

A Low-grade Myofibroblastic Sarcoma in the Abdominal Cavity

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Abstract. *Low-grade myofibroblastic sarcoma (LGMFS) is a fusiform cell tumor which develops in bone or soft tissues. This type of tumor frequently occurs in the oral cavity and extremities, while it is extremely rarely found in the abdominal cavity. This article reports a case of LGMFS exceeding 20 cm in diameter in the abdominal cavity observed in a 65-year-old male patient. The patient visited our hospital complaining of a heavy feeling of the stomach and abdominal distension. Imaging examinations revealed a giant solid tumor in the abdomen, and surgical treatment was scheduled. During the operation, a tumor about 20 cm in diameter with its anterior aspect covered with the greater omentum was found. The tumor had firm adhesions to the surrounding tissues, and it was excised with concomitant resections of the tail of the pancreas and the spleen. Histopathologically, fusiform cells were arranged in a complicated or storiform pattern, and immunohistochemical staining revealed that the tumor was positive for α -smooth muscle actin, negative for S100 β , H-caldesmon and c-KIT, and a diagnosis of LGMFS was made.*

Low-grade myofibroblastic sarcoma (LGMFS) is an uncommon tumor which develops mainly in the bone or soft tissues of the head and neck region, trunk, or extremities (1, 2). Such tumor is extremely rarely found in the abdominal cavity, and there have been only a few cases of LGMFS reported worldwide (1-3).

We recently encountered a giant LGMFS exceeding 20 cm in diameter which had developed in the abdominal cavity, and which was surgically resected with gratifying results.

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Case Report

A 65-year-old male patient visited our hospital with chief complaints of a heavy feeling of the stomach and abdominal distension. The patient's past history was unremarkable. These complaints had been observed since around January 2009, but the patient had not sought medical advice at that time. In January 2010, the patient underwent a medical checkup including upper gastrointestinal radiology, which revealed deformity of the gastric antrum. The patient was referred to the outpatient service of the Department of Internal Medicine of our hospital. A diagnosis of a primary giant solid tumor in the abdomen was made, and in April 2009, the patient was admitted to our department for surgical treatment.

On admission, the patient had a height of 160 cm and a body weight of 60 kg. His body temperature was 36.6°C; blood pressure, 140/90 mmHg; and his pulse rate with regular at 72 beats/min. A non-tender mass was palpable in the region from the epigastrium to the umbilicus and left flank.

Hematological and blood biochemical tests on admission showed no abnormal findings, and serum levels of tumor markers were within the normal limits: carcinoembryonic antigen (CEA), 1.9 ng/ml; and carbohydrate antigen (CA19-9), 17.0 U/ml.

Abdominal computed tomography (CT) disclosed a low-density, solid tumor measuring 22×18 cm, adjacent to the cardiac part of the stomach, duodenum and pancreas. Well-developed nourishing blood vessels were visualized in the tumor on early enhanced phase imaging (Figure 1). The image enhancement effect spread centrally over time, and a late phase image demonstrated a whorl-shaped blush in the entire tumor. A CT angiographic scan showed well-developed tumor blood vessels and a rightward deviation of the left gastric artery (Figure 2).

Abdominal magnetic resonance imaging (MRI) revealed a 22×18×9 cm tumor with a smooth rim, showing high intensity regions on the T2-weighted image (Figure 3), and low intensity areas on the T1-weighted image, situated on the dorsal aspect

