Solitary fibrous tumor: a 10-year retrospective analysis with several rare cases

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To the Editor: Solitary fibrous tumors (SFTs) are mesenchymal neoplasms and were first described in pleura, then subsequently found in extrapleural organs, such as peritoneum, lung, parotid gland, paranasal sinuses, orbit, skin, and intracranial areas.[1] However, extrapleural SFTs are limited to isolated case reports. Because of the rarity of them, the related information is far from sufficient and thus, leading this tumor under fully recognized. According to the World Health Organization (WHO) classification, SFTs are pluripotent neoplasms that could be benign, intermediate, or malignant. All of above drive us to further understand the epidemiology, clinical characteristics, medical therapy, and prognosis of this disease.

In this study, we explored 262 SFT patients treated in Second Xiangya Hospital, Central South University, over a 10-year period. Furthermore, we presented several exceedingly rare cases, with the hope that this retrospective study could provide guidance for the understanding and preoperative diagnosis of SFTs.

This study was approved by the Institutional Medical Ethics Board at the Second Xiangya Hospital (No. 2020047) and has obtained exemption from informed consent, all the data were analyzed anonymously. The clinical data collected and analyzed included patient age at diagnosis, sex, primary tumor site, symptoms, symptom duration, radiological findings, treatment, and recurrence. Samples were embedded in paraffin for histological and immunohistochemical analyses. Two experienced pathologists re-reviewed and confirmed each case. Descriptive statistics were utilized to summarize the clinical data. Continuous variables are presented as the means and range, and categorical variables are presented as percentages.

Of 262 patients, 199 (76.0%) were inpatients, and 136 (51.9%) were female. The median age at diagnosis was 46.6 ± 15.4 years (ranging from 3 months to 76 years old); and 247 patients (94.3%) were over 18 years old. The primary tumor site was pleura in 57 (21.8%), lung in 53 (20.2%), head/neck in 36 (13.7%), extremity in 26 (9.9%), retroperitoneal in 20 (7.6%), abdomen in 14 (5.3%), pelvis in 13 (5.0%), central nervous system (CNS) in 13 (5.0%), mediastinum in 12 (4.6%), trunk/skin in 6 (2.3%), groin in 6 (2.3%), and others (breast and pericardium) in 6 (2.3%). The initial symptom duration ranged from 3 days to 240 months (Supplementary Table 1, http://links.lww.com/CM9/A363).

Patients with primary tumor located in pleura, mediastinum, and lung were usually found during routine physical examination without clinical symptoms. The primary sites of abdomen, extremity, trunk/skin, and groin shared the symptom of a palpable mass. Patients with tumors in pelvis or retroperitoneum had an initial symptom of local swelling pain, and lesions in CNS usually caused dizziness and local pain. Tumor size varied among different sites: tumors in head/neck, CNS, lung, and extremities were more likely to be less than 5.0 cm, tumors in mediastinum, pelvis, and trunk/skin were more likely to be 5.1 to 9.9 cm, and tumors in pleura, abdomen, retroperitoneum, and groin were prone to grow to sizes over 10.0 cm (Supplementary Table 2, http://links.lww.com/CM9/A364).

Among all inpatients, 186 (93.5%) underwent surgical intervention, whereas 13 (6.3%) underwent needle biopsy for examination. For the patients who underwent surgical resection, 5 (2.5%) were combined with chemotherapy, and 4 (2.0%) were combined with radiation therapy. The pathology was reconfirmed as benign in 230 (87.8%)

