Giant solitary fibrous tumor of the pelvis invading the rectum and bladder: a rare case report in an adolescent

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Case Report

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Abstract

Background

Pelvic solitary fibrous tumor is a rare spindle-cell tumor arising from mesenchymal tissue, usually benign. However, its malignant form is an extremely rarer, and more aggressive disease. Our review of previous literature found that it has not been reported in adolescents younger than 18 years. Herein, we describe a case of pelvic giant solitary fibrous tumor with rectum and bladder invasion in a 16-year-old girl and summarize the diagnosis and treatment experience to further improve the existing management of solitary fibrous tumor.

Case Description

Retrospectively analyzed a 16-year-old girl admitted to our department with irregular menstruation and increased menstrual bleeding for 2 months. A computed tomography scan of the abdomen demonstrated an 11.2×7.5×8.9 cm isodense space-occupying lesion in the pelvis. Contrast-enhanced computed tomography showed heterogeneous enhancement, which was considered a tumor of mesenchymal origin. Pelvic mass resection, bladder repair, and right ureteral stent placement were performed. It was confirmed that the mass invaded the upper rectal and the right bladder wall during the operation. After complete resection of the tumor, malignant solitary fibrous tumor was diagnosed in combination with histopathology and immunohistochemistry. The patient survived well with no tumor metastasis or recurrence in 4 months of postoperative follow-up.

Conclusion

This case report suggests that pelvic solitary fibrous tumor is rarely seen clinically. Because of its non-specific clinical manifestation and imaging, definite diagnosis is mainly based on histopathology and immunohistochemistry. Complete resection of the tumor is the first-line treatment, and most patients have a good prognosis.

Introduction

Solitary fibrous tumor (SFT) was first identified and reported in the pleura by Klemperer and Rabin in 1931[1]. This type of neoplasm has been described as a rare mesenchymal tissue-derived spindle-cell tumor, mostly with benign or borderline biologic behavior. But malignant solitary fibrous tumor (MSFT) is even rarer than SFT. It may present with metastasis or recurrence after surgical intervention. SFT can occur in any site of the body, most common in the pleura (approximately 30%), followed by the meninges (approximately 27%)[2]. But MSFT in the pelvis accounts for only 6% of cases[3]. All of the reported SFTs are usually observed in middle-aged and older patients (ranging from 20 to 70 years), equally affecting
male and female patients[4]. However, to the best of our knowledge, this case is the first report of SFT arising from the pelvis with adjacent organ invasion in the adolescent age group.

**Case Presentation**

A 16-year-old girl attended the local hospital with irregular menstruation and increased menstrual bleeding for 2 months. The gynecologic sonography indicated a hypoechoic space-occupying lesion on the right side of the pelvis, considered a broad ligament myoma. She was admitted to our department for further diagnosis and treatment. On physical examination, a hard mass with obscure borders and poor mobility was palpated in the right lower abdomen, without tenderness and rebound tenderness. Rectal palpation did not reveal any obvious abnormality. The abdominal computed tomography (CT) revealed an isodense mass with clear borders on the right side of the rectum in the pelvis, measuring approximately 11.2×7.5×8.9 cm, and the contrast-enhanced CT images demonstrated heterogeneous enhancement with patchy non-enhancing areas within the lesion (Fig. 1A), which was considered as a tumor originating from mesenchymal tissue. Multiple tortuous and thickened vessels were seen within and surrounding the mass (Fig. 1B), and the rectum, uterus, and vagina were displaced by compression (Fig. 1C).

After excluding surgical contraindications, the patient received pelvic mass resection, bladder repair, and right ureteral stent placement under general anesthesia. Intraoperatively, there was a hard mass located in the posterior part of the uterus and bladder and the right side of the rectum, with a size of about 12.0×10.0×10.0 cm, pressing the uterus, bladder, and rectum to the left side, and it was surrounded by pronounced nourishing vessels with rich blood flow. The mass invaded the upper rectal and the right bladder wall, with obvious adhesions and poor mobility. After fully dissociating the tumor, it was completely excised and sent for pathological examination.

