The patient underwent radical excision of the neoplasm and has since not been treated with any further therapies. She is still alive, after four years, without any clinical-radiological evidence of recurrence.

Figure 1. Contrast-Enhanced CT-Scan of the Abdomen and Pelvis, Showing a Well-Defined Heterogeneous Enhancing Mass Strictly Adherent to the Uterus



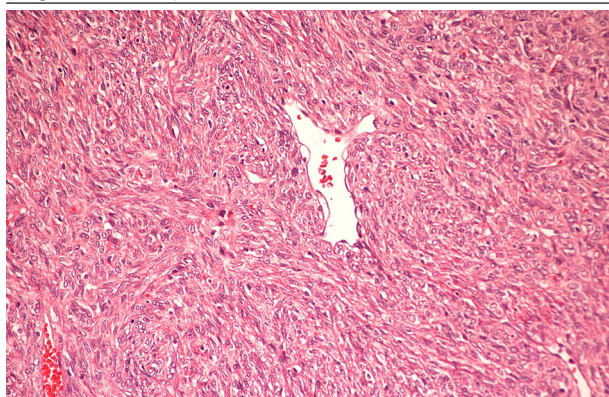
A: axial section, B: coronal section, C: sagittal section

Figure 2. Intrabdominal Image of the Mass



A: showing no connection with pelvic organs, B: and its location between cecal appendix and right iliac vessels

Figure 3. Microscopic Appearance: by Hematoxylin and Eosin Stains (Magnification, $20 \times$)



It shows patternless proliferation of bland-looking spindle to oval epithelioid cells, admixed with thin collagen bands. In some areas, tumor cells were arranged around ectasic vessels in a hemangiopericytoma-like pattern.

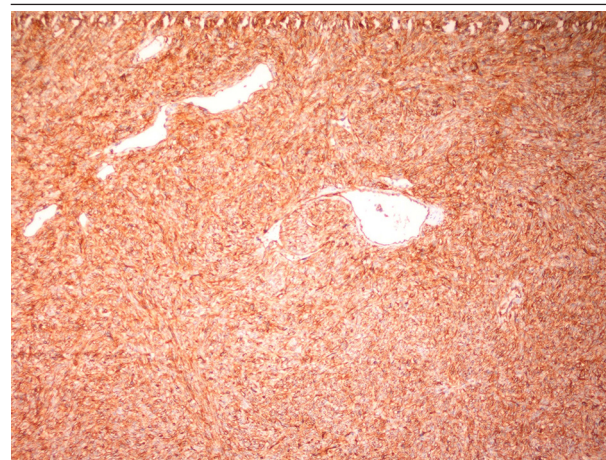


Figure 4. The Immunohistochemical Staining Revealed CD34 Positive Immunoreactivity

Table 1. Literature Reports About Retroperitoneal SFT^a

Author	Year	Country	Cases, No.	Age, y	Gender	Symptoms	Site	Biological Behaviour	Ref
Nielsen	1997	USA	1	Na	Na	Na	Retroperitoneum	Malignant	(13)
Morimitsu	2000	Japan	1	Na	Na	Na	Na	Na	(14)
Nakatani	2002	Japan	1	56	F	Palpable mass	Under left kidney	Malignant	(15)
Kume	2002	Japan	1	Na	Na	Na	Around SMA	Na	(16)
Nagasako	2004	Japan	1	81	M	Palpable mass	pelvis	Benign	(17)
Kunieda	2004	Japan	1	53	M	Right flank pain	Between right kidney and liver	Benign	(18)
Cristi	2005	Italy	1	28	F	Abdominal pain	Pelvis	Benign	(19)
Kakihara	2007	Japan	1	39	F	Fatigue	Left renal space	Malignant	(20)
Yamashita	2007	Japan	1	69	F	Palpable mass	Right renal space	Benign	(21)
Wat	2008	Canada	1	63	F	Abdominal pain	Pelvis	Benign	(22)
Ito	2008	Japan	1	48	F		Under left kidney	Malignant	(23)
Takizawa	2008	Japan	4	42-74	1 F/3 M	2 Asymptomatic, 1 Abdominal mass, 1 Dysuria	3 Perirenal, 1 Presacral	2 Malignant, 2 Benign	(6)
Chen ^b	2008	China	1	Na	Na	Na	Na	Na	(24)
Saint-Blancard ^c	2009	France	1	34	M	Hypoglycemia	Retroperitoneum	Na	(25)
Wagner	2009	Germany	1	28	M	Hypoglycemia	Pelvis	Malignant	(26)
Anchi ^d	2009	Japan	1	54	F	Incidental	Left adrenal space	Benign	(27)
Kawashima ^d	2009	Japan	1	56	M	Hypoglycemia	Retroperitoneum	Malignant	(28)
Nukui ^d	2009	Japan	1	63	M	Incidental	Pelvis	Benign	(29)
Mosquera	2009	USA	1	Na	Na	Na	Na	Na	(30)
Tadokoro ^d	2010	Japan	1	96	F	Right flank pain	Between right kidney and vena cava	Benign	(31)
Yamakawa	2010	Japan	1	65	M	Hypoglycemia	Right retroperitoneum	Malignant	(32)
Herrera ^e	2010	Spain	2	71, Na	F, Na	Abdominal pain, Na	Na, Na	Na, Na	(33)
Bae	2011	Korea	1	59	M	Incidental	Between stomach and left hepatic lobe	Malignant	(34)
Present case	2011	Italy	1	29	F	incidental	Pelvis	Benign	
Total			28						