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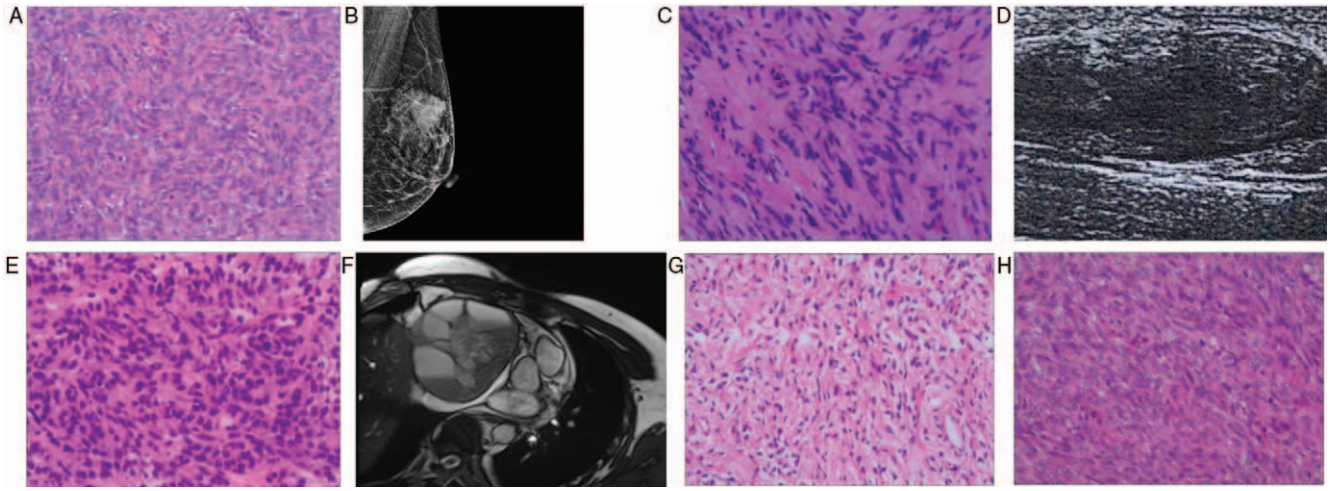

Figure 1: Radiological and histopathological findings of several rare SFTs. (A) HE staining (original magnification ×200) of the breast SFTs case 1 presented a spindle cell tumor composed of round cells with ovoid nuclei, relatively thin chromatin, and embedded in a patternless fibrous collagen stroma, with slight atypia and nuclear division < 3/10/HPF, no bleeding or necrosis was observed; (B) Mammography of breast SFTs case 1 showed an ovoid lobulated mass with unclear boundary in the upper left breast, punctate calcifications are scattered at surroundings; (C) HE staining (original magnification ×200) of the breast SFTs case 2 presented a spindle cell tumor with ovoid nuclei embedded in abundant fibrous interstitium without significant cell atypia; (D) Sonography examination of breast SFTs case 2 presented an ill-defined hypo-echoic ovoid lesion measuring 3.6 cm × 1.9 cm × 0.8 cm with abundant blood flow signal; (E) HE staining (original magnification ×200) of pericardium SFT presented spindle cells and ovoid cells were diffusely distributed with atypia, nuclear division was observed (> 10/10 HPF), and hemorrhage was observed in the interstitium; (F) MRI of pericardium SFT showed that T2WI with mixed high-intensity signals, and no significant enhancement was observed in the left ventricle; (G) HE staining (original magnification ×200) of inguinal SFTs case 1 showed a spindle cell tumor with few atypia and nuclear division. Irregular blood vessels could be found in the tumor interstitium, mixed with some non-atypical cartilage and few calcifications; (H) HE staining (original magnification ×200) of inguinal SFTs case 2 showed the tumor composed of spindle cells that were arranged in striated pattern. The cells presented atypia with nuclear division > 10/10 HPF. HE: Hematoxylin-eosin; HPF: Highpower field; SFTs: Solitary fibrous tumors.

patients and as malignant in 32 (12.2%). The malignancy at each site is shown in Supplementary Table 2, http://links.lww.com/CM9/A364. Recurrence was found in 14 (7.0%) inpatients (Supplementary Table 1, http://links.lww.com/CM9/A363).

Breast SFT case 1: A 56-year-old woman presented with a palpable nodule at 1'o'clock position of the left breast for 2 years. Mammography was shown in Supplementary Figure 1B, http://links.lww.com/CM9/A365. A surgical resection was performed, histologic and immunohistochemical analysis was shown in Figure 1A & Supplementary Figure 1A, http://links.lww.com/CM9/A365. A diagnosis of an intermediate SFT was made. The patient recovered well, with no recurrence during the 1-year follow-up.

Breast SFT case 2: A 66-year-old man presented with a palpable nodule located at 11 o'clock position of the left breast for half-a-month. Sonography examination presented in Figure 1D. A needle core biopsy was performed [Figure 1C] and a diagnosis of a benign SFT was made, no further treatment was performed except for observation. The patient was well during the 1.5-year follow-up.

