An Unusual Solitary Fibrous Tumor of the Ischiorectal Region

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Abstract

Solitary fibrous tumors (SFTs) are rare fibroblastic/myofibroblastic proliferations that occur in a wide range of anatomical sites. These tumors have nonspecific clinical presentations often with unpredictable biological behavior. SFTs can be slow growing low-risk tumors or rapidly growing high-risk tumors. They show a wide variety of histological features and typically are characterized by NAB2-STAT6 fusion. SFTs of the ischiorectal fossa are rare, with few studies reported in the literature to date. Here, we report a 90-year-old male who had a road traffic accident in October 2018. A pelvic computed tomography (CT) revealed a mass measuring 3.5 × 2.5 cm in the right ischiorectal fossa. Histopathology of the CT-guided biopsies confirmed the diagnosis of low-grade SFT. No surgical intervention was needed since the patient was asymptomatic. In January 2022, a follow-up CT showed a gradual increase in tumor size (5 × 3.5 × 3 cm), but not infiltrating the surrounding structures. However, the patient complained of constipation, which warranted a surgical excision of the mass. Subsequently, immunohistological examination reconfirmed the diagnosis of low-risk SFT. Here, we discussed the clinicopathological features of the case and the relevant literature about pelvic SFTs. In conclusion, SFTs should be considered in the differential diagnosis of any ischiorectal mass. It is recommended that tissue samples be obtained, and immunohistology should be performed.

Keywords: SFT; Rectum; Tumor; Immunohistochemistry

Introduction

Historical aspects of solitary fibrous tumor

Solitary fibrous tumor (SFT) is rare mesenchymal tumor that Stout and Murray first described in 1931 as a pleural fibroma. Other names for SFT include benign mesothelioma, localized mesothelioma, pleural fibroma, subserosal fibroma, submesothelial fibroma, and subpleural fibroma. The incidence of this tumor is one new case per million people per year [1-3]. In 1942, Stout and Murray reviewed historical neoplasm called the “hemangiopericytomas” group. They found that although these neoplasms have seemingly similar histology in the form of profuse stag-horn vascular pattern, surrounded by connective tissue sheath, they did not share the same biological behavior. It has been challenging to diagnose these hemangiopericytomas, as this hemangiopericytoma-like pattern has also been observed in several other soft tissue tumors. The latter include mesenchymal chondrosarcomas, fibrosarcomas, and synovial sarcomas [1-7].

Hemangiopericytoma and SFT

After the discovery of NAB2-STAT6 gene fusion, World Health Organization (WHO) defined hemangiopericytoma and SFT as a single entity in 2016 [4-11].

Clinical locations of SFTs

SFT occurs most frequently in the pleura but has been described in several anatomic sites, including the viscera and soft tissues, especially the deep soft tissues. The thorax (lung, mediastinum, and diaphragm) is the most common extra-pleural site. SFTs can occur at intraperitoneal, retroperitoneal, or pelvic locations in the abdomen, which is the second most common extra-pleural site for SFTs [12-14]. Other rare anatomic sites that are affected by SFTs include the head and neck region, extremities, and meninges [11, 14-16]. However, the sites of predilection for SFTs are the pleura, peritoneum, and meninges [17]. WHO classification (2020) of tumors considered SFT as a neoplasm with fibroblastic/myofibroblastic differentiation that rarely metastasizes. SFTs are sometimes classified as intermediate malignant tumors [18].
The clinical features of the pelvic SFTs

SFTs of the pelvic area are very rare. In addition to the sigmoid, rectum (serosa) and mesorectum, they can be found in the prostate, urinary bladder, spinal sacral canal, perineal area, and ischiorectal and ischioanal fossae [14-16, 19-28]. There have been only a few reported cases of SFTs involving the ischiorectal fossa in the literature [15, 16, 19, 21, 22, 25, 26, 28].

