A hypervascular malignant solitary fibrous tumor in the mesentery of the rectum with good response to preoperative radiation therapy

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Abstract Solitary fibrous tumor (SFT) is an uncommon neoplasm located in the pelvis. Here, we report a case of hypervascular malignant SFT in the mesentery of the rectum that showed a significant response to preoperative radiotherapy. A 36-year-old man presented to our department with constipation and dysuria. Abdominal computed tomography confirmed a large round neoformation (155 mm in the longest diameter) with an evident contrast-enhanced periphery and a central necrotic area in the pelvis. Considering the results of the examinations, a pelvic neoplasm was strongly suspected. Due to the difficulty of safe gross total removal, a diagnostic laparotomy was indicated. Histologically, hypercellularity with spindle-shaped cells and high mitotic activity was observed. Immunohistochemically, the tumor was positive for CD34, CD99, and bcl-2. The diagnosis of hypervascular malignant SFT was made. After the appropriate information had been given, the patient chose the use of preoperative radiotherapy. After local irradiation, the tumor size regressed to 105 mm in the longest diameter. Laparotomy revealed a tumor originating from the mesentery of the rectum, and the tumor was successfully excised. This case strongly suggested the

potential applicability of preoperative radiotherapy for the safe gross total removal of a large pelvic SFT.

Keywords Solitary fibrous tumor (SFT) · Mesentery · Radiotherapy · CD34 · CD99 · bcl-2

Introduction

Solitary fibrous tumor (SFT) is an uncommon neoplasm with a reported incidence of 2.8 cases per 100,000 people [1, 2]. SFT may develop in any organ or tissue. It is usually found in the pleura, but can also be found in other sites such as the meninges, orbit, nasal cavity, respiratory tract, retroperitoneum, soft tissue, and urogenital organs [3, 4]. The most effective therapeutic modality for SFTs is surgical resection. The role of the other treatment modalities, such as radiotherapy and chemotherapy, is presently unclear because of the rarity of SFTs. Here, we report a case of hypervascular large malignant SFT that developed in the mesentery of the rectum, which showed a significant response to preoperative pelvic radiotherapy for the safe gross total removal, and discuss the clinical, radiological, and pathological findings.

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Case report

A 36-year-old man presented to our department with constipation and dysuria. He complained of a dull lower abdominal pain, and on the physical examination, a huge firm mass was palpable in the pelvis. The blood test revealed a raised white blood cell count of $1.22 \times 10^4/\mu\text{l}$ and a high C-reactive protein level of 12.61 mg/dl. In urinalysis, the sediment contained innumerable white blood cells per high-power field.

An abdominal radiography showed a smooth-surfaced, well-demarcated mass in the pelvis (Fig. 1). Abdominal computed tomography (CT) confirmed a large round neoplasia (155 × 84 × 77 mm in size) displacing the bladder and rectum, without the evident invasion, with an evident contrast-enhanced periphery and a central necrotic area (Fig. 2a). Magnetic resonance imaging (MRI) also confirmed the large solid mass in the pelvis. T1-weighted imaging showed a low-signal-intensity area and T2-weighted image showed a low-signal-intensity area including high-signal intensity foci, which indicated the fibrous nature of the tumor (Fig. 3a, b). The mass was mainly fed by the inferior mesenteric artery.

Fig. 1 The abdominal X-ray before therapy. It showed a smooth-surfaced, well-demarcated mass in the pelvis

Fig. 2 Time course changes of the tumor. **a** Image of pre-radiotherapy demonstrating a large tumor (155 × 84 × 77 mm in size) with an evident contrast-enhanced periphery and a central necrotic area. **b** Image of post-radiotherapy showing a significant reduction of the tumor size (105 × 77 × 67 mm in size)

Taking the results of the physical, laboratory, and radiographic examinations, a neoplasm of the pelvis of unknown origin was strongly suspected. Due to the difficulty of a safe gross total removal dependent on the size and hypervascularity of the tumor, a diagnostic laparotomy with a small incision was indicated after adequate information had been given to the patient. Histologically, high-vascularity, hypercellularity with spindle-shaped cells, and high mitotic activity (5/10 high-power fields) were observed (Fig. 4a, b). Ki-67 was positive in approximately 15 % of the cellular population (Fig. 4c). Immunohistochemically, the tumor was diffusely positive for CD34, CD99, and bcl-2. Based on the pathological features and the clinical presentation, the diagnosis of hypervascular malignant SFT was made. Then, based on a few previous reports [5, 6], we planned preoperative radiation therapy in an attempt to reduce the risk of bleeding during the operation. Adequate information concerning the risks and benefits of the different kinds of therapeutic modalities was given, and the patient chose the performance of preoperative radiation therapy.

