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Hussein S.[1, 2], Parkhomenko L.[1], Maslova M.[2], Kondratieva K.[2]
[1] Belarusian State Medical University, Minsk, Belarus
[2] N.N. Alexandrov National Cancer Centre, Minsk, Belarus

Solitary Fibrous Tumor of the Abdominal Wall: Potential for Indolent Malignancy, A Rare Case Report, Ultrastructural Studies and Reviewing Relevant Publications

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Contacts: lar.bar.parkhomenko@gmail.com, husseinsaad.hpb.onco@mail.ru

Abstract

Solitary fibrous tumor (SFT) is a rare, mesenchymal-originating fibroblastic tumor with a hypervascular constitution, predominantly affecting serous membranes and soft tissue, most often arising in the pleura and peritoneum.
These spindle cell tumors develop from dendritic cells that express the haematopoietic progenitor cell marker CD34. SFTs are classified as fibroblast or myofibroblast origin, falling under the intermediate class or borderline behaviour. Solitary fibrous tumors originating from the abdominal wall are uncommon, with few case reports in the literature. These slow-growing tumors affect both sexes, and are more prevalent in middle-aged individuals. The progression of the disease is uncertain. There effectiveness of chemotherapy in SFT is limited. Long-term observation and monitoring are critical to preventing disease relapse. Individuals with high-risk features may require adjuvant therapy.
Keywords: solitary fibrous tumor, chemotherapy, mesenchymal tumor

Хуссейн С.Х.[1, 2], Пархоменко Л.Б.[1], Маслова М.С.[2], Кондратьева К.А.[2]
[1] Белорусский государственный медицинский университет, Минск, Беларусь
[2] Республиканский научно-практический центр онкологии и медицинской радиологии имени Н.Н. Александрова, Минск, Беларусь

Солитарная фиброзная опухоль брюшной стенки: потенциальная возможность индолентной злокачественности, клинический случай, ультраструктурные исследования и обзор литературы

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Контакты: lar.bar.parkhomenko@gmail.com, husseinsaad.hpb.onco@mail.ru

Резюме

Солитарная фиброзная опухоль – редкая фибробластическая опухоль мезенхимального происхождения с гиперваскулярным строением, преимущественно поражающая серозные оболочки и мягкие ткани, чаще всего возникающая в плевре и брюшине. Эти веретенообразные опухоли развиваются из дендритных клеток, которые экспрессируют маркер гемопоэтических клеток-предшественников CD34. Солитарные фиброзные опухоли классифицируются как фибробластные или миофибробластные, попадая в промежуточный класс или пограничное поведение. Солитарные фиброзные опухоли, возникающие в брюшной стенке, встречаются редко, в литературе описано мало таких случаев. Эти медленно растущие опухоли поражают оба пола, чаще встречаются у людей среднего возраста. Прогрессирование заболевания носит неопределенный характер. Эффективность химиотерапии при солитарных фиброзных опухолях ограничена. Долгосрочное наблюдение и мониторинг имеют решающее значение для предотвращения и раннего выявления рецидива заболевания. Лицам с признаками высокого риска прогрессирования может потребоваться адъювантная терапия.
Ключевые слова: солитарная фиброзная опухоль, химиотерапия, мезенхимальная опухоль

■ INTRODUCTION

Solitary fibrous tumor (SFT) is a relatively rare mesenchymal origin fibroblastic tumor with a hypervascular constitution, which most commonly originates in the pleura [1]. These tumors are a rare spindle cell tumor that develops from dendritic cells that express

the haematopoietic progenitor cell marker CD34 [2]. In accordance with the updated version of the World Health Organization's (2020) classification of soft tissue tumors, (SFTs) are categorized as tumors of fibroblast or myofibroblast origin, falling under the intermediate class or borderline behaviour (rare metastasis) [3]. Roughly 10–20% of these tumors have aggressive behaviour [4]. The tumor was initially described by Klemperer and Rabin in 1931 [5, 6], and it has been documented in several extrabdominal and extrapleural sites, including the brain and spinal cord [7]. This slow-growing tumor affects both sexes equitably, but is more prevalent in middle-aged individuals. The progression of the disease is uncertain. Consequently, most physicians have a lack of expertise in SFT, and preoperative diagnosis can frequently be challenging.

