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Primary leiomyosarcoma of the abdominal wall mimicking nodular fasciitis in a child

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ABSTRACT

We report the case of an 8-year-old boy with a 30-mm solid mass in the right lower quadrant of the abdominal wall. Computed tomography revealed that the tumor was on the lateral border of the rectus abdominis, and surgical resection was performed. Despite difficulty in differentiating this mass from nodular fasciitis, pathologic analysis and immunohistochemical staining led to the diagnosis of leiomyosarcoma.

1. Case report

An 8-year-old boy with an unremarkable medical history presented to our surgical service after noting the development of a hard indolent mass in the right lower quadrant of the abdomen over the course of a month. Detailed history taking revealed that he had a trauma history of a right lower abdominal bruise sustained a few weeks before noticing the mass. On presentation, a slight elevation was observed at the right lower lateral border of the rectus abdominis, and the smooth, round-shaped, nontender mass, measuring 30 mm in diameter, was recognized on palpation. There were no other abnormal findings on physical examination. A computed tomography (CT) scan of the abdomen revealed that the tumor was 23 × 21 mm in diameter, the mass being composed of a slightly lower-level lesion in comparison with the right rectus abdominis (Fig. 1A). Magnetic resonance imaging showed that the mass was a well-circumscribed oval lesion with low intensity on T1-weighted imaging and high intensity on T2-weighted imaging (Fig. 1B). Laboratory findings, including ferritin and lactase dehydrogenase levels, were within the normal range. Among the differential diagnoses, we considered fibroma for the benign tumor. Furthermore, although the possibility of malignancy was not denied completely, we strongly suspected nodular fasciitis owing to the patient's trauma history of abdominal bruising, because this tumor showed relatively rapid growth. He was therefore submitted to surgical resection and biopsy to diagnose the tumor. The elastic-soft tumor, measuring approximately 25 mm in diameter with a fibrous capsule, arose from the right lower lateral border of the frontal sheath of the rectus abdominis. Although we were able to separate the tumor from subcutaneous tissue easily, the posterior surface of the tumor adhered to the sheath of the rectus abdominis. We peeled off a part of the anterior sheath of the rectus abdominis so that we could resect the tumor without exposing the muscle itself. The retrieval specimen had an ovoid shape of 28 × 20 × 18 mm in size, surrounded by a fibrous capsule. On cross section it was smooth, grayish white, and solid, without bleeding and necrosis.

Pathologic analyses revealed that the tumor was composed of spindle-shaped cell proliferation with a blunt-ended, cigar-shaped nucleus (Fig. 2A and B). Partial palisade arrangements were observed in its nucleus, with a mitotic index of 40/50 high-power fields. Immunohistochemical staining was positive for α-smooth muscle actin, desmin, muscle-specific actin (HHF35), and caldesmon. Conversely, S-100 protein, Myo-D1, CD99, CD34, bcl-2, and EMA were all negative (Fig. 2C–F, Table 1). Although it was difficult to differentiate the mass from nodular fasciitis because of the trauma history of a right lower abdominal bruise sustained few

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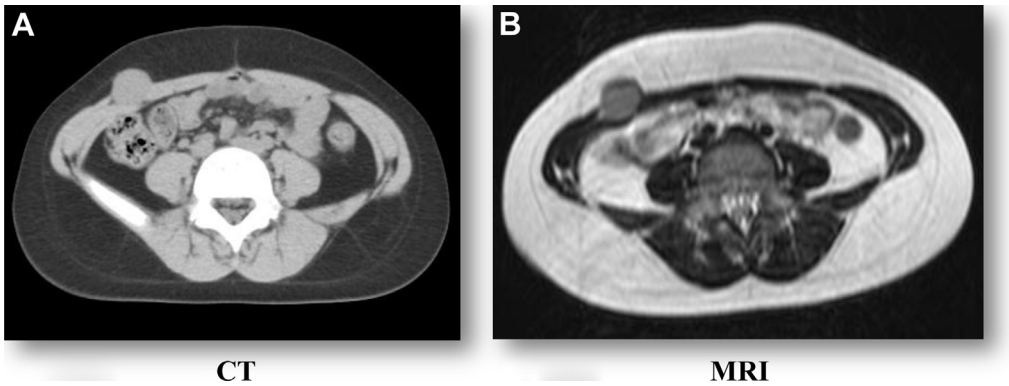

Fig. 1. Preoperative radiologic images. (A) Preoperative CT: The tumor size was 23 × 21 mm in diameter and the mass was composed of a slight low CT level lesion compared with the right rectus abdominis. (B) Preoperative magnetic resonance imaging: The mass had the well-circumscribed oval lesion with low intensity at T1WI and high intensity at T2WI.

weeks before the mass was noticed, pathologic findings led to the diagnosis of leiomyosarcoma because this tumor had the following characteristics: first, the process of peripheral blood vessel movement from smooth muscle to the tumor; second, the hypercellular appearance with abundant cell division; and third, the differentiation into smooth muscle with positive staining of caldesmon.

We carried out careful monitoring and observation without additional therapy because the tumor margin was negative and its pathologic malignancy was of low grade. Moreover, no distant metastasis was identified by postoperative positron emission tomography/CT. Follow-up was uneventful for 2 years.