SFTs of the pelvic region have no sex predilection, and the peak incidence is between 40 and 70 years [18]. The SFTs in the pelvis usually involve deep tissue rather than superficial. SFTs in the pelvis are generally asymptomatic until they become large enough to produce mass effects on the surrounding tissues. There are no specific clinical features of pelvic SFTs [15, 16, 19, 21, 22, 25, 26, 28]. They include abdominal pain [28], difficulty in urinating, urinary retention [27], numbness and weakness in lower extremities due to nerve compression, constipation, and dysuria [28, 29]. A summary of some previous cases is presented in Table 1 [14-16, 19-28, 30-32].

The paraneoplastic syndromes and the SFTs

SFTs may rarely be associated with paraneoplastic syndromes, most commonly non-islet cell hypoglycemia with the production of insulin-like growth factors (IGFs), specifically IGF-II. Most of these tumors are malignant; therefore, the presence of non-islet cell hypoglycemia is considered a poor prognostic factor [33, 34]. Rarely, SFTs may also be associated with other paraneoplastic syndromes like Pierre-Marie-Bamberger syndrome [35], Doege-Potter syndrome [36], cerebellar degeneration [37], and hypertrophic osteoarthropathy [38].

The radiological features of the SFTs

The diagnostic approach of pelvic SFTs is similar to other soft tissue tumors at varied anatomical sites. The diagnosis of SFT is often incidental on radiography or CT [39], with appearance being nonspecific [39]. Imaging studies include plain radiographs that typically reveal a well-defined mass. Contrast-enhanced computed tomography (CT) usually shows a well-de-
fined, hypervascular mass lesion.

SFT can be hypodense or hyperdense with respect to muscle. The attenuation of SFT depends on the content of the collagen bundles. The densely collagenized SFTs are usually hyperdense. Conversely, the hypodense SFTs usually have fewer collagen contents [39]. The SFTs are usually hypoechogenic on ultrasonographic examination but occasionally may have a heterogeneous appearance [39]. Alternatively, on magnetic resonance imaging (MRI), these tumors are isointense on T1-weighted images and can be variable on T2-weighted images [40]. Benign SFTs have low-grade activity (hypometabolic state), whereas their malignant counterparts usually have hypermetabolic and homogeneous activity on fluorodeoxyglucose-positron emission tomography (FDG-PET) [41].

The gross and histological features of the pelvic SFTs

SFTs are typically more than 1.0 cm in diameter (size ranging from 1.0 to 40 cm) [42, 43]. They are typically well-defined, can have a smooth surface or be lobulated, may or may not have a capsule. Cut sections of these masses are usually yellow or grayish white. Cystic changes or necrosis is rare [44].

SFTs in the pelvis are characterized histologically by a spindle-to-ovoid-shaped proliferations of fibroblasts/myofibroblasts associated with staghorn-like vasculature. Some SFTs may have hemangiopericytoma-like or storiform pattern or can be patternless. Tumor cells consist of monomorphic or mildly atypical spindle-shaped to ovoid-shaped cells with minimal cytoplasm, arranged in undulating, straight, curved, or patternless arrangements [26, 43, 45]. Tumor cells are embedded in a variable cellular fibrous stroma which are separated by bands of hyalinized, ropy collagen. Some tumors have poorly cellular, densely collagenized stroma, while others have an abundant cellular stroma [11, 18]. Some tumors are homogeneously cellular, while others have a cellular zone alternating with hypocellular or keloid-like areas. They are accompanied by hemangiopericytoma-like vessels [11, 18].

The grade of SFTs depends upon several histological features including the mitotic activity, necrosis and nuclear pleomorphism. The degree of mitotic activity may range from the complete absence or scarcity of mitotic activity to mitotically active stromal cells with some atypical mitotic figures. This feature is crucial in the risk stratification of SFTs. Moreover, the degree of nuclear pleomorphism, and necrosis are also important histological features for risk stratification of these tumors [11, 46].

