CASE REPORT

Low-Grade Myxofibrosarcoma of the Rectus Abdominis Muscle Infiltrating into Abdominal Cavity: A Case Report

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Objective: Myxofibrosarcoma (MFS) is a relatively rare tumor that is histologically characterized by myxoid stroma and spindle cell proliferation. This tumor most commonly arises as a slow growing, enlarging painless mass in the extremities of elderly patients. Methods: We report a case of a primary, low-grade MFS in the rectus abdominis muscle infiltrating the abdominal cavity of a 75-year-old man. Results: The patient underwent a wide excision of the right abdominal wall mass with a 3-cm surgical margin from the scar due to a biopsy. The tumor infiltrated the urinary bladder, peritoneum, and external iliac vessels. Twenty-six months after the initial operation, he had recurrences in his abdominal wall, urinary bladder, and right iliac vessels. Conclusions: To our knowledge, primary MFS of the muscle in the abdomen has not been documented previously. Although this case was histopathologically classified as a low-grade tumor, it infiltrated the abdominal cavity. The tumor is suspected to have penetrated the abdominal cavity below the linea arcuata, which lacks the posterior sheath of the rectus abdominis muscle; from there, it could easily spread without being blocked by any biological barriers.
of the muscles in the abdomen has not been previously documented. We report a case of a primary low-grade MFS in the rectus abdominis muscle invading the abdominal cavity of a 75-year-old man.

METHODS

A 75-year-old man experienced an induration with pain on his lower abdominal wall. Since the tumor increased gradually, he consulted a plastic surgeon. Magnetic resonance imaging revealed a tumor in his right rectus abdominis muscle. Magnetic resonance imaging showed a heterogeneously contrast-enhancing mass involving his right rectus abdominis muscle (Fig 1). The patient underwent a biopsy at the Department of Orthopaedic Surgery in our hospital. Low-grade MFS was diagnosed histopathologically and the patient was referred to our department for further management including reconstructive surgery. No other lesions were detected on whole-body fluorodeoxyglucose-positron emission tomography (PET) scanning. At this point, there was no apparent evidence whether the tumor invaded into the abdominal cavity or not.

Figure 1. (a). Axial section of gadolinium-enhanced T1-weighted magnetic resonance imaging. The gross tumor was located in the right rectus abdominis muscle and spread horizontally. (b). Sagittal section of T2-weighted magnetic resonance imaging. The abdominal tumor did not spread into the abdominal cavity. The yellow arrow indicates the urinary bladder.
The patient underwent a wide excision of the right abdominal wall mass with a 3-cm surgical margin from the scar due to a biopsy. The tumor infiltrated the urinary bladder, so we shaved the surface of the muscle layer of the bladder (Fig 2). However, the firm and fibrous mass was infiltrating the peritoneum and external iliac vessels. As the margin was described as positive or involved in intraoperative rapid diagnosis, further excision was performed. Consequently, the entire right rectus abdominis muscle along with the overlaying skin and the subcutaneous tissue, part of the bladder muscle, the right half of the posterior peritoneum, and the adventitia of the external iliac vessels. We attached Seprafilm (Kaken Pharmaceutical Co, Ltd, Tokyo, Japan) to the defect of the posterior peritoneum. We reconstructed the abdominal wall with Composix E/X Mesh (Davol, A Bard Company, Rhode Island).

RESULTS

Postoperatively, the patient had a paralytic ileus, which resolved after decompression with an ileus tube. The gross specimen measured $10 \times 4 \times 4$ cm, and the lesion within the specimen was semitranslucent, gelatin-like, and with hemorrhage. Histologic examination showed a multinodular lesion with a prominent myxoid matrix. A small number of irregular,
spindle-shaped, or stellate cells, with hyperchromatic nuclei, proliferated in the stroma (Fig 3). According to the modified French Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) grading system, the tumor in this case scored 3 (tumor differentiation; score 2, mitotic count; score 1, tumor necrosis; score 0), that is, grade 1. Twenty-six months after the initial operation, a recurrence was prominent in his right abdominal wall, urinary bladder, and the right iliac vessels. Considering the patient’s physical condition, it was very difficult for our multidisciplinary medical team to come to a consensus regarding ablative surgery including radical cystectomy and removal of the right external iliac vessels. During chemotherapy with doxorubicin, he complained of nausea and fatigue. At that point, he opted for the best supportive care without further medical intervention.

**DISCUSSION**

Myxofibrosarcoma is one of the most common soft tissue sarcomas in the elderly and occurs mostly on extremities. It was first described in 1977 as a distinct soft tissue sarcoma characterized by a mucoid and nodular appearance. Most cases of this tumor occur in the dermis or the subcutaneous tissue. It usually presents as a painless, slow-growing mass. Currently, the World Health Organization classifies MFS as a fibroblastic/myofibroblastic malignant tumor with specific clinicopathological characteristics. Histopathological features show multinodular growth with incomplete fibrous septa and a myxoid stroma composed of
Hyaluronic acid. In low-grade lesions, mitotic figures are infrequent. The present case had a multinodular lesion with a prominent myxoid matrix and slightly increased mitotic activity on light microscopic examination. A small number of irregular, spindle-shaped, or stellate cells, with hyperchromatic nuclei, proliferated in the stroma. This case was diagnosed as a malignant soft tissue tumor, MFS. In FNCLCC grading system, the tumor scored 3 (tumor differentiation; score 2, mitotic count; score 1, tumor necrosis; score 0), that was grade 1 (low-grade) disease.

In the present case, this large tumor might have originally been located in the rectus abdominis muscle. It extended into the abdominal cavity, involving the urinary bladder and the right half side of the peritoneum. To our knowledge, MFS in the skeletal muscle of the trunk is rare, with few cases of thoracic lesions being reported previously. There are only 2 previously reported cases of MFS of the retroperitoneum from Japan. Moreover, there have been no reports in the English literature describing cases similar to our case, with infiltrative growth into the abdominal cavity. In this case, the tumor penetrated the abdominal cavity below the linea arcuata, which lacks the posterior sheath of rectus abdominis muscle; from there, the tumor could easily further infiltrate surrounding areas without being blocked by biological barriers.
Potential therapeutic options for MFS have been reported, including wide local excision, chemotherapy, and radiation. Wide local excision is the most recommended strategy for low-grade MFS. The local recurrence rate in patients with MFS after resection is 16% to 54%. Patients with close surgical margins should be considered for radiation therapy, and recurring masses should be widely excised, including the scar from the previous surgery. In the present case, owing to the wide defect of the abdominal wall after wide resection, we had to reconstruct the abdominal wall, using artificial mesh material. Despite the possibility of an inadequate excision, we could not administer postoperative radiation in order to prevent complications, such as an adhesive intestinal obstruction or exposure of the mesh. Chemotherapy with doxorubicin had been suspended because of side effects. Consequently, we could not perform curative treatment for this recurrent mass. Palliative therapy to increase the patient’s comfort was important.

CONCLUSION

We presented a rare case of a large tumor with clinical and histopathological features of low-grade MFS in the rectus abdominis muscle with recurrence. The tumor was suspected to have penetrated the abdominal cavity below the linea arcuata, which lacks the posterior sheath of the rectus abdominis muscle; from there, the tumor could easily infiltrate the surrounding areas without being blocked by biological barriers.
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