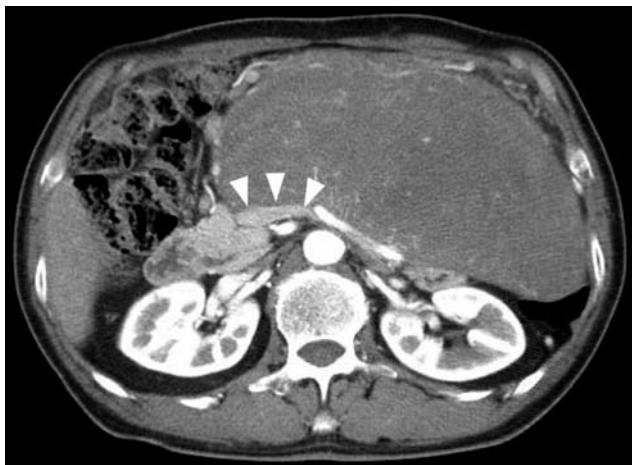


Figure 1. Computed tomography (CT) showing a tumor (arrowheads) in contact with the anterior surface of the pancreas. The major axis of the tumor is about 22 cm.

of the gastric body and the ventral aspect of the pancreatic head. There was relatively homogenous image enhancement inside the tumor. The tumor was situated in contact with the pancreas, spleen and stomach, yet with no obvious infiltrations.

Upper gastrointestinal endoscopy revealed no abnormalities on the mucosal surfaces of the stomach or duodenum but disclosed inward displacement of the anterior gastric wall by the mass outside the wall.

A diagnosis of intra-abdominal giant tumor was made, and surgical resection of the tumor was scheduled.

Upon laparotomy by a midline incision, a smooth-surfaced, huge tumor exceeding 20 cm in diameter was noted within the omental sac. There was neither ascites nor peritoneal dissemination. The anterior aspect of the tumor was covered with the greater omentum, and the dorsal aspect showed an extensive adhesion, primarily to the tail of the pancreas. The superior aspect of the tumor was adhered to the posterior aspect of the stomach, and it was possible to detach the tumor from the stomach. Since the dorsal aspect of the tumor was firmly adhered to the tail of the pancreas, the tail of the pancreas and the spleen were resected concurrently. Furthermore, there were firm adhesions around the common hepatic artery and it was difficult to detach the left gastric artery; therefore, these arteries were ligated and severed at their roots, to complete the resection of the giant tumor. The estimated intraoperative blood loss was 5,500 ml.

The resected tumor specimen measured 215×174×105 mm and weighed 2,528 g. The tumor was capsulated. Sections through the tumor revealed solid, grayish-white tissues, showing relatively homogenous internal structures (Figure 4).

Histopathological examination revealed that the anterior aspect of the tumor was virtually entirely capsulated, while the posterior aspect showed a slight infiltration into the pancreatic

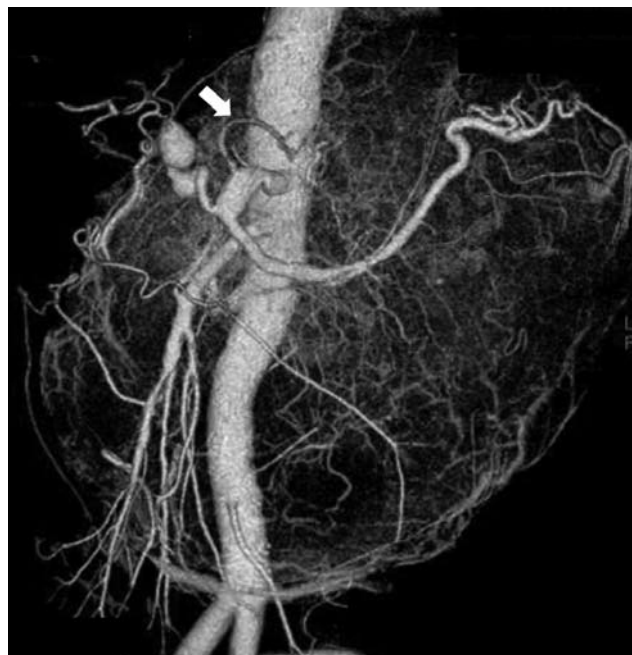


Figure 2. CT angiography showing developed tumor vessels and a rightward deviation of the left gastric artery (arrow).