The major goal of this study is to deliberate the clinical features, histopathological and radiologic findings of SFT emerging from the abdominal wall. A more thorough comprehension of these traits will increase diagnostic accuracy.

■ CASE REPORT

A 70 year-old patient with mental issues was admitted to the hospital with abdominal distension. Due to the mental issues the patient can't admit any further symptoms, including, fever, cough, nausea, vomiting, gastric discomfort, diarrhea, anorexia, fatigue or bloody stool. There was no history of cancer or surgery.

Clinical observations

During the physical examination, the abdomen was soft with bulging mass extending from left upper and middle sides of the abdomen. The mas measured approximately 18×16 cm.

Diagnostic Assessment

Tumor markers such as CEA, CA19-9, PSA total and CA72-4, were within normal range. All other laboratory tests, including full blood count, biochemistry, coagulation profile were within normal limits. The abdominal CT scan revealed a large solid-cystic tumor of the abdominal cavity measuring approximately 21.2×17.2 cm (Fig. 1).

Tumor compressing other adjacent organs and tissues located on the middle and left between the splenic flexure of the colon, left kidney, stomach. Impaired perfusion of the left kidney. Extensive venous network around the tumor and along the anterior abdominal wall. We considered the potential of a mesenchymal tumor There was no evidence of regional lymphadenopathy or distant metastases. Preoperatively, we contemplated that this tumor could be surgically resected.

■ HISTOPATHOLOGY AND MICROSCOPIC EXAMINATION

The surgical specimen was preserved in 4% buffered formaldehyde and then processed using standard techniques. Macroscopically, the lesion was a nodular, well-circumscribed, encapsulated mass and was measured as 21×19×12 cm. The capsule was focally damaged with a soldered area of fatty tissue 17×6.5×0.2 cm and a coagulated area of soft tissues. Resection margins were marked by ink. In serial sections, the tumor is a greyish-white color of a softly elastic consistency with areas of disintegration and small cystic cavities filled with serous content. In microscopic examination, a soft tissue tumor with extensive necrosis and the presence of a thin connective tissue capsule. Tumor with different cell