Variants of the SFTs

SFTs have several histological variants [11]. The giant cell rich SFTs (formerly known as giant cell-angiofibroma) are common in the head and neck region and tend to be indolent. The differentiated SFTs (also known as anaplastic SFTs) are the most aggressive variants and contain areas of high-grade sarcoma encompassing heterologous elements such as bone-forming sarcoma or rhabdomyosarcoma [47, 48]. Fat-forming SFTs contain mature adipose tissue and have an indolent course similar to giant cell rich SFTs. Alternatively, this variant may contain lipoblast (lipomatous SFTs) and present with malignant behavior [47, 49].

The immunohistochemical and ultrastructural features of the SFTs

Immunohistochemically, the neoplastic cells in SFTs are usually strongly reactive to CD34 and STAT6. Although CD34 is a conventional marker in SFT, it can be expressed by other soft tissue tumors and therefore, lack specificity. Alternatively, STAT6 is considered the most sensitive and specific marker for SFTs. The tumors are also variably reactive to other markers, including CD99, BCL-2, EMA, and nuclear β-catenin [28, 29, 50]. The tumor cells of SFT are negative for SOX10, desmin, CD31, pancytokeratin (AE1/AE3), and S-100 [11]. An ultrastructural description of SFTs is that they are composed of cells that are fibroblast-like and have a well-developed rough endoplasmic reticulum. These cells are seen amid collagen fibers [51]. Some SFTs may also have a myxoid stroma [47].

Risk stratification of the SFTs

The behavior of SFTs has been difficult to predict. Our knowledge about the biological behavior and malignant potential of SFTs is rudimentary. Nonetheless, available case reports indicate that most of these tumors are indolent and slowly growing [44]. However, SFTs can metastasize in 5-25% of cases [46]. England et al proposed five criteria for judging malignant change in SFT. They include necrosis, mitotic activity (more than four mitotic figures per 10 high-power fields (HPFs)), high cellularity, pleomorphism, and hemorrhage [52].

The conventional sarcoma grading systems have proved to be poorly applicable to SFT. Also, the validity of the current sarcoma staging systems for traditional sarcomas has not been tested for these tumors. SFTs with sarcomatous changes usually behave aggressively and can metastasize like other high-grade sarcomas. Alternatively, it is difficult to predict the biological behavior of tumors lacking overt sarcomatous changes [53]. Based on the risk stratification, SFTs include low aggressive, highly aggressive, and dedifferentiated SFT. Out of the several risk classification models, only two of them have been validated. One model is based on age, size, mitotic count, and tumor necrosis and distributes the patients into three different risk categories. The other risk model estimates the individual risk for local and metastatic recurrence [11, 46].

The genetic features of the SFTs

NGFI-A binding (NAB) proteins can repress or activate transcription induced by some members of the EGR (early growth response) family of transactivators [54]. The STAT6 is a tran-
The somatic fusions of the two genes, located at chromosomal region 12q13, namely NGFI-A-binding protein 2 (NAB2) and STAT6, have been recently considered as the tumor-initiating events in SFTs [60]. STAT6 nuclear reactivity in SFTs has diagnostic sensitivity and specificity [58, 59]. The most common NAB2-STAT6 fusion variants in these tumors are NAB2ex4-STAT6ex2, NAB2ex6-STAT6ex16, and NAB2ex6-STAT6ex17. The NAB2ex4-fused SFTs represent a distinct from non-NAB2ex4-fused counterparts in several clinical and pathological aspects [61].

The treatment strategies of the SFTs

Given our rudimentary knowledge about SFTs, there are no clearly defined treatment protocols. Surgery (tumorectomy with wide negative surgical margin) is the mainstay. Surgical excision may be the only treatment necessary for SFTs of the pelvic region with no or low recurrence rate. Radiotherapy is not generally recommended after surgical excision as most of these tumors are indolent or low risk. Chemotherapy using certain drugs (such as bevacizumab, sunitinib, pazopanib, and sorafenib) that target the vascular endothelial growth factor and other tyrosine kinase signaling pathways are sometimes used. These pathways interfere with the blood supply to the tumor and are used to stop the progression of the tumor [62-65]. Patients with malignant SFTs or tumors with positive margins or unresectable or recurrent tumors may benefit from radiation therapy [66].

