

Case Report

Mesenteric Myxofibrosarcoma: A Case Report

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Abstract

Myxofibrosarcoma is a soft tissue sarcoma that occurs in elderly patients. Primary myxofibrosarcoma rarely arises in the mesentery; this is the fourth known case of myxofibrosarcoma presenting as a mesenteric tumor. A 62-year-old male with a mesenteric myxofibrosarcoma presented with an abdominal mass; his symptoms were frequent urination and a sense of abdominal pressure. He was admitted for further examination. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a mesenteric lesion. The preoperative diagnosis was a suspected malignant myxoid tumor. We performed a curative resection with wide margins. The histopathological and immunohistochemical findings confirmed that the tumor was mesenteric myxofibrosarcoma. The postoperative course was uneventful, and there have been no signs of relapse for three years to date after surgery. It is difficult to make a definitive diagnosis of mesenteric myxofibrosarcoma using only CT or MRI. However, when the preoperative findings suggest a malignant mesenteric tumor, then the best practice is resection with sufficient margins.

Keywords

myxofibrosarcoma, mesentery, tumor

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Introduction

Myxofibrosarcoma is a common soft tissue sarcoma that occurs mostly in elderly patients and in the lower extremities. Primary myxofibrosarcoma rarely arises in the mesentery. A preoperative diagnosis is difficult to make using only computed tomography (CT) and magnetic resonance imaging (MRI), and the first-choice treatment is curative resection with wide adequate margins. We report a rare case involving a 62-year-old male with mesenteric myxofibrosarcoma. To the best of our knowledge, this is only the fourth known case in the literature of a myxofibrosarcoma presenting as a mesenteric tumor.

Case Report

A 62-year-old male, who was referred to us for examination, presented with an abdominal mass that had increased considerably in size in the past month. His symptoms included frequent urination with a sense of abdominal pressure. He reported no other notable changes in his health condition. There was a palpable, solid, mobile, and non-tender mass in the lower abdomen. Ultrasound showed an ill-defined heterogeneous mass in the abdomen. Enhanced CT revealed ascites and multiple heterogeneous hyperdense masses, measuring 11 cm in greatest dimension, which appeared to arise from the mesentery. There was no clear evidence of any abnormality. T1-weighted and T1-weighted

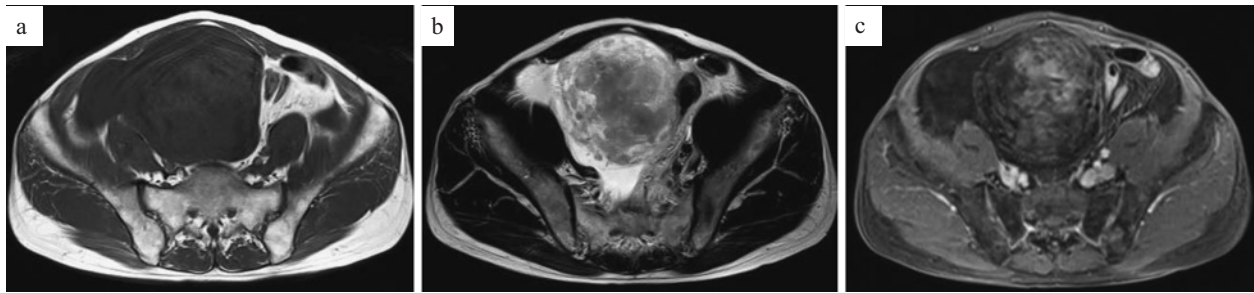


Figure 1. Magnetic resonance imaging. a T1-weighted image revealed a tumor with a clear border. b T2-weighted imaging revealed a low- and high-signal tumor. c Contrast-enhanced imaging revealed a tumor with a smooth edge.