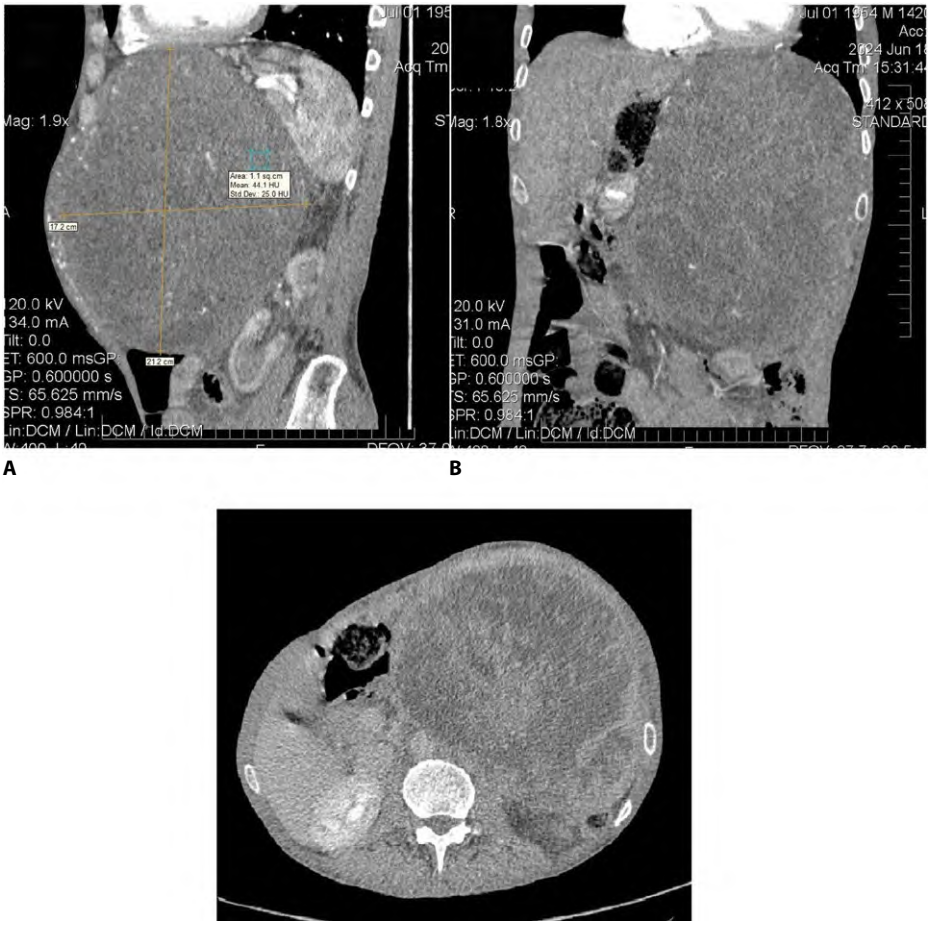


Fig. 1. Preoperative enhanced CT scan of the abdomen: A – sagittal section of the tumor; B – coronal section of the tumor; C – axial section of the tumor

density with hypocellular and hypercellular areas (Fig. 2), consisting of spindle-shaped and rounded cells, forming solid fields and short multidirectional bundles.

There were areas with fibromyxoid stroma and thickened collagen fibres. The branching blood vessels of various sizes imparted a "staghorn"-like appearance (Fig. 3).

Tumor cells with eosinophilic and focally light cytoplasm, ovoid and round nuclei, with a small nucleolus in some cells, without pronounced cytological atypia (Fig. 4).

Mitotic rate was 3 mitoses per 1 mm^2. In the marked resection margin, there was a focus of painted tumor cells, which refers to a positive resection margin (R1). In immunohistochemical analysis, it was revealed that the tumor was positive for $STAT6^+$, $CD34^+$, and $CD99^+$ with focally weak intensity (Fig. 5), and $TLE1^+$ weak intensity expression in single cells.

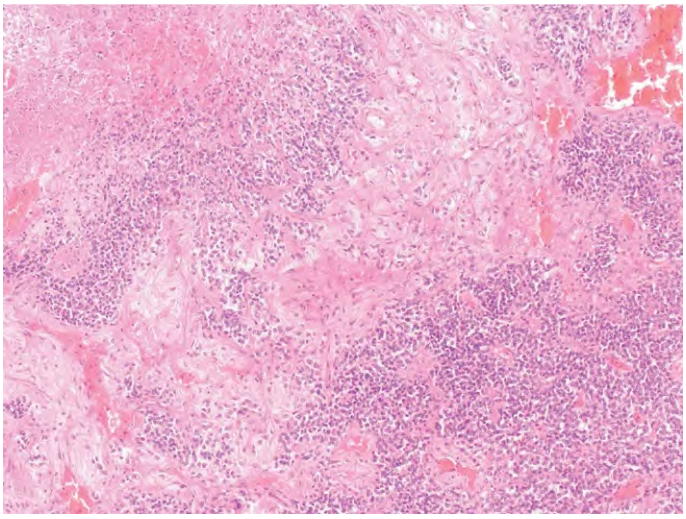


Fig. 2. Low power view of H&E staining (×100) shows hypercellular and hypocellular areas with focus of necrosis