Case Report

A 90-year-old male, otherwise healthy, had a road traffic accident in October 2018. A CT scan was requested as a routine workup of trauma assessment. It revealed an incidental finding of asymptomatic round, well-defined soft tissue density with variable enhancement in the right ischiorectal fossa measuring 3.5 × 2.5 cm. Following this, MRI revealed a well-defined mass lesion of abnormal signal intensity measuring 3.5 × 2.5 cm in the right ischiorectal fossa, inseparable from the pelvic sidewall muscles laterally. The mass was in close contact with the rectum medially and superiorly. It was seen separable from but displacing the pelvic floor muscles. The mass lesion displayed mixed intermediate and hyperintense signals on the T2-weighted image. These findings were impressive of a neoplastic process. A summary of these findings is shown in Figure 1.

CT-guided biopsy was taken, and the histological examination revealed a spindle-cell-shaped neoplasm composed of patternless proliferation of short spindly cells with mitotically inactive nuclei, inconspicuous nucleoli, and pink cytoplasm with some collagen fibril network. The proliferating cells were admixed with small blood vessels (Fig. 2). The tumor cells were positive for CD34, BCL2, CD99, and STAT6. They were negative for desmin, S100, and pancytokeratin (AE1/AE3), supporting the diagnosis of SFT. The patient was offered an excisional biopsy, but he refused.

More than 3 years later (January 2022), the patient came back with newly developed vague symptoms related to the mass lesion (constipation). A follow-up imaging showed only a mild increase in the lesion size. Radiological studies revealed well-defined heterogeneous predominantly low-signal right ischiorectal fossa cone-shaped mass (Figs. 3, 4). The patient agreed to surgical intervention (excision of the mass). The tumor was below the pelvic floor and therefore abdominal approach is not an option. The patient was put in lithotomy position, and under general anesthesia, a longitudinal incision was made just lateral to the site of the external sphincter muscles. The surgical dissection was extended upward till the site of the mass. The lesion was soft, movable, and compressible.

Figure 1. Radiological MRI features of solitary fibrous tumor (MRI: October 2018). (a) Coronal T2WI with fat saturation shows a well-defined heterogeneous predominantly low signal intensity right ischiorectal fossa ice-cone shaped mass (white asterisk). (b, c) Pre and post contrast-enhanced coronal T1WI with fat suppression showing homogenous lesion isointense to muscles in pre-contrast and avidly enhancing following contrast administration (orange asterisk). MRI: magnetic resonance imaging.
The mass could not be grasped and taken out without applying a digital transrectal pushing the mass outward. The mass was released, delivered out, and excised completely. The wound was closed in a layer, and a Penrose drain was used to drain the cavity and prevent fluid collection and infection.

Gross examination revealed a well-defined, thinly encapsulated mass measuring 5 × 3.5 × 3 cm with a pink cut section and firm consistency. Histological examination revealed haphazard proliferation of spindly and oval mitotically inactive cells with bland short spindly and oval nuclei, pale eosinophilic cytoplasm, and variable amounts of the collagenous stroma. The tumor was cellular, and the mitotic count was 0/10 HPFs. There was no evidence of necrosis or hemorrhage (Fig. 5). The resection margins were free. The postoperative recovery was

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**Figure 2.** Histological features of solitary fibrous tumors (true cut needle biopsy: October 2018). (a-d) The tumor consists of a haphazard proliferation of short spindle to ovoid cells with banal-looking oval to spindle nuclei, pale eosinophilic cytoplasm, and some collagenized stroma (star), and the hemangiopericytoma-like vascular structures (arrow). The tumor cells are negative for SMA (e) (original magnifications: (a) × 20; (b) × 200; (c) × 400; (d) × 200; and (e) × 200).