After performance of local irradiation at a total of 50 Gy, the tumor size regressed dramatically from 155 to 105 mm in the longest diameter (partial response, 32.3 % reduction according to RECIST, Fig. 2b) during 3 months. The CT scan also confirmed the tumor with an evident contrast-enhanced periphery like in the pre-radiotherapy exam. By laparotomy, the origin of the tumor was confirmed to be the mesentery of the rectum, which showed fibrous changes attributed to the radiation, and the tumor was successfully excised with a negative resection margin. The patient's postoperative course was uneventful.

The resected tumor was 115 × 70 × 60 mm in size. The cross-section surface of the SFT revealed a yellowish-white solid mass with a focal area of necrosis (Fig. 5A). Compared with the pre-radiotherapy specimen, the histological examination revealed decreased cellularity and increased fibrous bands and collagen, with inflammatory

Fig. 3 MRI of pre-radiotherapy. **a** T1-weighted image showed hypointense mass. **b** T2-weighted image showed hypointense mass including hyperintense foci

Fig. 4 Histological findings at the time of diagnostic laparotomy. **a** Histopathological examination of the tumor revealed high vascularity and hypercellularity with spindle-shaped cells arranged in

“patternless pattern.” **b** High mitotic activity (5/10 high-power fields) was observed. **c** Ki-67 was expressed in approximately 15 % of the tumor cells

cells easily identified. A slight reduction of vascularity was observed. Less mitotic activity was also observed (Ki-67 was positive in approximately 10 % of the cellular population) (Fig. 5B). Immunohistochemical examination showed that tumor cells were CD34 (+), CD99 (+), bcl-2 (+), KIT (–), SMA (–), S-100 (–), and epithelial membrane antigen (EMA) (–) (Fig. 5C).

No recurrence of the tumor was confirmed at 26 months of follow-up.

Discussion

SFT was first described by Klemperer and Rabin in 1931 as a primary spindle-cell tumor of the pleura [7]. Studies of pleural SFT were prolific in the 1980s, and the largest study indicated that SFT occurred almost equally in both sexes (195 female, 165 male) and that it occurred in all age groups (range 5–87 years, average 51 years) [5, 8]. The reported tumor size ranged from 1–33 cm (average 11 cm), and the malignant potential was reported to be 12 % [5]. On the other hand, various extrapleural SFTs have been

reported since the 1990s. In 2007, Kawamura et al. [5] analyzed 17 cases of SFT arising in the pelvic cavity from 10 studies. Pelvic SFT occurred in both sexes (7 female, 10 male) and in a patient age range from 23 to 73 years (average 50 years). The tumor size range was 5–22 cm (average 13 cm), and 4 out of 16 cases were designated as malignant (25 %, status of one case was uncertain). From these data, it was concluded that the pelvic SFT had a tendency to be larger, with higher malignant potential than those originating from the pleura. The histological criteria to classify SFT as benign or malignant include more than four mitoses per ten high-power fields, the presence of necrosis, hypercellularity with crowding and overlapping of nuclei and nuclear atypia [9]. In the present case, mitosis were easily identified (5 mitoses per 10 high-power fields), and local necrosis was observed. These were the important diagnostic criteria for malignant SFT.

The histological appearance of SFT may resemble and overlap that of other benign and malignant diagnostic entities, such as hemangiopericytoma, sarcomas (e.g., leiomyosarcoma, synovial sarcoma), gastrointestinal stromal tumor (GIST), inflammatory myofibroblastic tumor, hamartoma,

Fig. 5 A *a* Gross appearance of the SFT. It was a solid, well-circumscribed tumor. *b* Gross appearance of the cut surface. It revealed a *yellowish-white solid mass* with a focal area of necrosis. The tumor was $115 \times 70 \times 60$ mm in size.