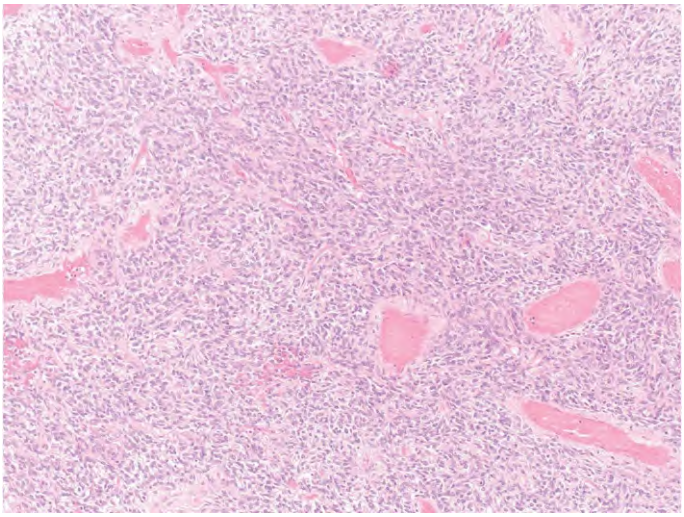


Fig. 3. Low power view with branching staghorn-like blood vessels of various size. H&E staining (×100)

It was negative for PanCK, Desmin, S100, and SMA. Proliferative activity index (Ki67) – 1–3%. The diagnosis of solitary fibrous tumor with positive resection margin was histologically established. The metastatic risk was assessed as high 7 points according to the modified four-variable risk models (Table 1).

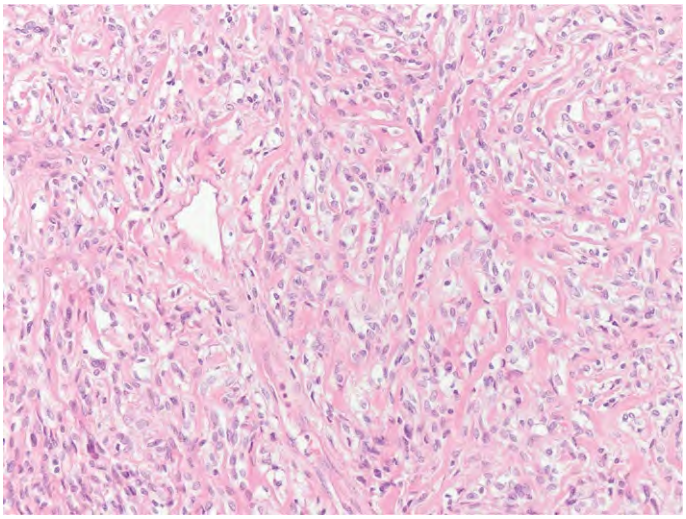
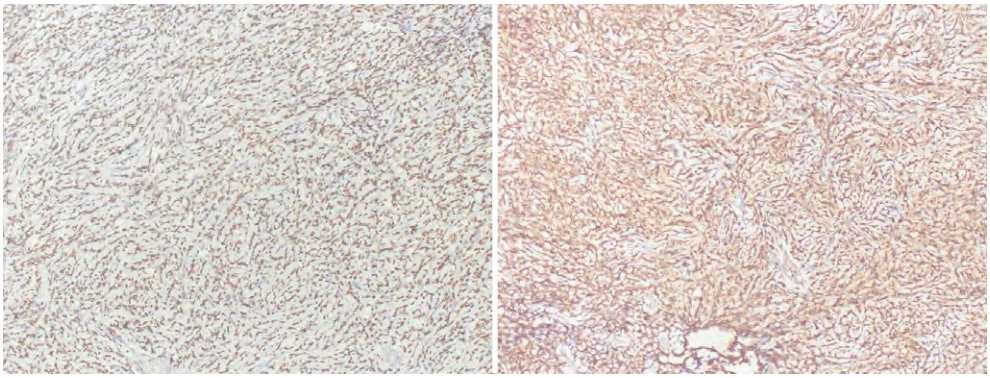


Fig. 4. High power view shows cells with eosinophilic and focally light cytoplasm, ovoid and round nuclei, without pronounced cytological atypia. H&E staining (×200)



A **B**

Fig. 5. A – STAT6 immunohistochemical stain with nuclear staining; B – Diffuse membranous CD34 positivity