**Figure 3.** Radiological CT features of solitary fibrous tumor (CT: January 2022). (a, b) Contrast-enhanced CT of the pelvis shows a well-defined right ischiorectal fossa oval-shaped mass (white arrow) with progressive contrast enhancement on delayed CT (b). (c) Contrast-enhanced CT coronal reformat shows a well-defined elongated right ischiorectal fossa mass (orange arrow). CT: computed tomography.
uneventful, and the drain was removed 2 weeks later without any complications. According to the four-variable model for risk stratification, the tumor was considered as low risk (score of 3) [43].

Discussion

Given that SFTs of the pelvic region are rare neoplasms, much of the literature has emerged from the occasional case reports. Therefore, our knowledge about this entity is not only limited but also fragmented. We pursued this study to improve our understanding of these neoplasms. To achieve our goal, we presented a literature review about pelvic SFTs. Also, herein, we presented a case of ischiorectal SFT in an elderly male patient. The characteristic immunohistological profiles of the tumor and the nonspecific clinical manifestations of the case presented here concur with previous studies [14-16, 19-28]. Nassif

Figure 4. Radiological MRI features of solitary fibrous tumor (MRI: January 2022). (a) Coronal T2WI shows right well-defined heterogeneous predominantly low signal right ischiorectal fossa ice cone-shaped mass (white asterisk) upward displacing the right levator ani muscle (yellow arrow) without invasion. (b, c) Pre- and post-contrast-enhanced coronal T1WI with fat suppression show homogenous lesion isointense to muscles in pre-contrast and avidly enhancing following contrast administration (orange asterisk). MRI: magnetic resonance imaging.

Figure 5. Gross and histological features of the solitary fibrous tumor (excisional biopsy: January 2022). Gross examination of the mass reveals well-defined, thinly encapsulated mass measuring about 5 × 3.5 × 3 cm (a). Histologic sections reveal cellular spindle cell neoplasm in a collagenous stroma with variable-sized blood vessels having stag-horn vascular morphology (b). The neoplastic cells are ovoid, short, or fusiform spindle-shaped cells with indistinct cell borders, bland looking nuclei, and indistinct nucleoli. They are arranged in short, poorly defined bundles, haphazardly fashion, and patternless pattern. The cells are streamed among the dermal collagens. No mitotic activity was seen. No significant nuclear atypia or necrosis was seen (c, d). The tumor cells are diffusely and strongly positive for CD34, BCL2, and CD99 (e, f, and g, respectively) and are negative for desmin, S100, and pancytokeratin (AE1/AE3) (original magnifications: (b) × 40, (c) × 200, (d) × 400, (e) × 200, (f) × 200, and (g) × 200).
et al [44] presented a literature review of five reported cases of anorectal SFTs [14, 19, 24, 67, 68]. The tumors occurred in four males and one female. The size of the tumors ranged from 7 cm to 13 cm. The surgical resection with a clear margin was the mainstay of treatment in all cases. The patient did not receive chemotherapy or radiotherapy. None of the patients received adjuvant chemotherapy radiation [44].

In the case reported herein, there was a slow increase in the size of the tumor over a period of more than 3 years. This indicates the indolent nature of the tumor. Moreover, the assessment of risk stratification suggests that the tumor belongs to the low-risk group following the scheme proposed by Demicco et al and Bhat et al [30, 46]. In 2012, Demicco et al initially reported a three-variable scheme (age, tumor size, and mitosis) on the risk stratification for SFT [46]. In 2017, the authors revised their scheme by incorporating tumor necrosis (necrosis representing 10% or more of the tumor) as a new risk factor to their previously reported variables (patient age, tumor size, and mitotic activity). According to this revised scheme, the revised risk stratification model for SFT comprised of low risk (0 - 3), intermediate-risk (4 - 5), and high risk (6 - 7) groups. The authors validated their revised scheme in 79 patients with primary non-meningeal SFTs. Most of the patients (66%) were at low risk and had no metastasis at 10 years. Some patients (24%) were scored as an intermediate risk with a 10% risk of metastasis at 10 years. The high risk included 10% of the patients with a 73% risk of metastasis at 5 years [46]. According to this risk stratification, the case presented here is considered as the low-risk tumor. The slow growth and lack of invasion of the surrounding ischiorectal structures support the indolent behavior of the tumor.