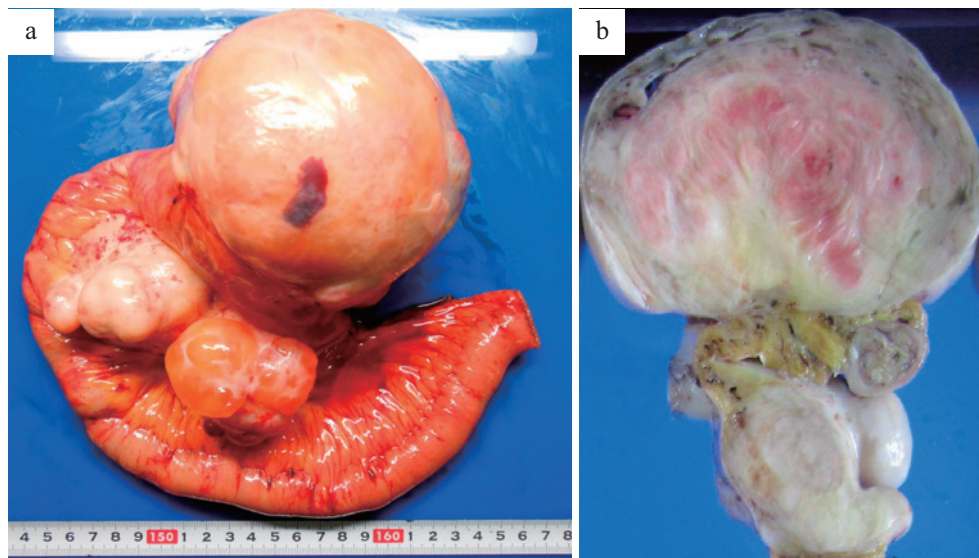


Figure 2. Gross appearance. a The size of the tumor was 11×10 cm. b Split face showed pink and white solid mass.

fat-suppressed images showed a little heterogeneous region (Figure 1a), while T2-weighted images showed a high-intensity heterogeneous region (Figure 1b). The apparent diffusion coefficient was not very low, and contrast-enhanced MRI showed nonuniform contrast (Figure 1c). Together, these findings suggested a possible malignancy, and curative resection was decided. There were extensive serous ascites although cytology indicated no malignancy. The tumor appeared to arise from the mesojejunum and then extend into the abdominal cavity, involving the jejunum. The tumor was resected en bloc with wide margins. The resected specimen showed that the tumor was approximately 11 cm × 10 cm in size (Figure 2). Hematoxylin and eosin staining showed a storiform pattern (Figure 3a), and over 50% of the tumor was composed of myxoid areas (Figure 3b). High cell density and mitotic figures were relatively frequent (Figure 3c). Immunohistochemical examination revealed high Ki67 expression (Figure 4a), and the tumor cells were positive for alcian blue (Figure 4b). The tumor cells were negative for CD34, c-kit, aSMA, and p53. These findings indicated that

the lesion was myxofibrosarcoma. This tumor was T3N0M0 Stage III B according to the American Joint Committee on Cancer system (eight edition). To date, the patient is alive and has not relapsed for three years post-operatively.

Discussion

Myxofibrosarcoma is one of the most common soft tissue sarcomas occurring in elderly patients. It was first described in 1977 as a malignant soft tissue tumor, characterized by a mucoid and nodular appearance, with a coarse plexiform capillary pattern[1]. In 2002, the World Health Organization declassified malignant fibrous histiocytoma as a diagnostic entity and determined that myxoid malignant fibrous histiocytoma without myogenic, lipoblastic and chondrogenic features can be diagnosed as myxofibrosarcoma[2]. Most cases of this tumor occur in the dermis or subcutaneous tissue[3]. It usually presents as a painless, slow-growing mass and occurs mostly on the extremities[4]. Myxofibrosarcoma as a mesenteric tumor is extremely rare. We searched for the

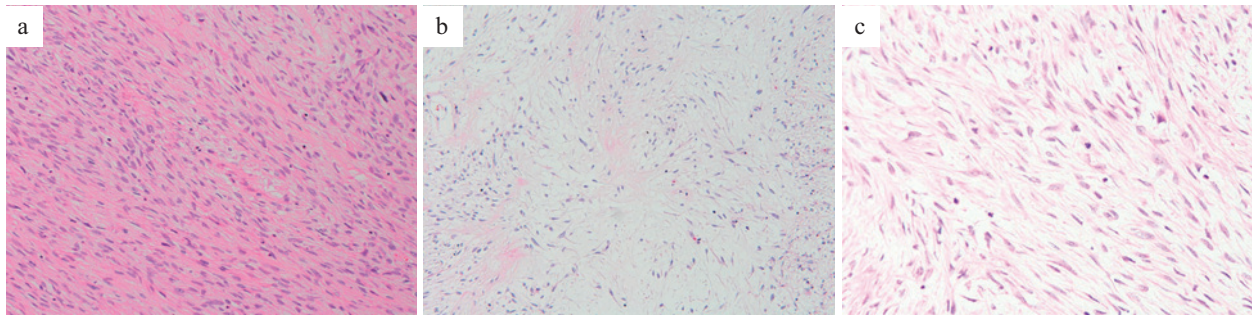


Figure 3. Histopathological findings.

a Microscopic examination showed a storiform pattern (H.E. staining). b Over 50% of the tumor was composed of myxoid areas. c Mitotic figures were present relatively frequently.