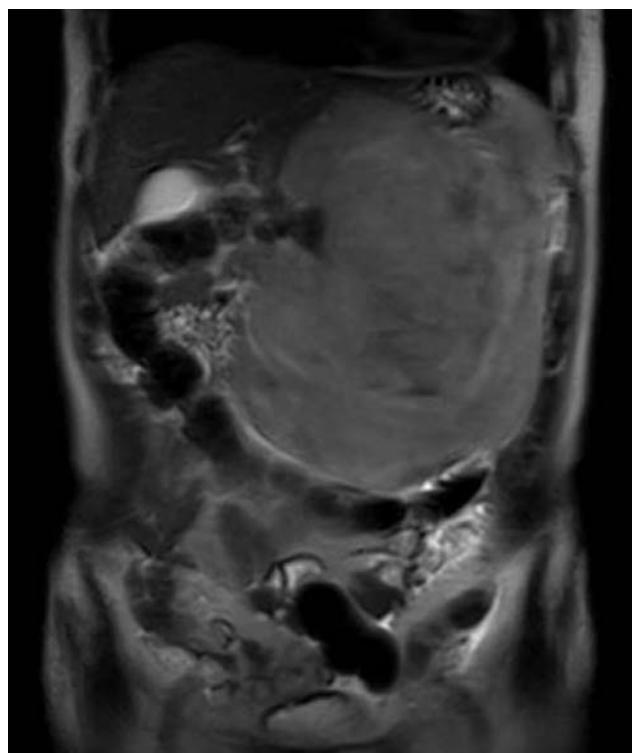


Figure 3. Magnetic resonance imaging (MRI) showing a tumor with smooth limbs. Internal construction of the tumor was relatively smooth, showing high intensity regions on the T2-weighted image.

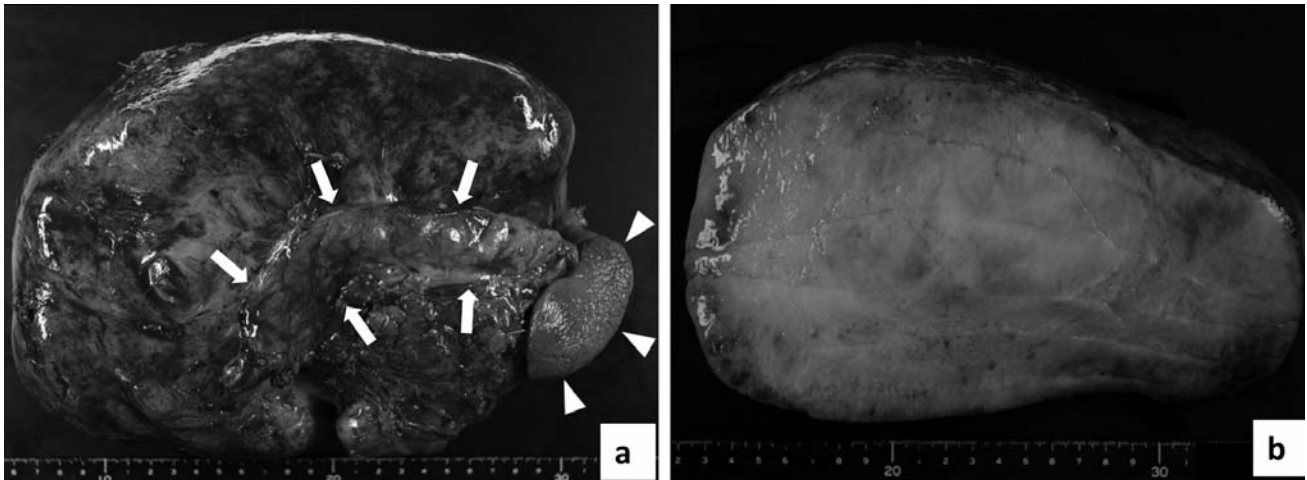


Figure 4. *a*: The gross specimen comprised the tumor, the body and tail of the pancreas (arrows) and spleen (arrowheads). The tumor size was 215×174×105 mm. *b*: The section revealed grayish-white and uniformly structured tissues.