■ DISCUSSION

Solitary fibrous tumors are uncommon spindle cell neoplasms that often occur in the thorax, although up to 50–70% can occur beyond. It accounts for less than 2% of all soft tissue tumors [6, 8]. These tumors grow in a variable anatomical locations like bones, brain, oral cavity, head and neck regions. A 30% from the thoracic cavity, primarily from pleura [9, 10], and also about 30% from the peritoneal cavity, mainly from retroperitoneum and pelvic soft tissues [11]. SFTs originating from the abdominal wall are exceedingly uncommon. There were not many case reports of SFTs arising from the abdomen wall in

Table 1
Three-variable and modified four-variable risk models for the prediction of metastatic risk in solitary fibrous tumors

Risk factor	Cut-off	Points assigned	
		3-variable model	4-variable model
Patient age in years	<55	0	0
	≥55	1	1
Mitoses/mm^2 (mitoses per 10 HPFs)	0 (0)	0	0
	0.5–1.5 (1–3)	1	1
	≥2 (≥4)	2	2
Tumor size in cm	0–4.9	0	0
	5–9.9	1	1
	10–14.9	2	2
	≥15	3	3
Tumor necrosis	<10%	n/a	0
	≥10%	n/a	1
Risk	Low	0–2 points	0–3 points
	Intermediate	3–4 points	4–5 points
	High	5–6 points	6–7 points

Notes: HPF- high-power field; n/a – not applicable.

the currently available literature. Described cases of SFTs that originate from the abdominal wall [12–18], all of which received surgical resection with no tumor recurrence at the stipulated follow-up intervals, except patient reported by Bi et al. [16], a complete surgical resection was performed on a solitary fibrous tumor of the abdominal wall 3 years ago. and the patient was diagnosed with a recurrent solitary fibrous tumor that metastasized to the lung, and manifested with a unilateral pleural effusion. She was not a candidate for systemic chemotherapy and died one year later of severe respiratory failure. While reviewing the literature, SFTs are historically considered indolent tumors that are rare to metastasize. Unfortunately, the behaviour is unpredictable and encompasses a wide range of biologic behaviour. Although most SFTs are benign following surgical excision, some may recur locally or spread to distant sites. Therefore, long-term observation is required. In a study of 24 patients with extrathoracic SFTs, two experienced local recurrences at 6 months and 5 years, while one developed pulmonary metastases after 7 years [19]. Furthermore, there is no clear association between morphology and clinical outcomes. Most histologically benign SFTs are non-recurring and non-metastasising lesions. while tumors in the mediastinum, abdomen, pelvis, and retroperitoneum exhibit more aggressive behaviour than pleuropulmonary SFTs [20]. In 2002, Gold et al. published the first large series analysing clinicopathologic correlates of tumor behaviour in 79 patients with SFTs. The study found a low rate of local recurrence and metastasis after surgical treatment, with a small but statistically significant increase in risk for extrathoracic SFTs. There was no difference in metastasis-free survival for thoracic and extrathoracic SFTs [8]. In 2013, van Houdt et al. Published a retrospective study of 81 patients treated for SFTs from 1995 to 2012 found that positive resection margins were associated with local recurrence, while tumor size >10 cm and high mitosis rate were associated with higher

metastasis rates and a trend towards poor overall survival [21]. Demicco et al. published a risk assessment model for SFTs of all locations except meninges in 2012 [22], based on a clinicopathological examination of 110 cases. The model used patient age at presentation, tumor size, and mitotic index to identify patients at high risk for poor outcomes (Table 1). Patients with SFTs in the high-risk group had a 5- and 10-year disease-specific survival rate of 60 and 0%, respectively, whereas those in the moderate-risk group have a survival rate of 93 and 93% [22]. However, the model has not been verified in large-scale investigations or clinical settings. Long-term follow-up is crucial for patients with SFTs, especially those with high-risk characteristics, due to the disease's slow progression and potential for late recurrence.