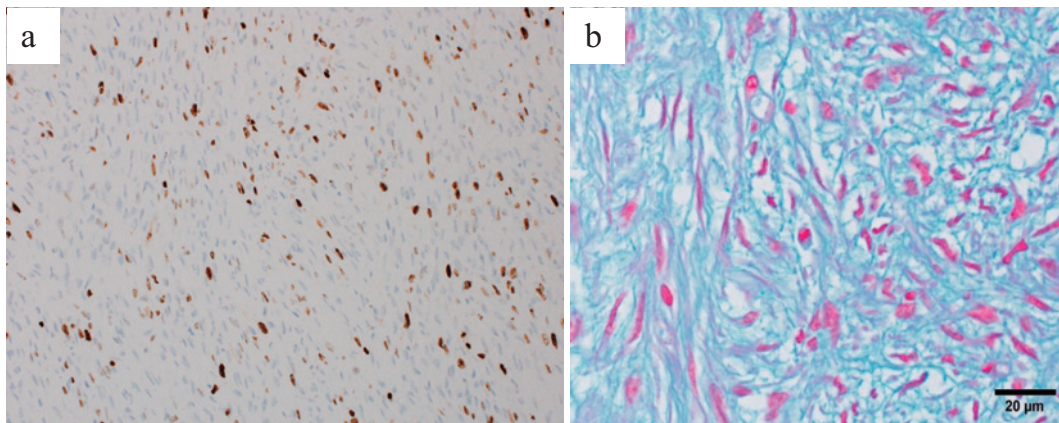


Figure 4. Immunohistochemical findings.

a Ki67 immunostaining was positive. b Alcian-blue immunostaining was positive.

Table 1. Reported Cases of Mesenteric Myxofibrosarcoma.

Authors, Year	Age, Sex	Symptom	Location	Size (cm)	Preoperative diagnosis	Operation	Tumor capsule	Capsule damage	Adjuvant chemotherapy	Re-lapse	Prognosis
Maeura et al., 2000	62, M	Abdominal tumor	Mesocolon	8×7	Retroperitoneal tumor	Sigmoidectomy	+	-	EPI, CDDP, VCR	-	Alive (7Y)
Kadoya et al., 2003	71, F	Abdominal tumor	Mesoileum	13×12	Abdominal tumor	Ileocecal resection	+	-	None	-	Alive
Kitayama et al., 2005	77, F	Loss of appetite	Mesojejunum	15×15	Abdominal tumor	Tumor resection	+	-	None	-	Alive (14M)
This study, 2019	62, M	Abdominal tumor	Mesojejunum	11×10	Malignant mesenteric tumor	Partial jejunectomy	+	-	None	-	Alive (3Y)

keywords “myxofibrosarcoma” and “mesentery” in PubMed and Japan Medical Abstracts Society since 1990. Moreover, we searched for the keywords “malignant fibrous histiocytoma” and “mesentery” and found three cases. That was because myxofibrosarcoma had been classified as a myxoid type of malignant fibrous histiocytoma before 2002. In Japan, we found three cases[5-7] (Table 1); however we did not find reports in literature from other countries.

Myxofibrosarcoma is an asymptomatic disease. In the present case, it was not until the tumor grew to approximately 11 cm that the patient noticed the abdominal mass, with symptoms of frequent urination and a sense of abdominal fullness. All three previous mesenteric myxofibrosarcomas were larger than 8 cm in size. The large size means that wide margins for a safe curative resection are less attainable. Myxofibrosarcoma symptoms depend on the size and occur-

rence site.

It is difficult to make a specific diagnosis of a mesenteric tumor without a biopsy or surgical resection. In all previous cases, mesenteric myxofibrosarcoma could not be diagnosed preoperatively. However, in the present case, the tumor could be diagnosed as malignant by CT and MRI findings.

On CT, there were three tumors. Moreover, the tumors were not connected with any abdominal organ, such as the intestines, and were seen to arise from the mesenterium.