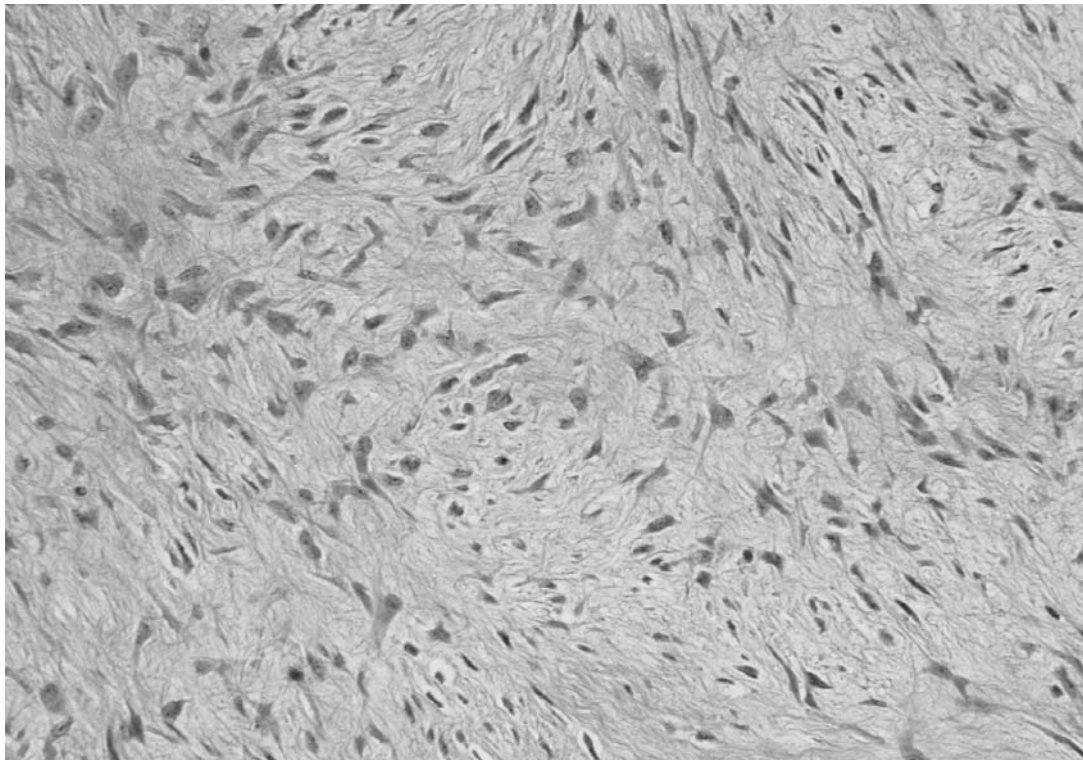


Figure 5. *Histopathological findings*. Tumor cells were fusiform, arranged in sheet-like or storiform patterns, showing proliferation with a moderate cell density (HE, ×200).

tail. Tumor cells were fusiform, containing a constricted slender nucleus and eosinophilic and fibrous cytoplasm. The cells were arranged in sheet-like, complicated or, in part, storiform patterns, showing growth with a moderate cell density (Figure 5). Mitotic figures were only sparsely seen. In

the interstice, there was an admixture of eosinophilic, hyalinized collagen fibers with abundant capillaries, but no evidence of necrosis. Immunohistochemical staining disclosed that the tumor tissue was positive for α -smooth muscle actin (α -SMA), negative for S100 β , H-caldesmon and c-KIT