Our patient underwent a thorough preoperative evaluation, with no contraindications to surgery. A large abdominal tumor was completely resected with open total resection without intraoperative complications. Due to the positive resection margin (R1),

Table 2
Published cases of SFTs that originated from the abdominal wall

№	Author	Age	Sex	Size (cm)	SFT-location	Interven tion	CD 34 expression	Malignancy	Follow up intervals (months)	Recurrence
1	Huang et al. [12] (2002)	50	F	4	abdominal wall	Surgical Resection	Positive	–	14 months	No recurrence
2	Huang et al. [12] (2002)	38	F	7.5	abdominal wall	Surgical Resection	Positive	–	12 months	No recurrence
3	Ouazzani et al. [14] (2008)	66	M	16	abdominal wall	Surgical Resection	Positive	+	6 months	No recurrence
4	Migita et al. [13] (2009)	75	M	12	abdominal wall	Surgical Resection	Positive	–	10 months	No recurrence
5	Mosquera et al. [15] (2009)	50	F	6.4	abdominal wall	Surgical Resection	Positive	–	12 months	No recurrence
6	Bi et al. [16] (2017)	79	F	15	abdominal wall	Surgical Resection	Positive	+/ with MTS	48 months	Died
7	Ruiz-Juliá et al. [17]	45	F	6	abdominal wall	Surgical Resection	Positive	+	NA	NA
8	Testa et al. [18] (2019)	64	M	4.8	abdominal wall	Surgical Resection	Positive	–	6 months	No recurrence
9	Presented case	70	M	21	abdominal wall	Surgical Resection	Positive	+	6 months	No recurrence

an adjuvant single-agent chemotherapy dacarbazine was considered. There are currently no prospective trials on the effectiveness of chemotherapy in SFT. A poor response rate (RR) following conventional anthracycline-based chemotherapy, ranging from 0% to 20%, was seen in a small number of retrospective datasets [23, 24]. There are very few documented responses in individuals receiving anthracycline plus dacarbazine +/– ifosfamide [25]. In 2013 S. Stacchiotti et al. published a retrospective study to investigate triazines' (Dacarbazine, temozolomide) and antiangiogenics (Sunitinib and bevacizumab) beneficial effects for solitary fibrous tumors (SFT). The combination of bevacizumab and temozolomide did not have an improved therapeutic effect than temozolomide alone. These findings support the cytotoxic impact of temozolomide and dacarbazine, as opposed to the cytostatic effects of antiangiogenics [26]. As predicted, temozolomide and dacarbazine were equivalent in terms of tumor reduction, treatment effect duration, and pathologic response. On other hand Stacchiotti et al. Also compered the effectivness of dacarbazine – sunitinib in tumor shrinkage, the results showed that dacarbazine may be more effective in achieving tumor size reduction than sunitinib, at least in more aggressive SFT types. Most patients in this study had sunitinib prior to dacarbazine, with responses to dacarbazine found in those who progressed on sunitinib and vice versa [26]. Results demonstrate the possibility of dacarbazine and temozolomide can be used as a single agent in SFT. In 2023 S. Stacchiotti et al. Published a phase II randomised clinical trial (STRADA) to investigate the efficacy of trabectedin (Ta) vs adriamycin-dacarbazine (A–D) in patients with advanced SFT, the results indicated that no responses were found to either (Ta) or (A–D) among all SFT subtypes; hence, the study was closed to the enrolment for T-SFT and M-SFT following the interim analysis. It is important to highlight that both Ta and A–D stabilised the diseases in around 40% of previously progressing patients, and over 30% of Ta-treated patients were free of progression after one year. However, no conclusions can be obtained about the action of Ta/A–D in this SFT subtype [27]. More research must be conducted to determine the therapeutic value of high-risk traits in devising post-treatment monitoring for extrapleural SFTs.

■ CONCLUSION

SFTs are uncommon mesenchymal tumors that originates primarily in the pleura but have been reported in other locations. Although surgical resection is the primary treatment, long-term monitoring and surveillance are important to avoid disease recurrence. Adjuvant treatment may also be required for individuals with high-risk characteristics. More research is needed to validate risk categorisation models and establish post-treatment monitoring protocols for extrathoracic SFTs.