Pericardium SFT: A 68-year-old man with a 3-month history of progressive exertional dyspnea and chest tightness was admitted. Magnetic resonance imaging (MRI) demonstrated in Figure 1F. The tumor was partly dissected, histologic examination [Figure 1E] suggesting aggressive SFT. However, the patient was lost to follow-up after he was discharged, and no further information was available.

Inguinal SFT case 1 (Benign): A 46-year-old man with a 6-year history of a gradually growing mass in the right inguinal region was admitted. A wide excision of the lesion was performed, and the mass was well circumscribed without evidence of adjacent soft tissue involvement. The histological examination and immunohistochemistry (Figure G & Supplementary Figure B, http://links.lww.com/CM9/A365) indicating the diagnosis of SFT.

Inguinal SFT case 2 (Malignant): A 42-year-old man with a 9-month history of a gradually growing mass in the left inguinal region came to the clinic. A wide excision was performed, showing that the tumor was partly capsulated and infiltrating the peripheral tissue. Histological and immunohistochemistry (Figure 1H & Supplementary Figure 1C, http://links.lww.com/CM9/A365) suggesting atypical/malignant SFT. Although the patient underwent radiation therapy after surgery, local recurrence occurred 9-months later, and he was treated again with surgery and radiation. The patient had an uneventful recovery and had no evidence of disease during a 1-year follow-up.

Reports have shown that the period from fifty to seventy is usually the age of diagnosis of SFTs, in which the youngest patient reported was 5-year-old, we consider that the large size of our series makes our data slightly different from those in former, and the 3-month-old patient in our study might be the youngest SFT patient in all literature. Furthermore, we notice that the symptoms usually depend on anatomic location and the size of the tumor; and the size of tumor depends on the location on the other. Ample space allows the tumor to grow to a

large size, so the symptoms such as compression will appear later.

It has been shown that "benign is not always benign".[2] Several patients were found to have advanced secondary malignancies concurrently or after the diagnosis of SFTs, regardless of whether the resection was complete; this is understandable because the WHO classifies SFTs as neoplasms with intermediate biological potential that run a relatively indolent course but still prone to metastasis or recurrence. Therefore, complete surgical excision is the mainstay treatment and offers the best long-term prognosis. Radiotherapy might be used to shrink tumor or minimize incomplete surgical margins and to improve prognosis.[3] Though under debate, molecular-targeted chemotherapy might be a promising way to treat the disease. In our study, 7.04% of inpatients developed recurrence; considering the varying skill levels of surgeons and the patients who were lost to follow-up, we believe that more data should be collected for validation.

There are only 21 cases of breast SFTs reported in English literature. The typical clinical characteristics are a slowly growing mass without any other symptoms regardless of sex. Sonography examination is one of the most important strategies for diagnosis; however, the preoperative differential diagnosis is often easily confused with other benign tumors, especially fibroadenoma. Change in size of tumor during the menstrual cycle is a typical feature of fibroadenoma, which could help with differentiation. Surgical excision is always the first line treatment for breast SFTs.[4]

Pericardium SFT is exceedingly rare. Their symptoms are similar to those of other intrapericardial tumors, including dyspnea, chest tightness, fatigue, palpitation, syncope, or peripheral edema, and can cause tamponade or congestive heart failure in the long term. Benign SFTs often appear as large masses with clear margins on imaging because of their slowly growing nature and are surrounded by a capsule without infiltration.[5] The basic treatment is surgery to relieve the symptoms and to elucidate the pathological diagnosis.

There are merely 5 cases of SFTs occurring in inguinal region presented in English literature. The most common symptom is a palpable painless mass in the deep soft tissue, thus, this tumor is easily ignored by patients, and we consider this as one of the reasons for the limited number of reported cases. Despite its rarity, inguinal SFTs have similar treatment and prognoses to general SFTs.

Our study has several limitations. First, some SFT patients were treated in the outpatient clinic, making it impossible to obtain a detailed medical history. Second, our pathology review was not blinded; therefore, we included two experienced pathologists to re-review to minimize bias.

In conclusion, we presented one of the largest series of SFT patients over a 10-year period in correlate the clinical characteristics, incidence distribution, and prognosis, and our analysis presented the latest epidemiology of SFTs. Importantly, we reported the youngest SFT patient, and presented several exceedingly rare cases. Together with the above, we hope that these data could provide important assistance with the preoperative differentiation and diagnosis of SFTs.

Conflicts of interests

None.

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