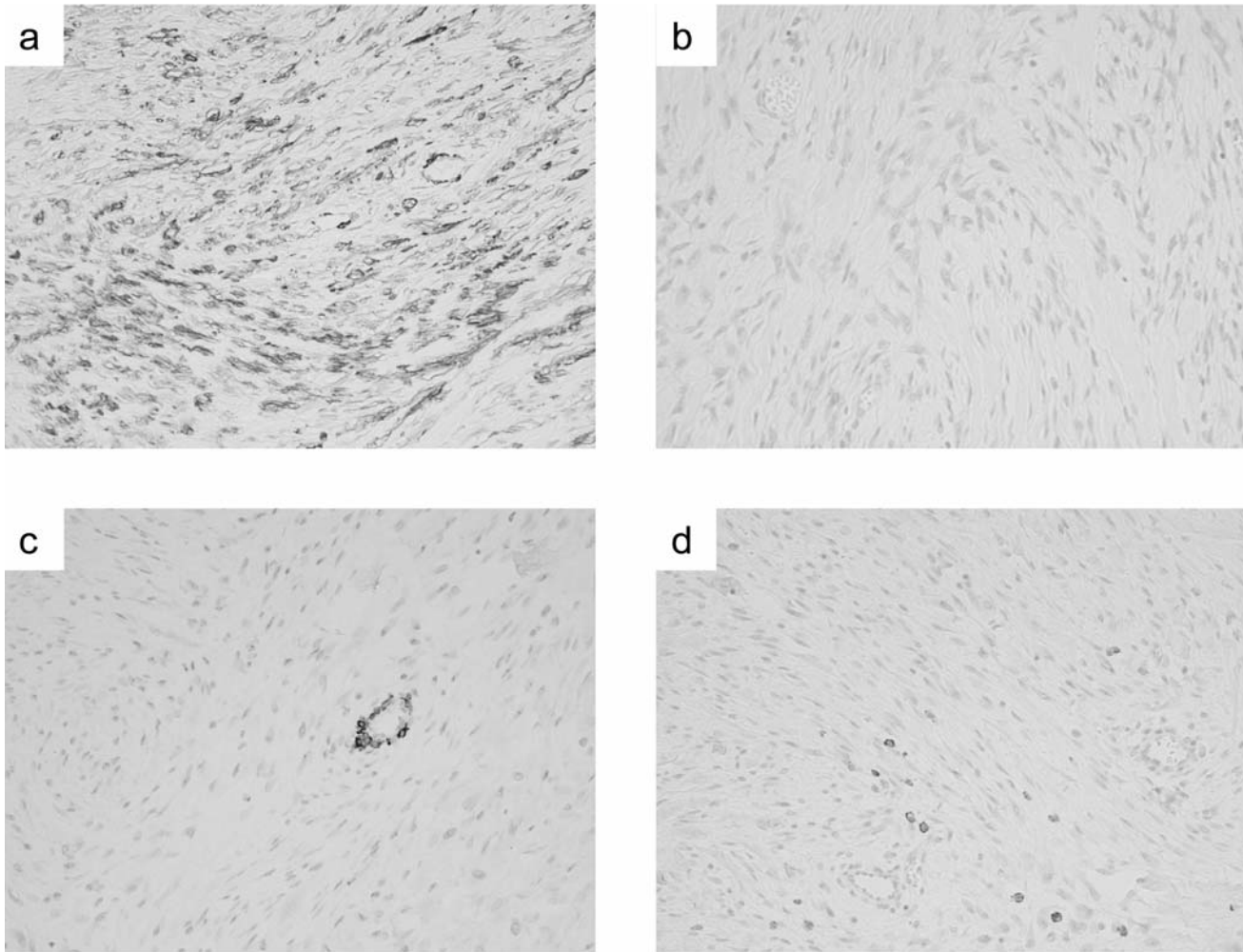


Figure 6. Immunohistochemical findings. a: The cytoplasm of the tumor cells is stained with antibody to α -SMA ($\times 200$). b: The tumor cells are negative for S100 β ($\times 200$). c: Smooth muscles of the vessel are stained; however, the tumor cells did not stain with antibody for H-caldesmon ($\times 200$). d: Mast cells are stained, but the tumor cells are negative for c-KIT ($\times 200$).

