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Low-grade fibromyxoid sarcoma of the abdominal wall in an 8-year-old boy

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ABSTRACT

Low-grade fibromyxoid sarcoma (LGFMS) is an extremely rare neoplasm classified as a fibroblastic soft tissue tumor. It has been described by its characteristic deceptively benign histological appearance with metastatic potential during long-term follow-up. We herein describe an 8-year-old boy with LGFMS located in the internal oblique muscle. Histological findings of the resected specimen included contrasting fibrous and myxoid areas with a swirling, whorled growth pattern; bland, benign-appearing fibroblastic spindle cells; and large collagen rosettes. Immunohistologic examination revealed mucin 4 positivity. The surgical margin of the tumor was obscure; therefore, we performed an additional resection 2 months later with a 1 cm margin from the edge of the tumor. No recurrence or metastasis was identified throughout 1 year of follow-up.

Low-grade fibromyxoid sarcoma (LGFMS) is an uncommon deep soft tissue neoplasm usually found in the lower limbs, trunk, upper limbs, buttocks, head, and neck. It typically occurs in young to middle-aged adults, but is sometimes found in children. LGFMS has a deceptively benign histological appearance with malignant potential, and long-term follow-up has indicated high rates of local recurrence and eventual metastasis. We herein report a rare case of LGFMS in the internal oblique muscle of an 8-year-old boy.

1. Case report

An 8-year-old boy had a 2-year history of a rapidly growing abdominal mass in the right abdominal wall. He had no associated pain, distention, rebound, or guarding of the abdomen. The mass measured 4.0 × 3.7 × 3.0 cm, and magnetic resonance imaging showed a low-intensity area on T1-weighted images and a partially high-intensity area on T2-weighted images (Fig. 1). We performed surgical excision of the tumor. The tumor was located in the internal oblique muscle, and its surface was smooth and grayish white (Fig. 2). The postoperative course was uneventful, and the patient was discharged after 7 days.

Histologically, the tumor consisted of contrasting fibrous and myxoid areas with a swirling, whorled growth pattern; bland, benign-appearing fibroblastic spindle cells; and large collagen rosettes (Fig. 3). The surgical margin of the tumor was obscure. Immunohistochemical examination revealed positivity for mucin 4 (MUC4) and epithelial membrane antigen (Fig. 4). The pathological diagnosis was LGFMS.

As the lesion had a marginal excision, 2 months later, we performed an additional wide resection to remove a margin 1 cm from the border of the previous resection. No tumor remnant was found in the additional specimen. No local recurrence or metastasis was identified by lung computed tomography 1 year after surgery.

2. Discussion

LGFMS was first described in 1987 [1] as a deceptively benign-appearing, unclassifiable but very similar fibromyxoid sarcomas characterized histologically by bland, innocuous-appearing fibroblastic cells and a swirling, whorled growth pattern were presented. The reported incidence of LGFMS was

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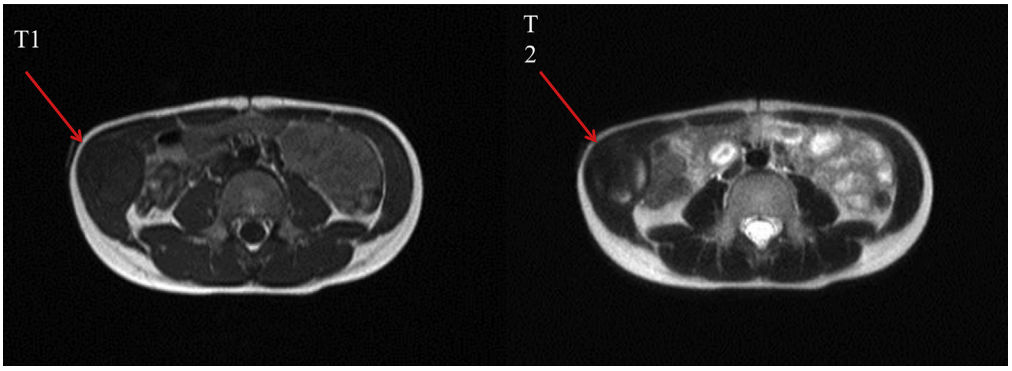

Fig. 1. Magnetic resonance images demonstrate a large tumor exhibiting partial hyperintensity on T2-weighted images and low signal intensity on T1-weighted images.