Follow up and outcomes

Abdominal CT scans were conducted 6 months after resection, and there were no evidence of recurrence or metastasis.

■ REFERENCES

1. Yuza K., Sakata J., Nagaro H., et al. A giant pelvic solitary fibrous tumor with Doege-Potter syndrome successfully treated with transcatheter arterial embolization followed by surgical resection: a case report. *Surgical case reports*. 2020;6(1):299. Available at: https://doi.org/10.1186/s40792-020-01076-5

2. Fang H., Wei Z., Lai J., et al. Multidisciplinary treatment of giant presacral solitary fibrous tumor: a case report and literature review. *The Journal of international medical research*. 2022;50(11):3000605221135458. Available at: https://doi.org/10.1177/03000605221135458
3. Sbaraglia M., Bellan E., Dei Tos A.P. The 2020 WHO Classification of Soft Tissue Tumors: news and perspectives. *Pathologica.* 2021 Apr;113(2):70–84. doi: 10.32074/1591-951X-213
4. Yanik F., Karamustafaoglu Y.A., Yoruk Y. Surgical outcomes and clinical courses of solitary fibrous tumors of pleura. *Nigerian journal of clinical practice*. 2019;22(10):1412–1416. Available at: https://doi.org/10.4103/njcp.njcp_213_18
5. Klemperer P., Coleman B.R. Primary neoplasms of the pleura. A report of five cases. *American journal of industrial medicine*. 1992;22(1):1–31. Available at: https://doi.org/10.1002/ajim.4700220103
6. Wang S., Guan J., Liu M., et al. Solitary fibrous tumor in the subcutaneous soft tissues of the left abdominal wall: A case report. *Clinical case reports*. 2019;7(7):1370–1373. Available at: https://doi.org/10.1002/ccr3.2217
7. Takenouchi T., Pannullo S.C., Stieg P.E., Lavi E. Solitary fibrous tumor with multiple intracranial and spinal lesions: case report. *Neurosurgery*. 2011 ;68(4):E1148–E1151. Available at: https://doi.org/10.1227/NEU.0b013e31820a1573
8. Gold J.S., Antonescu C.R., Hajdu C., et al. Clinicopathologic correlates of solitary fibrous tumors. *Cancer.* 2002;94(4):1057–1068.
9. Robinson L.A. Solitary fibrous tumor of the pleura. *Cancer control: journal of the Moffitt Cancer Center*. 2006;13(4):264–269. Available at: https://doi.org/10.1177/107327480601300403
10. Cardillo G., Lococo F., Carleo F., Martelli M. Solitary fibrous tumors of the pleura. *Current opinion in pulmonary medicine*. 2012;18(4):339–346. Available at: https://doi.org/10.1097/MCP.0b013e328352f696.
11. Zhou Y., Chu X., Yi Y., et al. Malignant solitary fibrous tumor in retroperitoneum: A case report and literature review. *Medicine*. 2017;96(11):e6373. Available at: https://doi.org/10.1097/MD.0000000000006373
12. Huang H.Y., Sung M.T., Eng H.L., et al. Solitary fibrous tumor of the abdominal wall: a report of two cases immunohistochemical, flow cytometric, and ultrastructural studies and literature review. *APMIS: acta pathologica, microbiologica, et immunologica Scandinavica*. 2002;110(3):253–262. Available at: https://doi.org/10.1034/j.1600-0463.2002.100308
13. Migita K., Watanabe A., Nakagawa K., et al. Solitary fibrous tumor of the abdominal wall. *International journal of clinical oncology*. 2009;14(6):555–559. Available at: https://doi.org/10.1007/s10147-009-0891-6
14. Ouazzani A., Delrée P., De Saint Aubain N., et al. Tumeur fibreuse solitaire maligne de la paroi abdominale chez un homme [Malignant solitary fibrous tumor of the abdominal wall in a man]. *Annales de chirurgie plastique et esthetique.* 2008;53(6):517–520. Available at: https://doi.org/10.1016/j.anplas.2008.03.003