CT revealed no other lesions, such as metastases in the lung, liver, or lymph nodes. Myxofibrosarcoma is often infiltrative, tending to spread along fascial planes in a curvilinear fashion, which is visible on MRI as a curvilinear projection (called a tail sign). Previous studies have suggested that this tail sign was moderately specific and sensitive for myxofibrosarcoma rather than for any other myxoid-containing tumors[8]. Although MRI in the present case did not show the tail sign, it did help determine malignancy through the high-intensity heterogeneous findings on T1-weighted images, suggesting that the tumor consisted of predominantly myxoid content, did not have a fibrous or bleeding nature, and was not a cyst or lipoma. Liposarcoma with necrosis can appear heterogeneous and could not be excluded. The T2-weighted images showed the lesion was heterogeneous, indicating myxoid content. Hemangioma was excluded by these MRI findings. Gastrointestinal stromal tumors and malignant lymphoma, which are common mesenteric tumors, were also excluded because they appear homogeneous on contrast-enhanced MRI. The apparent diffusion coefficient was not very low, suggesting the tumor might have been malignant.

Histologically, MFS features a multinodular growth pattern of spindle to polygonal sarcoma cells within variably myxoid stroma containing long curvilinear vessels[9]. Myxofibrosarcoma is distinct from other similarly named tumors, such as myxoid pleomorphic sarcoma and fibromyxoid sarcoma. Weiss et al. reported that a myxofibrosarcoma definition required at least 50% of the tumor to be composed of myxoid areas[10]. In addition to myxoid findings, the histopathology in the present case showed typical cellular fascicles of spindle cells with numerous and often atypical mitoses. In addition, microscopic examination showed mild pleomorphism. Immunohistochemical examination revealed the present tumor cells were positive for alcian blue, indicating the production of mucin, which is consistent with myxofibrosarcoma. Myxofibrosarcoma commonly shows strong and diffuse staining for vimentin, as found in the present case. Moreover, the resected lesion was negative for c-kit, aSMA, MDM2 and CDK4, thereby excluding gastrointestinal stromal tumors and liposarcoma. Therefore, the postoperative findings confirmed the differential diagnosis as high-grade myxofibrosarcoma because high cell density and mitotic figures were relatively frequent (Figure 3c). In addition,

Ki67 expression was high (Figure 4a).

The optimum strategy for treating primary myxofibrosarcoma is surgery. Wide local excision is the most recommended strategy for mesenteric myxofibrosarcoma, and Clarke urged a safe margin of at least 2 cm[11]. Moreover, the previous mesenteric myxofibrosarcomas had capsules. In the case of mesenteric myxofibrosarcoma, it is also important that we should not damage the capsule to prevent local recurrence. The present case and three previous patients with mesenteric myxofibrosarcomas are currently alive (to the best of our knowledge). It may be partly because they were resected with adequate margins.

Mentzel et al. reported that the incidence rate of local recurrence after surgery for myxofibrosarcoma was 55% and that of distant metastasis was 33%[3]. A few reports suggested lymph node metastasis in myxofibrosarcoma, so follow-up CT should be performed every three months. There are few effective chemotherapeutic regimens for relapsed myxofibrosarcoma; the standard treatment is re-excision of the relapsed myxofibrosarcoma, where possible. If it is inoperable, standard regimen of chemotherapy for myxofibrosarcoma is doxorubicin[12]. Fletcher et al. reported that the overall five-year survival rate after surgery for myxofibrosarcoma was 60%-70%[2]. Zumarraga et al. reported that any local recurrence increased the potential for distant metastasis, and overall survival was significantly correlated with positive surgical margins, local recurrence, and distant metastasis[13]. Lin et al. reported that distant metastasis-free survival after myxofibrosarcoma resection was associated with mitotic activity and margin status[14]. These reports indicated the need to ensure wide safe margins with en bloc resection as a curative treatment to achieve the best prognosis.

In conclusion, it is difficult to make a definitive diagnosis of mesenteric myxofibrosarcoma using only CT or MRI. However, when preoperative findings suggest a malignant mesenteric tumor, then the best course involves resection with sufficient margins.

Conflicts of Interest

There are no conflicts of interest.

Author Contributions

Kunihiro Ozaki is the first author and prepared the manuscript under the supervision of Fumihiko Fujita and Yoshito Akagi. Fumiki Koga, Ichitaro Shiratsuchi, Shintaro Yokoyama, Koichi Yoshiyama, Yutaka Nishimura, and Ryoza Hayashida participated in the data analysis. Takuya Furuta provided the pathologic data. All authors read and approved the final manuscript.

Informed Consent

The patient provided informed consent for this case re-

port.

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