2. Discussion

Leiomyosarcoma is a malignant neoplasm arising from smooth muscle cells, which make up the involuntary muscles such as the walls of blood vessels, internal organs, and skin. This tumor is one of the most common sarcomas in adults, accounting for 10–15% of all primary soft-tissue malignancies [1]. However, this tumor is extremely uncommon in childhood, particularly in soft tissue, where it probably represents less than 1% of primary malignant neoplasm in pediatric soft tissue [2,3]. Based on a large and well-documented series of soft-tissue sarcoma in children, the histologic diagnosis in 71% of the cases was either rhabdomyosarcoma or undifferentiated sarcoma, with only 1.9% of tumors classified as leiomyosarcoma [4]. Regarding the location of leiomyosarcoma, to date there are only two case reports of primary leiomyosarcoma derived from the abdominal wall, one in a 4-year-old boy and the other in a 22-year-old woman [5,6].

Nodular fasciitis is a self-limited pseudosarcomatous proliferation composed of fibroblasts and myofibroblasts [7,8]. Both the clinical and pathologic diagnoses present pitfalls in recognition, because nodular fasciitis simulates a sarcoma with its rapid growth and high cellularity. In some cases, the mitotic activity makes it difficult to distinguish nodular fasciitis from higher grade neoplasms, as in our case [7]. Histologically, the cellular phase of nodular fasciitis consists of plump, spindled, myofibroblastic and ganglion-like mesenchymal cells in whorls, interlacing fascicles, and sheets. Nodular fasciitis is typically nonreactive for caldesmon in immunohistochemical staining. Therefore, in this case we considered the tumor to be leiomyosarcoma rather than nodular fasciitis.

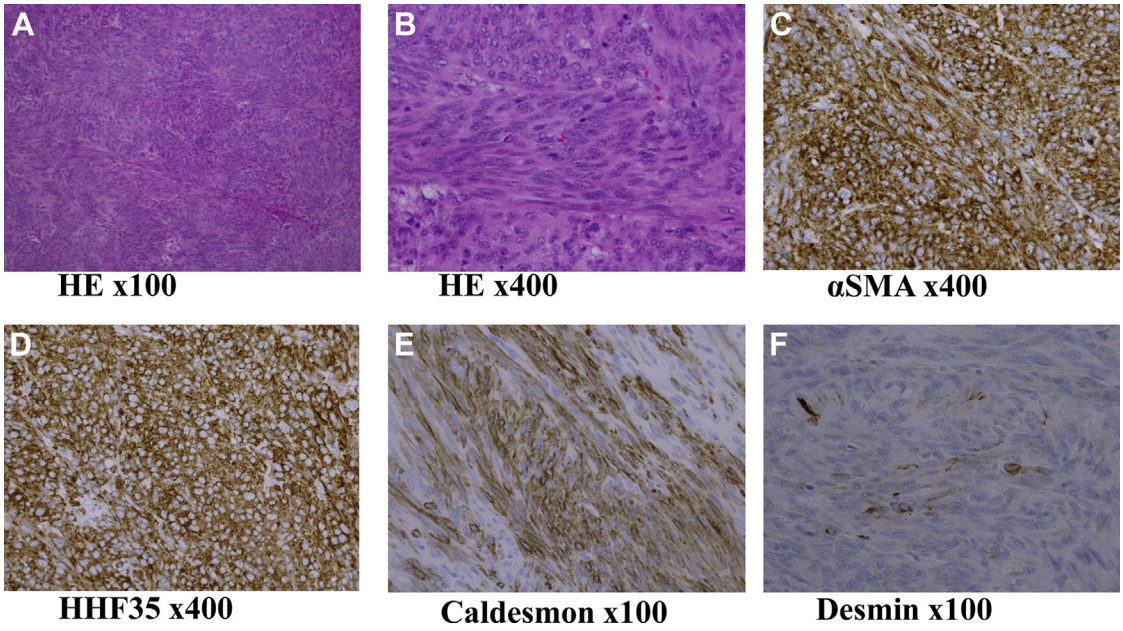

Fig. 2. Pathologic findings. (A) hematoxylin and eosin (H&E) ×100. (B) H&E ×400. (C) αSMA ×400. (D) HHF35 ×400. (E) Caldesmon ×100. (F) Desmin ×100.

Table 1
The results of immunohistochemical stainings

	Marker	Results
Myogenic marker	α-SMA	+
	Desmin	+
	HHF35	+
	Caldesmon	+
	Myo D1	–
Neurologic marker	S100	–
Leukocyte marker	CD99	–
Mesenchymal marker	CD34	–
Cell proliferation marker	bcl-2	–
Epithelial marker	panCK	+
	EMA	–

Regarding the treatment of leiomyosarcoma, almost all previous case reports selected only surgical resection with a pathologic negative margin (chemotherapy or radiotherapy were added in some cases) [5,6,9,10]. Lack reported that leiomyosarcomas in children showed clinical behavior similar to those in adults, with mitotic rates probably the single best histologic predictor of malignancy [9]. However, contrary to the results of adult studies, age at diagnosis appeared to have no prognostic significance in patients aged younger than 21 years [11]. Judging from these previous reports, pediatric leiomyosarcoma seems to carry a good prognosis compared with the adult counterpart, although the number of cases and currently available follow-up data do not allow for a definitive explanation.

3. Conclusion

Although extremely rare, soft-tissue primary leiomyosarcoma of the abdominal wall does occur in childhood. One should keep in mind that there is a possibility of primary abdominal wall leiomyosarcoma mimicking nodular fasciitis, and close follow-up is essential to define the pathologic factors and clinical behavior of this tumor.

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