0.18 per million in western Denmark and comprised 0.6% of all soft tissue sarcomas [2]. Tang et al. [3] reported the largest literature review of 273 cases of LGFMS, including 136 females and 137 males. Notably, the median onset age was 35 years, and 35 cases (13%) occurred in patients aged <18 years. Tumors were commonly located in the lower limbs, trunk, upper limbs, buttocks, head, and neck [3]. High rates of local recurrence and distant metastasis reported by several investigators are shown in Table 1. Their results varied widely; the local recurrence rate ranged from 9% to 73%, and the metastasis rate ranged from 6% to 45%. Folpe et al. [4] considered that earlier studies might have overstated the aggressive potential of LGFMS because of the presence of a larger group of patients with undiagnosed LGFMS who had never had a subsequent adverse event and who therefore had not been identified. It has also been argued that the insufficient length of follow-up has not allowed for complete assessment of the behavior of LGFMS. Local recurrence may reportedly develop up to 15 years later, and distant metastasis may occur 45 years later [8]; however, most investigators have described cases that they followed for <1 year. Furthermore, incompleteness of the excisional margin is a risk factor for recurrence and metastasis. Folpe et al. [4] performed an additional wide excision in 35 cases (69%) when the original surgical margin was not ascertained to be negative, whereas some case series included marginally positive cases without additional treatment. Most LGFMSs are deceptively well circumscribed but not encapsulated, and resection is often incomplete. Complete surgical excision with a safety margin is the mainstay of treatment for patients with LGFMS. The role of radiotherapy and chemotherapy remains controversial.

Histologically, LGFMS consists of bland fibroblasts with a whorled or linear arrangement alternating with less cellular areas

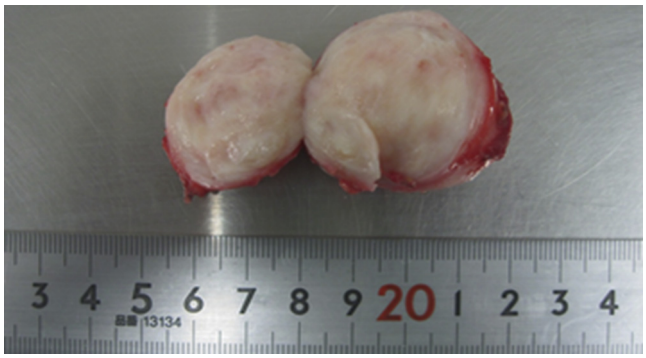

Fig. 2. Macroscopically, the tumor was solid with a grayish-white cut surface.

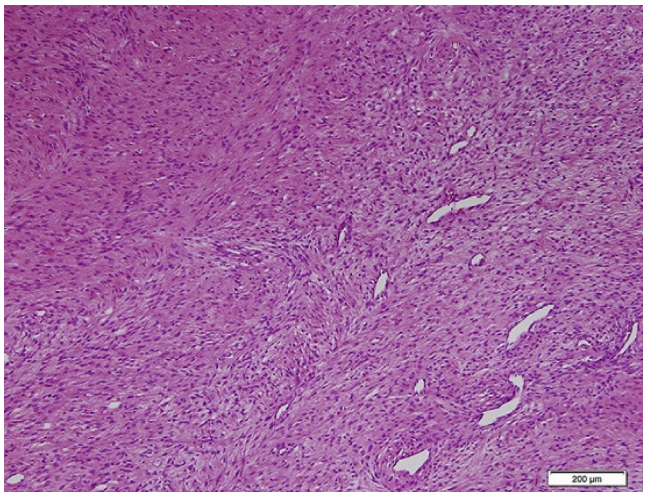

Fig. 3. Microscopic examination revealed a whorled growth pattern with bland spindle-shaped tumor cells.

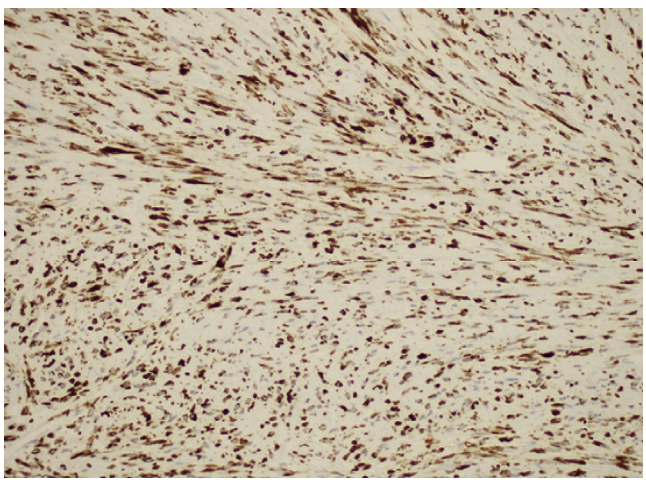

Fig. 4. Tumor cells showed diffuse and strong immunoreactivity for MUC4.

with a myxoid stroma [9]. Approximately 40% of LGFMSs show focal poorly formed collagen rosettes [10]. Diagnosis of LGFMS is less difficult when distinctive hyalinized collagen rosettes are encountered. The differential diagnoses of LGFMS include desmoid-type fibromatosis, nodular fasciitis, soft tissue perineurioma, neurofibroma, dermatofibrosarcoma protuberance, and