^a Abbreviation: Na: Not available.

^b Article in Chinese.

^c Article in French.

^d Article in Japanese.

^e Article in Spanish.

3. Discussion

A SFT is a relatively unusual neoplasm first described as a distinctive tumor arising from pleura. Extra thoracic SFT by now have been reported at almost all anatomical location, but reports of tumor in the retroperitoneum is quite rare with only 28 cases, including the present, reported in literature (Table 1) The most part of retroperitoneal SFT are reported by eastern authors especially Japanese. They were aged from 28 to 96 years reflecting a wide range of onset. The clinical presentation of SFTs varies greatly, presenting with symptoms related to the tumor site and size, or with systemic symptoms which include hypoglycemia, arthralgia, osteoarthropathy and clubbing (4). Briselli et al. have reported that approxi-

mately 4% of solitary fibrous tumors that develop in the pleura are associated with hypoglycaemia (5), while the incidence of hypoglycemia in the case of retroperitoneal solitary fibrous tumors has been reported to be approximately 11.5% (6). Our review shows an incidence of hypoglycemia of 14.2%. A correct pre-operative diagnosis of SFTs is difficult because clinical manifestations, physical examination findings, and imaging results are not distinctive. On computed tomography findings, SFTs appear as well-defined, solid tumors containing cystic portions. In the present case, she had no symptom related to tumor. But, the mass was founded incidentally on gynaecological control examination. An abdominal CT revealed a well-defined heterogeneous enhancing mass. Only histopathologic evidence can allow a correct diagnosis of

SFTs. An intact layer of mesothelium overlying the tumor characterizes SFTs histopathologically. The pathological features of SFT were “patentless”, characterized by haphazard, storiform arrangement of short spindle or ovoid cells, and “hemangiopericytoma-like appearance” with prominent vascularity by thin-walled vessels. Immunohistochemically, expression of CD34 have been reported (7) as well as positivity for bcl-2 (8). Our patient had both patterns and coexpresses CD34 and bcl-2. Hasegawa et al. described that the numbers of Mib1 positive nuclei were low in all histologically benign cases, ranging from 0.5 to 10% (mean 1.8%) (8). For this reason we defined our case as benign SFT. For the differential diagnosis of retroperitoneal SFT, the following malignant or benign spindle cell tumors should be considered: synovial sarcoma, nerve sheath tumors, hemangiopericytoma, leiomyoma, desmoid tumor, cellular angiofibroma, and gastrointestinal stromal tumor (9). Despite their unpredictable behavior, 10-15% of SFTs are aggressive. Takizawa et al. reported malignancy in 14.6% of patients with retroperitoneal solitary fibrous tumors (6). Local recurrence or onset of metastasis depends mainly on the following prognostic parameters: histological features (nuclear atypia, increased cellularity, necrosis, and more than 4 mitoses per 10 HPF), localization (approximately 10 to 15% of SFTs located outside the thoracic cavity are recurrent or metastatic), dimensions (> 10 cm) and resectability that is the most important factor (3, 10). Lesions located in the mediastinum, abdomen, pelvis or retroperitoneum tend to behave more aggressively than those in the limbs. Even the tumor appearance is a prognostic indicator, so polypoid, peduncolated tumours behave in a benign fashion regardless of their histological appearance, and the malignant cases are often poorly circumscribed, infiltrative masses (11). Because of the unpredictable behavior that SFTs display and the lack of consistent data in literature, surgery is considered the treatment of choice whenever possible, and only when radical resection is not feasible radiation and chemotherapy are applied. In literature surgical resection of these tumors is reported by open approach, often due to the large size of the mass. Only one case of bleeding SFT of the great omentum was approached and resected by laparoscopy (12). Our case is the first case of SFT of the retroperitoneum removed laproscopically.