15. Mosquera J.M., Fletcher C.D. Expanding the spectrum of malignant progression in solitary fibrous tumors: a study of 8 cases with a discrete anaplastic component – is this dedifferentiated SFT? *The American journal of surgical pathology*. 2009;33(9):1314–1321. Available at: https://doi.org/10.1097/pas.0b013e3181a6cd33
16. Bi X., Zhai J., Chun C.D. Solitary fibrous tumor of the abdominal wall re-surfacing as unilateral pleural effusion and mass: A case report and review of the literature. *Respiratory medicine case reports.* 2017;23:4–7. Available at: https://doi.org/10.1016/j.rmcr.2017.10.013
17. Ruiz-Julià M.L., Ruiz-Luque V., Ariza-Estepa M., et al. Solitary fibrous tumor of the abdominal wall: a new case with literature review. Tumor fibroso solitario de la pared abdominal: un nuevo caso con revisión de la literatura. *Cirugia y cirujanos.* 2024;92(6):823–828. Available at: https://doi.org/10.24875/CIRU.22000220
18. Testa D.C., Selvaggi F., Andreano T., et al. Solitary fibrous tumor of the anterior abdominal wall. A case report and review of the literature. *Annali italiani di chirurgia*. 2019;8:S2239253X19030068.
19. Hasegawa T., Matsuno Y., Shimoda T., et al. Extrathoracic solitary fibrous tumors: their histological variability and potentially aggressive behavior. *Human pathology*. 1999;30(12):1464–1473. Available at: https://doi.org/10.1016/s0046-8177(99)90169-7
20. Wilky B.A., Montgomery E.A., Guzzetta A.A., et al. Extrathoracic location and "borderline" histology are associated with recurrence of solitary fibrous tumors after surgical resection. *Annals of surgical oncology*. 2013;20(13):4080–4089. Available at: https://doi.org/10.1245/s10434-013-3241-x
21. Van Houdt W.J., Westerveld C.M., Vrijenhoek J.E., et al. Prognosis of solitary fibrous tumors: a multicenter study. *Annals of surgical oncology.* 2013;20(13):4090–4095. https://doi.org/10.1245/s10434-013-3242-9
22. Demicco E.G., Park M.S., Araujo D.M., et al. Solitary fibrous tumor: a clinicopathological study of 110 cases and proposed risk assessment model. *Modern pathology: an official journal of the United States and Canadian Academy of Pathology, Inc,* 2012;25(9):1298–1306. Available at: https://doi.org/10.1038/modpathol.2012.83
23. Chamberlain M.C., Glantz M.J. Sequential salvage chemotherapy for recurrent intracranial hemangiopericytoma. *Neurosurgery,* 2008;63(4):720–727. Available at: https://doi.org/10.1227/01.NEU.0000325494.69836.51
24. Galanis E., Buckner J.C., Scheithauer B.W., et al. Management of recurrent meningeal hemangiopericytoma. *Cancer*, 1998;82(10):1915–1920.
25. Ferigo N., Cottalorda J., Allard D., et al. Successful treatment via chemotherapy and surgical resection of a femoral hemangiopericytoma with pulmonary metastasis. *Journal of pediatric hematology/oncology.* 2006;28(4):237–240. Available at: https://doi.org/10.1097/01.mph.0000212903.61276.4b.
26. Stacchiotti S., Libertini M., Negri T., et al. Response to chemotherapy of solitary fibrous tumor: a retrospective study. *European journal of cancer (Oxford, England: 1990)*. 2013;49(10):2376–2383. Available at: https://doi.org/10.1016/j.ejca.2013.03.017
27. Stacchiotti S., Grignani G., Vincenzi B., et al. Trabectedin (T) versus adriamycin plus dacarbazine (A-DA) in advanced solitary fibrous tumor (SFT): Results from a phase II randomised clinical study (STRADA). *JCO.* 2023;41:11571–11571. DOI: 10.1200/JCO.2023.41.16_suppl.11571