The gross specimen of the pelvic tumor was a lobulated mass partially encapsulated on the surface (Fig. 2A). Cross section showed gray-red and gray-white, fish flesh-like, and partly granular. Microscopically, the hemangiopericytoma-like arrangement was observed, with abundant tumor cells in some areas, mild to moderate atypia and mitotic figures of 4/10HPF. There was no necrosis (Fig. 2B, C). Immunohistochemical staining showed positive CD34, Bcl2, STAT6, and Vimentin, focal positive CD99 and CR, along with notably weaker positive P53 and Ki67, but was negative for CK, D2-40, S-100, SMA, and Desmin (Fig. 2D-F). The diagnosis of pelvic malignant solitary fibrous tumor was made.

The intraoperative blood loss of the patient was approximately 2500 mL, and blood transfusion was given in time during and after the operation. The right ureteral stent was removed 40 days after surgery, with no obvious complications at the discharge time.

Our patient recovered well after surgery without any adjuvant therapy. There was no evidence of tumor recurrence or metastasis 4 months after pelvic tumor resection (Fig. 3). The follow-up period was too short to comment on the long-term survival outcomes. The girl and her parents were very satisfied with our treatment and were willing to accept long-term close follow-up and further therapeutic measures.
Discussion And Conclusions

SFT is a clinically indolent neoplasm originating from mesenchymal cells, commonly found in the pleura, meninges, and extrapleural soft tissues, accounting for less than 2% of all soft tissue tumors[5]. In comparison, SFT in the pelvis is extremely rare and usually more aggressive than the pleural form[2].

Pelvic SFT typically presents as a slow-growing painless mass with no noticeable clinical symptoms in its early stages. However, it can push and press the nearby organs and cause relevant symptoms, such as abdominal discomfort, urination disturbance, and bowel obstruction, until it is large enough. A small number of patients can be characterized by non-islet cell hypoglycemia[6]. In this case, the girl presented with irregular menstruation and increased menstrual bleeding as the initial symptoms, which were considered to be related to the compression of the uterus and adnexa by the mass.

Imaging examinations are of great value in the pretreatment diagnosis of SFT. Ultrasonography, often used as the first diagnostic and screening method, generally shows hypoechoic or moderate-hypoechoic space-occupying lesions with clear boundaries from adjacent tissues and organs. Although SFT is hypervascular, it does not always show obvious blood flow signals during Doppler imaging[7]. The appearance of SFT on CT and magnetic resonance imaging (MRI) is similar to that of other soft tissue tumors in the pelvis. It is generally a single, well-defined, round, or oval soft tissue mass. In larger cases, it can be lobulated or irregular, pushing the surrounding organs to cause compression or displacement. The lesions were isodensity on CT scan, isointensity on T1-weighted imaging, iso/hyperintensity on T2-weighted imaging, and hyperintensity on diffusion-weighted imaging. Because of the mature fibrous tissue within the tumor and the rich blood supply, the enhanced CT images can reveal characteristic heterogeneous "map-like" enhancement, and multiple enhanced vascular can be seen in and around the tumor[8].

Due to the rarity of this tumor, imaging examinations are mainly used to find, locate, and display morphological features in diagnosing SFT, which have great limitations in the qualitative and differential diagnosis of the tumor. The definite diagnosis requires the combination of histopathological features and immunohistochemical results. Classically, SFT is microscopically composed of ovoid to spindle tumor cells arranged in patternless distribution, with intercellular fibrous deposits, unclear cell boundaries, homogeneous chromatin, nuclear vacuoles, no obvious atypia, and rare mitotic figures. In addition to the above manifestations, MSFT is usually characterized by abundant and dense tumor cells, frequent mitotic figures (≥ 4/10HPF), obvious pleomorphism, hemorrhage, necrosis, and infiltration of surrounding tissues[9]. In one study, patients were divided into low-risk, intermediate-risk, and high-risk by incorporating the four variables, including patient age, tumor size, mitotic activity, and tumor necrosis, and then a risk stratification scheme for SFT metastasis was constructed[10]. According to the above criteria, our patient could be considered the intermediate-risk. Regarding immunohistochemical staining, SFT typically shows positive for CD34, Bcl2, CD99, and Vimentin, while Desmin, CD68, CD117, S100 protein, and epithelial markers such as EMA and CK proteins are generally stained negative. Although CD34, Bcl2, CD99, and Vimentin have long been used as differential markers to distinguish SFT from
other histologically similar lesions, all of them lack specificity, and it is not easy to make a clear diagnosis in the case of atypical histology. Recently, a research group found that the NAB2 gene located at the 12q13 locus in SFT was fused with the STAT6 gene, resulting in the generation of the NAB2-STAT6 fusion gene, which plays a key role in the occurrence and development of SFT[11]. Several studies have shown that this fusion gene product, STAT6 protein, is a highly sensitive and specific marker for SFT, with an overall sensitivity of 98% and a specificity of greater than 85%[12–14].