The histological differential diagnosis of the ischiorectal region SFT reported herein includes other soft tissue tumors with the hemangiopericytic-like vascular pattern, including synovial sarcoma (negative for STAT6 and CD34 and has SS18-SSX gene fusions) [69], dermatofibrosarcoma protuberans (positive for CD34 but negative for STAT6, and has OLA1-PDGFB gene fusions) [70], deep fibrous histiocytoma (negative for STAT6) [71], and liposarcoma (MDM2 gene amplification by fluorescence in situ hybridisation (FISH)) [72]. The other tumors in that should be separated from SFTs include mesenchymal chondrosarcoma (chondroid component, negative for STAT6 and the presence of HEY1-NCOA2 gene fusions) [73, 74], myopericytoma (positive for smooth muscle actin and negative for STAT6) [75], gastrointestinal stromal tumor (positive for CD34, DOG1, and CD117 but negative for STAT6) [76], and mammary type myofibroblastoma (positive for desmin and ER but negative for STAT6) [77].

To conclude, SFTs are mesenchymal neoplasms with fibroblastic/myofibroblastic differentiation and unpredictable biological behavior. The physicians should be aware of this entity whenever presented with any ischiorectal mass lesion.

### Learning points

Although SFTs were first reported in the pleura (localized fibrous mesothelioma), they were subsequently reported in several anatomical sites.

- Pelvic SFT, including the tumors of the ischiorectal and ischioanal fossae, perineal region, sigmoid (serosa), rectum (serosa), mesorectum, prostate, urinary bladder, sacral spinal canal, obturator area, are rare.
- There are no sex predilections in SFTs.
- SFTs most commonly occur in the age range between 20 -70 years.
- The somatic fusions of NAB2 and STAT6 genes are the tumor-initiating events in SFT.
- The clinical presentations of the pelvic SFTs are nonspecific, and they include constipation, dysuria, bleeding per rectum, pain in the lower extremities associated with numbness, and muscle weakness.
- SFTs are histologically composed of spindle cells and a hemangiopericytoma-like vasculature. The spindled cells can show variable cytologic atypia, pleomorphism, and mitotic figures/10HPFs, but no hemorrhage or necrosis. Immunohistochemically, the tumor cells show strong and diffuse positivity for CD34 and STAT6. The neoplastic cells are negative for desmin, smooth muscle actin, inhibin, and CD117.
- The hemangiopericytoma-like vasculature and the salient histological features of SFTs.
- The histological variants of SFTs include the fat-forming SFTs, the giant cell-rich SFTs, and the dedifferentiated SFTs. STAT6 nuclear reactivity in SFTs has diagnostic sensitivity and specificity [58, 59].
- The features of malignancy in SFTs include increased cellularity, mitosis, and necrosis.
- The behavior of SFTs has been difficult to predict.
- Risk-stratification of SFTs is based on the patient’s age, size of the tumor, mitotic activity, and tumor necrosis.
- Based on the risk stratification, SFTs include low-risk, intermediate-risk, and high-risk tumors.
- Surgical excision (tumorectomy) is the standard management in most cases of pelvic SFTs.
- The beneficial roles of adjuvant chemotherapy or radiotherapy is still unclear.

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### Conflict of Interest

None to declare.

### Informed Consent

The informed consent was obtained from the patient.
Author Contributions

All authors certify that he or she has equally participated sufficiently in the intellectual content, the analysis of data and the writing up of the manuscript. Each author has reviewed the final version of the manuscript and approved it for publication.

Data Availability

The authors declare that data supporting the findings of this study are available within the article.

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