Primary Malignant Fibrous Histiocytoma of the Mesentery: A Case Report

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A malignant fibrous histiocytoma of the mesentery is rare, and multifocal involvement as a primary tumor is very rare. In this report, a case of malignant fibrous histiocytoma of the mesentery presenting with two masses and multiple peritoneal seeding in a 48-year-old man is described. A physical examination revealed a large, firm, and non-tender mass in the right lower abdomen of the patient. Computed tomography of the lesion revealed a partially, indistinctly marginated and heterogeneously enhancing mass with irregular peritumoral strands in the mesentery of the right lower abdomen, while sonograms of the lesion revealed an ill-defined low-echoic mass. The final pathology demonstrated the presence of a storiform-pleomorphic malignant fibrous histiocytoma.

Index words: Mesentery
Malignant fibrous histiocytoma
Ultrasonography
Computed tomography (CT)

Case Report

A 48-year-old man visited our clinic because of a painless palpable mass in the right lower abdomen. The patient detected the mass two weeks prior to the clinic visit. The patient had no history of trauma or any previous surgery. A physical examination revealed the presence of a large, firm mass in the right lower abdomen. The level of serum carbohydrate antigen 19-9 was elevated, but the other laboratory findings were normal. An initial abdominal CT scan showed an approximate 4.5 cm sized heterogeneously enhancing mass with a partially indistinct margin and irregular peritumoral strands in the mesentery of the right lower abdomen [Fig. 1A]. This mass was causing right hydronephrosis. Sonograms showed the presence of an ill-defined low-echoic mass. The presumptive diagnoses in-
Fig. 1. A 48-year-old man with a palpable mass in the right lower abdomen.

A. Computed tomography shows a partially indistinct and heterogeneously enhancing mass with irregular peritumoral strands (white arrows) in the mesentery of the right lower abdomen.

B, C. Three months later, follow-up computed tomography shows an increased size of the known mass (white arrows) in the mesentery of the right lower abdomen and another mass (black arrows) with a similar nature in the mesentery of the upper abdomen.

D. A microphotograph shows mononuclear histiocytie-like cells and multinucleated giant cells (H & E staining, × 200).

E. Immunohistochemical staining for smooth muscle actin shows a positive reaction in the histiocytie-like cells (smooth muscle actin, × 200).

F. Microphotographs show irregular peritumoral strands including infiltrating tumor cells and desmoplastic reaction in the mesentery (H & E staining, × 200).
cluded a variety of tumor or tumor-like conditions of the mesentery causing a desmoplastic reaction, such as a metastatic tumor, carcinoid tumor and sclerosing mesenteritis. An ultrasound-guided aspiration was performed, and the possibility of a benign spindle cell tumor was suggested. Three days later, a laparoscopic biopsy was performed, and the pathologist suggested the possibility of fibromatosis or a gastrointestinal stromal tumor. A complete excision of the mass was recommended for an accurate diagnosis. An excisional biopsy was scheduled, but the biopsy was delayed. Two months later, the abdominal pain and distension became aggravated. A follow-up abdominal CT scan showed an increase in the size of the known mass in the mesentery of the right lower abdomen, and demonstrated the appearance of a new similar mass in the mesentery of the upper abdomen. In retrospect, the initial CT images revealed the presence of multiple mesenteric nodules (Fig. 1B, C). The multiple small, enhancing nodules in the mesentery and the omentum had grown within the two-month period. An extended right hemicolectomy with jejunal resection was performed for palliative treatment. Macroscopically, the surgical specimen contained two ill-defined serosal-intramuscular masses in the mesentery of the jejunum and terminal ileum. Neither necrosis nor hemorrhage was present in the gross specimen. The remaining serosa showed multiple mesenteric nodules, measuring from 0.5 cm to 1.5 cm in the longest dimension. A histopathological examination revealed some areas of a storiform pattern, high cellularity, and marked nuclear pleomorphism with multinucleated giant cells in the tumors of the mesentery (Fig. 1D). Immunohistochemical staining was positive for expression of smooth muscle actin and negative for expression of CD34, C-kit, S-100 protein, desmin, and cytokeratin. These findings were suggestive of a storiform-pleomorphic malignant histiocytoma (Fig. 1E).