In the management of STF, no standard of care has been established, but complete surgical resection has been the first-line therapy[15]. If there is peripheral organs invasion, the whole resection should be performed as far as possible according to the intraoperative results. Pelvic SFT surgery is difficult and risky. The narrow pelvic space, poor surgical field, limited operation, and the fact that SFT is often rich in nourishing vessels, as well as the long-term compression of the surrounding veins by the tumor leading to stasis expansion, all of which make surgical resection extremely difficult[15, 16]. Therefore, complete tumor resection is the key to the prognosis of patients with pelvic SFT. Postoperative adjuvant chemotherapy or radiotherapy is not used as a routine treatment. However, some studies have shown that surgery combined with radiotherapy can enhance the local inhibition of tumor growth and reduce the recurrence rate. For patients whose tumors cannot be completely removed, postoperative radiotherapy can achieve better results[17]. SFT is poorly sensitive to conventional chemotherapeutic agents, but in recent years the targeted drug Sunitinib has achieved certain efficacy in the treatment of SFT, which has potential value in controlling the progression of SFT[18].

In conclusion, the relatively low incidence of pelvic SFT, which lacks specific clinical symptoms and imaging manifestations, has made it require to be diagnosed through histopathology and immunohistochemistry. Complete surgical resection is the first choice for treatment. As for potential malignant tumors with positive surgical margins, postoperative radiotherapy or targeted therapy can be used. Since SFT tends to recur, long-term, close follow-up is necessary.

Through this case report, we have summarized the following lessons to provide a reference for the subsequent management of pelvic SFT. First, a preoperative needle biopsy is necessary, which can better define the nature of the tumor and make targeted clinical decisions. Second, when the mass is suspected of having rich nourishing vessels on imaging, interventional embolization of the tumor-feeding artery should be performed preoperatively to reduce intraoperative bleeding, shorten the operation time, and speed up the postoperative recovery of patients. Furthermore, we recommend that all patients with pelvic SFT should be closely followed up and regularly reviewed based on SFT prognostic scheme.

**Abbreviations**

SFT: Solitary fibrous tumor; MSFT: Malignant solitary fibrous tumor; CT: Computed tomography; MRI: Magnetic resonance imaging.

**Declarations**
Ethics approval and consent to participate

This report was reviewed and approved by the Ethics Committee of China-Japan Union Hospital of Jilin University.

Consent for publication

Written informed consent was obtained from the patient and her family for the publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Availability of data and materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

YJ designed and revised the work. QC participated in all aspects of this report: management of the patient, conceptualization of the report, collection of the data, and writing of the draft. DS and BS performed the operation and made revisions. All authors read and approved the final manuscript.

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References


Figures

**Figure 1**

Abdominal contrast-enhanced CT scan shows a pelvic tumor. (A) Contrast-enhanced CT images showed a round, well-defined mass with obvious enhancement and patchy non-enhanced areas inside. (B) The tumor is hypervascular. Multiple tortuous and thickened vessels are seen within and around it (white arrows). (C) The circular dashed box is marked as the compressed vagina, and the rectangular dashed box is marked as the compressed rectum.
Figure 2

Pathological features of the pelvic SFT. (A) The gross specimen of the pelvic SFT reveals a well-circumscribed and lobulated mass. (B) (H&E ×100) Low-power view shows the hemangiopericytoma-like arrangement. (C) (H&E ×400) High-power view shows frequent mitoses. (D) (×100) Immunohistochemistry shows positive CD34. (E) (×100) Immunohistochemistry shows positive Bcl-2. (F) (×100) Immunohistochemistry shows positive STAT6.
Figure 3

Timeline of the case. Postoperative CT images show no evidence of tumor recurrence and metastasis.