In conclusion we report a case of retroperitoneal SFT to emphasize the importance of bearing this entity in mind, to show that the characteristic microscopic appearance of this unusual tumour and immunohistochemistry are useful for differentiating it from other spindle cell neoplasms, and to underline the importance of a minimally invasive approach. Laparoscopy, offering all advantages of a minimally invasive technique, is useful to remove SFT located in the pelvis, great omentum, gastric or small bowel wall, in the mesentere, and at list, as in the present case, even in peduncolated retroperitoneal tumor with intrabdominal growing.

Authors' Contributions

Study concept and design: Maria Sofia, Domenico Russello. Acquisition of data: Saverio Latteri, Valentina Randazzo. Analysis and interpretation of data: Antonio Galia, Giuseppe Scibilia. Drafting of the manuscript: Maria Sofia. Critical revision of the manuscript for important intellectual content: Maria Sofia, Gaetano La Greca. Statistical analysis: Maria Sofia, Giuseppe Scibilia. Administrative, technical, and material support: Saverio Latteri, Valentina Randazzo. Study supervision: Gaetano La Greca, Domenico Russello.

References

1. Klemperer P, Rabin CB. Primary neoplasms of the pleura. A report of the five cases. *Arch Pathol.* 1931;**11**:385-412.
2. Hanau CA, Miettinen M. Solitary fibrous tumor: histological and immunohistochemical spectrum of benign and malignant variants presenting at different sites. *Hum Pathol.* 1995;**26**(4):440-9.
3. Bruzzzone A, Varaldo M, Ferrarazzo C, Tunesi G, Mencoboni M. Solitary fibrous tumor. *Rare Tumors.* 2010;**2**(4).
4. Lowbeer L. Hypoglycemia-producing extrapancreatic neoplasms. A review. *Am J Clin Pathol.* 1961;**35**:233-43.
5. 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**(11):2678-89.
6. Takizawa I, Saito T, Kitamura Y, Arai K, Kawaguchi M, Takahashi K, et al. Primary solitary fibrous tumor (SFT) in the retroperitoneum. *Urol Oncol.* 2008;**26**(3):254-9.
7. 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**(8):814-20.
8. Hasegawa T, Matsuno Y, Shimoda T, Hirohashi S, Hirose T, Sano T. Frequent expression of bcl-2 protein in solitary fibrous tumors. *Jpn J Clin Oncol.* 1998;**28**(2):86-91.
9. Suster S, Nascimento AG, Miettinen M, Sickel JZ, Moran CA. Solitary fibrous tumors of soft tissue. A clinicopathologic and immunohistochemical study of 12 cases. *Am J Surg Pathol.* 1995;**19**(11):1257-66.
10. 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**(8):640-58.
11. Moran CA, Suster S, Koss MN. The spectrum of histologic growth patterns in benign and malignant fibrous tumors of the pleura. *Semin Diagn Pathol.* 1992;**9**(2):169-80.
12. Patriti A, Rondelli F, Gulla N, Donini A. Laparoscopic treatment of a solitary fibrous tumor of the greater omentum presenting as spontaneous haemoperitoneum. *Ann Ital Chir.* 2006;**77**(4):351-3.
13. 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**(10):1028-37.
14. Morimitsu Y, Nakajima M, Hisaoka M, Hashimoto H. Extrapleural solitary fibrous tumor: clinicopathologic study of 17 cases and molecular analysis of the p53 pathway. *APMIS.* 2000;**108**(9):617-25.
15. Nakatani T, Tamada S, Iwai Y, Tanimoto Y. Solitary fibrous tumor in the retroperitoneum: a case with infiltrative growth. *Hinyokika Kyo.* 2002;**48**(10):637-41.
16. Kume M, Komori K, Inoguchi H, Shoji T, Furuyama T, Sakamoto A, et al. Solitary fibrous tumor in the retroperitoneal space: report of a case. *Surg Today.* 2002;**32**(12):1099-101.
17. Nagasako Y, Misawa K, Kohashi S, Sano H. Solitary fibrous tumor in the retroperitoneum. *J Am Coll Surg.* 2004;**198**(2):322-3.
18. Kunieda K, Tanaka Y, Nagao N, Yamaguchi K, Sano J, Osada S, et al. Large solitary fibrous tumor of the retroperitoneum: report of a case. *Surg Today.* 2004;**34**(1):90-3.
19. Cristi E, Perrone G, Battista C, Benedetti-Panici P, Rabitti C. A rare case of solitary fibrous tumour of the pre-sacral space:

- morphological and immunohistochemical features. *In Vivo*. 2005;**19**(4):777-80.
20. Kakihara D, Yoshimitsu K, Eto M, Matsuura S, Honda H. MRI of retroperitoneal solitary fibrous tumor in the suprarenal region. *AJR Am J Roentgenol*. 2007;**188**(6):W512-4.
 21. Yamashita S, Tochigi T, Kawamura S, Aoki H, Tateno H, Kuwahara M. Case of retroperitoneal solitary fibrous tumor. *Hinyokika Kyo*. 2007;**53**(7):477-80.
 22. Wat SY, Sur M, Dhamanaskar K. Solitary fibrous tumor (SFT) of the pelvis. *Clin Imaging*. 2008;**32**(2):152-6.
 23. Ito H, Fukuda M, Imamura Y, Fuse H. A malignant solitary fibrous tumor in the retroperitoneum. *Int J Clin Oncol*. 2008;**13**(2):173-5.
 24. Chen YY. [Retroperitoneal solitary fibrous tumour: report of a case]. *Zhonghua Bing Li Xue Za Zhi*. 2008;**37**(5):355-6.
 25. Saint-Blancard P, Jancovici R. [Solitary fibrous tumor of the retroperitoneum]. *Rev Med Interne*. 2009;**30**(2):181-5.
 26. Wagner S, Greco F, Hamza A, Hoda RM, Holzhausen HJ, Fornara P. Retroperitoneal malignant solitary fibrous tumor of the small pelvis causing recurrent hypoglycemia by secretion of insulin-like growth factor 2. *Eur Urol*. 2009;**55**(3):739-42.
 27. Anchi T, Tamura K, Inoue K, Fukata S, Nishikawa H, Moriki T, et al. [A case of retroperitoneal solitary fibrous tumor]. *Hinyokika Kyo*. 2009;**55**(7):401-3.
 28. Kawashima A, Ujike T, Nin M, Nishimura K, Miyoshi S, Kawano K, et al. [A case of retroperitoneal solitary fibrous tumor associated with hypoglycemia]. *Hinyokika Kyo*. 2009;**55**(7):395-9.
 29. Nukui A, Ochi M, Suzuki K, Yuzawa M, Morita T. [Retroperitoneal solitary fibrous tumor: a case report]. *Hinyokika Kyo*. 2009;**55**(8):503-7.
 30. Mosquera JM, Fletcher CD. Expanding the spectrum of malignant progression in solitary fibrous tumors: a study of 8 cases with a discrete anaplastic component—is this dedifferentiated SFT? *Am J Surg Pathol*. 2009;**33**(9):1314-21.
 31. Tadokoro A, Kawaguchi M, Koike H. [A case of retroperitoneal solitary fibrous tumor]. *Hinyokika Kyo*. 2010;**56**(6):311-4.
 32. Yamakawa-Yokota F, Ozaki N, Okajima A, Nishio H, Nagasaka T, Oiso Y. Retroperitoneal solitary fibrous tumor-induced hypoglycemia associated with high molecular weight insulin-like growth factor II. *Clin Med Res*. 2010;**8**(3-4):159-62.
 33. Carnerero Herrera V, Dominguez-Perez A, Gonzalez-Martin R, Nacarino-Mejias V, Alcazar Iribarren-Marin M. [Solitary fibrous tumor of the abdomen. A rare neoplasm of vascular origin]. *Gastroenterol Hepatol*. 2010;**33**(8):578-81.
 34. Bae JM, Kim SW, Kim SW, Song SK. Malignant solitary fibrous tumor of retroperitoneum mimicking gastric submucosal tumor. *Korean J Gastroenterol*. 2011;**57**(1):47-50.