Five months have elapsed since the surgery, and the patient is doing relatively well without any evident tumor recurrence or distant metastasis.

Discussion

Primary tumors arising in the mesentery are rare, but the mesentery is a frequent route for the spread of a malignant neoplasm through the peritoneal cavity and between the peritoneal space and the retroperitoneum (5). Most primary lesions are mesenchymal in origin, and most are histologically benign. Malignant mesenchymal lesions include liposarcomas, leiomyosarcomas, malignant schwannomas, synovial sarcomas, fibrosarcomas and MFHs. Unfortunately, with the exception of a minority of lesions, such as lipomas, hemangiomas and subacute hematomas, the radiological appearance of most soft-tissue masses remains nonspecific (1).

An MFH in the abdominal cavity represents 16% of all MFHs, and most of these lesions are located in the retroperitoneum. If secondary involvement of the abdominal cavity can be ruled out, the presence of an intra-abdominal, extra-retroperitoneal MFH could be considered even though multifocal involvement is very rare. The anatomic distribution of an intra-abdominal, extra-retroperitoneal MFH is as follows: the stomach, the small intestine, the colon, the appendix, the mesentery, the liver, and the spleen (1-3).

The pathological diagnosis of MFH can be inaccurate if the whole mass cannot be excised as it is composed of variably proportioned histiocytic and fibrous elements. The complete excision of the mass is therefore recommended for an accurate diagnosis.

In general, on a CT scan, MFH appears as a relatively well-defined and homogenous mass, or a mass with internal low attenuation due to necrosis or hemorrhage. The frequency of internal necrosis or hemorrhage increases in proportion to the mass size (6). Lee et al. reported 13 masses in the abdomen out of seven cases that were pathologically confirmed to be MFHs; eight of the lesions (62%) had a peritumoral-vascular-appearing structure. Previous reports had not mentioned this finding (4), and Lee and colleagues first reported this finding but there was no pathological confirmation of the presence of a peritumoral-vascular-appearing structure.

For MFHs, sonography demonstrates the presence of three patterns, including a hypoechoic pattern (the most frequent), a mixed pattern with an extensive necrotic area, and a predominantly anechoic pattern with thick septa (6).

On a CT scan, the MFH appeared as an irregularly marginated, heterogeneously enhancing mass with irregular peritumoral strands in the mesentery, and as an ill-defined low-echoic mass on the ultrasonogram. The irregular peritumoral strands in this case are similar to the peritumoral-vascular-appearing structure that Lee and colleagues have reported. When these findings were correlated with the pathological findings, they mainly a desmoplastic reaction that included infiltrating tumor cells (Fig. 1B, F).

The differential diagnoses of soft-tissue masses with ir-
regular peritumoral strands in the mesentery include sclerosing mesenteritis, mesenteric edema or hemorrhage, mesenteric inflammation secondary to pancreatitis, fibrofatty mesenteric proliferation related to Crohn’s disease, primary mesenteric neoplasms (e.g., a desmoid or carcinoid tumor), peritoneal mesotheliomas, and metastatic neoplasms [5, 7]. Although rare, a MFH should be included in these differential diagnoses.

In general, an extra-retroperitoneal abdominal MFH recurs in 37-51% of all cases and metastases occur in 42% of all cases, essentially in the lungs and lymph nodes. The overall two-year and five-year survival rates for patients afflicted with MFH have been reported to be about 60% and 47%, respectively [2]. The prognosis of an MFH is unclear, but most of the reports suggested that it is poor. One group of investigators reported that postoperative chemotherapy increased the survival rate, but another group of investigators has reported that postoperative chemotherapy did not achieve curative results. There is a report stating that the tumor was not radiosensitive. However, it is clear that complete surgical excision in the early phase is essential to improve the prognosis, although the role of adjuvant chemotherapy after surgery is still unclear [8-10].

In the current case, surgical resection was delayed as the ultrasound-guided aspiration and laparoscopic biopsy suggested that the tumor was benign. Unfortunately, this outcome might have influenced the prognosis of the patient.

In conclusion, if CT scan images reveal a large soft-tissue mass with irregular peritumoral strands in the mesentery, especially in a middle-aged adult, an MFH must be included in the differential diagnoses, along with other benign or malignant disorders. Furthermore, for an accurate diagnosis of an MFH, the whole mass must be excised.

References

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