(Figure 6), and negative for CD34, β -catenin and epithelial membrane antigen (EMA), with a low MIB-1 index of about 5%. The above findings led to a diagnosis of LGMFS, although the primary neoplastic tissue was unknown.

On account of the massive intraoperative blood loss, the patient was managed in the intensive care unit postoperatively and was returned to the ward with improvement on the second postoperative day. The patient was begun on water by mouth on the fifth postoperative day, and oral food intake was slightly delayed because of hiccup and vomiting due to gastric hypomotility from the seventh postoperative day. The patient improved thereafter and was discharged on the 49th postoperative day. No recurrence has occurred as of 11 months post operation.

Discussion

LGMFS is a rare fusiform cell neoplastic disease that arises in the bone or soft tissues (1, 2). Such tumor may be found in people of all age groups, but is slightly more frequently observed among males. Mentzel *et al.* (1) found this tumor type in 11 male and 7 female patients aged between 10 and 72 years. The oral cavity is the most frequent site of the tumor development, followed in order by the extremities, pelvis, lung, and mammary gland (1, 2, 4, 5), as well as regions such as the salivary gland and perineum though low in frequency (4, 6-8). The tumor is found most commonly in deep soft tissues but may also arise in the subcutis, submucosal tissue, and bone (9). Development of the tumor

Table I. Reported cases of LGMFS in the abdominopelvic cavity.

Case	Author, (ref)	Age, years gender	Site	Size (cm)	Immunohistochemistry	Treatment	Outcome
1	Mentzel <i>et al.</i> , 1998 ⁽¹⁾	72/M	Mesentery of small bowel	17	Desmin (+), α -SMA (-) S100 (-), EMA (-)	Excision	Local recurrence at 4 months, No sign of recurrence at 7 months
2	Mentzel <i>et al.</i> , 1998 ⁽¹⁾	36/F	Omentum	13	Desmin (+), α -SMA (-) CD99 (+) S100 (-), EMA (-)	Excision	No sign of recurrence at 10 months
3	Mentzel <i>et al.</i> , 1998 ⁽¹⁾	36/M	Pelvis	11	Desmin (+), α -SMA (-) S100 (-), EMA (-) Fibronectin (+/-), CD34 (+/-) collagen typeIV(+/-)	Excision	Local recurrence at 20 months, No sign of recurrence at 9 months
4	Mentzel <i>et al.</i> , 1998 ⁽¹⁾	46/F	Pelvic side wall, iliacus muscle, psoas muscle, multiple nodule ileal serosa	5-12	Desmin (+), α -SMA (-) CD34 (+/-) S100 (-), EMA (-)	Partial excision	Not available
5	Montgomery <i>et al.</i> , 2001 ⁽²⁾	73/M	Mesentery	12	α -SMA (+), desmin (+) α -SMA (+), desmin (+)	Excision and radiation therapy	No sign of recurrence at 11 months
6	Agaimy <i>et al.</i> , 2008 ⁽³⁾	70s/F	Mesentery	21	Vimentin (+), α -SMA (+/-)	Excision	Three recurrences at 5, 13 and 25 months, alive with disease
7	Agaimy <i>et al.</i> , 2008 ⁽³⁾	60s/F	Periuterine peritoneum, pelvic peritoneum	3	Vimentin (+), α -SMA (+) Fibronectin (+), CD34 (+/-)	Ruptured intraoperatively	Four recurrences at 10, 19, 25 and 32 months, alive with disease
8	Present case, 2010	65/M	Unknown	21	α -SMA (+), S100 β (-) CD34 (-), β -catenin (-) H-caldesmon (-) c-KIT (-), EMA (-)	Excision with distal pancreatectomy	No sign of recurrence at 11 months

in the abdomen, as seen in our patient, is extremely rare, and no such cases have been reported in Japan, while there have been no more than 7 cases documented throughout the world (Table I). Of these 7 patients, the tumor was found in the greater omentum or mesenterium in 4 and the pelvis in the remaining 3. We failed to identify a tissue of primary development in the present patient based on the preoperative imaging and postoperative histopathological findings.