B Post-radiation histopathological findings. *a* Compared with the pre-radiotherapy specimen, the histological examination revealed decreased cellularity and increased fibrous bands and collagen, with inflammatory cells easily identified (HE). *b* Compared with the pre-radiotherapy specimen, less mitotic activity was observed. *c* Ki-67 was expressed in approximately 10 % of the tumor cells. **C** Results of immunohistochemical staining ($\times 400$). *a* Tumor cells were positive for CD34. *b* Tumor cells were positive for CD99. *c* Tumor cells were positive for bcl-2. *d* Tumor cells were negative for KIT. *e* Tumor cells were negative for SMA. *f* Tumor cells were negative for S-100. *g* Tumor cells were negative for EMA

and peripheral nerve sheath tumor, among others. Since it lacks typical imaging features, the clinical and radiological diagnosis is usually very difficult. In radiography, an opaque and solitary mass is commonly found [10]. The contrast-enhanced CT reveals an almost homogeneous mass surrounded by a well-defined capsule with progressive

enhancement from the arterial to the venous phase and occasionally multiple small non-enhancing portions [11]. Fine-needle aspiration biopsies rarely provide enough tissue for a definitive diagnosis because of the heterogeneous distribution of cellular density within the tumor [12, 13]. Histologically, the “patternless pattern” is the most

characteristic, but not unique, finding in SFT. Thus, immunohistochemical staining is necessary to rule out other differential diagnoses. Typical immunohistochemical features of SFT are the positive expression of CD34, CD99, and bcl-2 and the negative expression of KIT, SMA, S-100, and EMA. Given that in the present case the surgical specimen revealed cells strongly positive to CD34 antibody and focal CD99 membranous expression, and EMA expression was negative, the diagnosis was in favor of SFT. Moreover, immunohistochemical study with KIT marker did not show any reactivity, excluding the possibility of GIST and hamartoma. The immunohistochemical study also did not show any reactivity for SMA or S-100, eliminating the possibility of leiomyosarcoma and peripheral nerve sheath tumor. Myofibroblastic tumor usually shows focal myxoid stroma and is normally CD34 negative [14]. Hemangiopericytoma is known to have a histopathological and immunohistochemical overlap, and it is questionable if it can be clearly distinguished from SFT. Recently, there has been a tendency to consider SFT and hemangiopericytoma as part of the same entity, with a broad morphological spectrum of differentiation.

SFT is usually a slow-growing tumor with favorable prognosis. The patient is usually asymptomatic until the tumor becomes large enough to compress the neighboring organs [3]. The clinical behavior of SFT is benign. However, it can manifest an aggressive clinical behavior by acquiring the ability to invade locally or recur, and also occasional development of distant metastases has been documented. The most effective therapeutic modality for SFTs is surgical resection. The role of other treatment modalities in the management of SFT is unclear. Some reports in the literature have demonstrated the effectiveness of radiotherapy for the control of SFT. Pelvic radiotherapy (total dose 50 Gy) effectively controlled a pelvic SFT with multiple lung metastases [5], and external thoracic radiotherapy (total dose 60 Gy) reduced a recurrent SFT of the right lung [15]. Also, preoperative radiotherapy (total dose 40 Gy) significantly reduced an intracranial SFT [6]. In the present case, the preoperative pelvic radiotherapy (total dose 50 Gy) successfully reduced the tumor size of a pelvic SFT. On the other hand, some reports showed controversial results. Dietrich et al. [16] reported accelerated tumor progression during the course of radiotherapy in a patient with a thoracic wall SFT. Also, Shiono et al. [2] reported a case of malignant SFT with superior vena cava syndrome that rapidly relapsed following surgery and radiotherapy. Other alternative treatment modalities have also been attempted. The novel targeted drug imatinib mesylate seems to exert some activity on SFT expressing the wild-type platelet-derived growth factor receptor- β : its in vitro [17] and in vivo [18] inhibitory activity in chemo- and radioresistant malignant SFT has recently been demonstrated [11]. Consensus has not been achieved yet

concerning radiotherapy and chemotherapy against SFT, but our present report is one supportive of the effectiveness of preoperative radiotherapy for the safe gross total removal of a large pelvic malignant SFT.

Conclusion

We experienced a rare case of hypervascular large malignant SFT in the mesentery of the rectum, which showed a good response to preoperative radiation therapy. This case strongly supports the potential effectiveness of preoperative radiotherapy for the safe gross total removal of a large pelvic SFT, and it should be carefully considered as a therapeutic modality.

Conflict of interest The authors declare that they have no conflict of interest.

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