Indolent enlargement of the mass is a typical clinical manifestation, and the patient may also develop pyrexia, chills and leukocytosis (4, 10). In the present case, the tumor grew without pain and it was not until displacement of the stomach became overt that the symptoms appeared.

The size of the tumor has been reported to be measured at 1.4 to 21 cm in cases in which the tumor development site included the head and neck region or extremities, whereas the lesions were relatively large at 10 cm or more in 6 out of 7 patients with the tumor developing in the abdominal cavity and pelvis (1-3). The present patient also had the lesion measured at 21 cm. This may be due to the fact that tumors growing in the abdominal cavity or pelvis are less palpable through the body surface as compared to those growing in the oral cavity, or other regions such as the extremities, and they are likely to be discovered only after having enlarged.

Histopathologically, LGMFS is characterized by fusiform tumor cells which are arranged in complicated, sheet-like or storiform patterns and show a diffuse, infiltrative growth (1, 2, 9). The cytoplasm is indiscrete and faintly eosinophilic, and the slender, undulate nucleus contains uniformly distributed chromatin and a small nucleolus. Mitotic figures vary in frequency. The interstitial tissue consists of collagenous fibers and is often hyalinized. Inflammatory cell infiltrates are sparse with no necrosis (1, 9, 11). Both the tumor cells and the stroma presented microscopic features characteristic of LGMFS in the present patient.

It is generally recognized that immunohistochemical staining characteristics of this disorder include positive staining for α -SMA, muscle-specific actin (MSA), desmin, calponin and fibronectin, and negativity for laminin, S100 β and EMA (1, 2). An analysis of 18 LGMFS cases reported by Mentzel *et al.* (1) showed that the staining was positive for at least one muscular marker, with positivity rates of 61% for α -SMA, 40% for MSA and 67% for desmin, whereas all cases were negative for laminin. The staining was positive for CD34 and CD99 in 3 each of the 9 patients examined. H-Caldesmon was rarely positive in their study. In the present case, the immunohistochemical staining was negative for S100 β , H-caldesmon, CD34, β -catenin,

c-KIT and EMA; therefore, the possibility of peripheral schwannoma, leiomyoma, solitary fibroma, desmoids, gastrointestinal stromal tumor (GIST), perineurinoma, or synovial sarcoma, among other fusiform cell soft tissue tumors, was ruled out. Thus, as the tumor was positive for α -SMA, a diagnosis of LGMFS was made.

Treatment of this disorder usually consists of surgical resection. Some patients have been treated concomitantly with radiation therapy and chemotherapy (1, 2), but therapeutic effects in these patients have not been documented.

The outcome of LGMFS treatment included local recurrence in 2 out of 11 patients reported by Mentzel *et al.* (1) and in 7 out of 13 patients reported by Montgomery *et al.* (2), when reviewed including tumors of the head/neck and the extremities. The median duration to recurrence varies between these reports, *i.e.*, 29 and 10 months, respectively; hence the tumor recurred roughly 1 to 3 years post operation. When only intra-abdominal and intrapelvic tumors were assessed, local recurrence occurred in 4 out of 7 patients, including one that had recurrence on a total of 4 occasions within 32 months after the first operation (3). The initial recurrence in these patients was observed between 4 and 20 months post operation, and the recurring lesions were resected again. Distant metastases were rare (1, 2) and did not occur in any of the 7 patients with tumor which had developed in the abdominal or pelvic cavity. Of these 7 patients, only one patient, who underwent an incomplete resection at the first intervention, died (1).

The above findings suggest that a favorable prognosis may be attained in LGMFS patients by complete resection of the primary lesion and local recurrent lesions. No recurrence has been noted in our patient; however, any local recurrence should be detected as soon as possible by periodic imaging examinations.

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