

A child with intravascular fasciitis mimicking deep vein thrombosis: a case report

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ABSTRACT

Background. Intravascular fasciitis (IF) is a benign, reactive, myofibroblastic proliferation that originates from the superficial or deep fascia of small / medium-sized arteries and veins.

Case Report. An 8-year-old male patient was admitted to a health center with the complaint of swelling in the inguinal region. Lower extremity venous Doppler ultrasonography showed deep vein thrombosis (DVT) of the femoral vein and anticoagulation with low-molecular weight heparin (LMWH) was initiated. The patient was referred to our center for follow-up. The D-dimer level was detected within normal limits. Doppler ultrasonography was repeated and showed an intraluminal expanding mass lesion with increasing vascularity, without distinct borders and LMWH was discontinued. This lesion at the sapheno-femoral junction was excised surgically and the histopathological examination revealed intravascular fasciitis.

Conclusion. Clinicians should be aware that the clinical findings of IF may mimic sarcoma and thrombosis.

Key words: fasciitis, intravascular fasciitis, thrombus.

Intravascular fasciitis (IF) is a benign, reactive, myofibroblastic proliferation affecting small/medium-sized veins and arteries. It is accepted to be a rare variant of nodular fasciitis and it may easily be mistaken for sarcoma and thrombosis. Seventeen IF cases were initially defined by Patchefsky and Enzinger in 1981.¹ Although only 3 adult patients with large vessel IF have been reported to date, no pediatric cases have been described.²⁻⁴ Herein, we report a rare case of an 8-year-old boy initially diagnosed with deep vein thrombosis (DVT), and a mass lesion in the main femoral vein was noticed afterward, leading to a diagnosis of IF.

Case presentation

An eight-year-old boy was admitted to a health center due to inguinal swelling, which was identified two weeks prior to admission. There were no additional symptoms, such as pain or tenderness. There was no family history of a similar disease or consanguinity between the parents of the patient. Right lower extremity venous doppler ultrasonography showed a thrombus with a length of 3 centimeters in the right femoral vein lumen, initiating from the saphenofemoral junction, and the patient was referred to our center with a diagnosis of DVT. Physical examination showed extensive superficial collateral veins and swelling in the inguinal region. The right lower extremity circumference was 1 cm wider than the left. There was no pain, tenderness, or immobility in the inguinal region. He had no history of trauma or infection. Routine laboratory parameters were within normal limits. So, low-molecular weight heparin treatment was initiated due to

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the diagnosis of thrombosis. But the patient had findings of chronic thrombosis (collateral superficial veins) rather than acute symptoms and his D-dimer level was within normal limits (383 ng/ml). The patient was re-evaluated with a venous doppler ultrasonography on the first day of treatment in our center. An intraluminal low-echogenicity expansion lesion with internal vascularization within the right femoral vein was detected, extending to the proximal saphena magna. Venous lumens at the inferior and superior of the mass lesions were intact (Fig. 1-2). Due to the ultrasonographic findings, anticoagulant therapy was discontinued. Magnetic resonance imaging of the right femoral vein was performed to exclude angiosarcoma and vascular tumors and showed soft tissue with internal vascularization. The mass lesion did not show extravascular extension (Fig. 2). The 15x10x7 mm mass lesion was surgically extracted, and histopathological examination revealed the lesion to be IF (Fig. 3). The patient had complications after the surgery and was discharged in 1 week. He has been followed up at three-month intervals for nearly 1 year after surgery without complications.

Informed consent was obtained from the parents of the patient for the publication of this case report.

Discussion

IF was initially defined by Patchefsky and Enzinger in 1981, as a rare variant of nodular fasciitis.¹ IF is a non-tender nodule that is clinically benign, and simple surgical excision is usually sufficient to cure the lesion. Nevertheless, the intravascular expansion ability of the nodules may cause them to be mistaken for a vascular invasion of malignancies such as myofibrosarcoma, fibrosarcoma, liposarcoma, or leiomyosarcoma.^{5,6} For this reason, some authors have used the term “pseudo sarcoma” to define those lesions.⁷ *USP6* gene rearrangement is used as a confirmation tool, which has been shown in nodular fasciitis recently.⁸ The underlying etiology of IF is not known. The myofibroblasts within the vessels are assumed to lead to local proliferative changes without any underlying vascular changes.¹ Although trauma is included among the etiological factors, a scarce number of patients have a history of trauma. Some studies have suggested viral infections and previous thromboses to be possible causes of IF.⁷⁻¹⁰ Although our patient did not have any history of trauma, infection, or previous thrombosis, the clinical findings at the time of admission were initially thought to be suggestive of

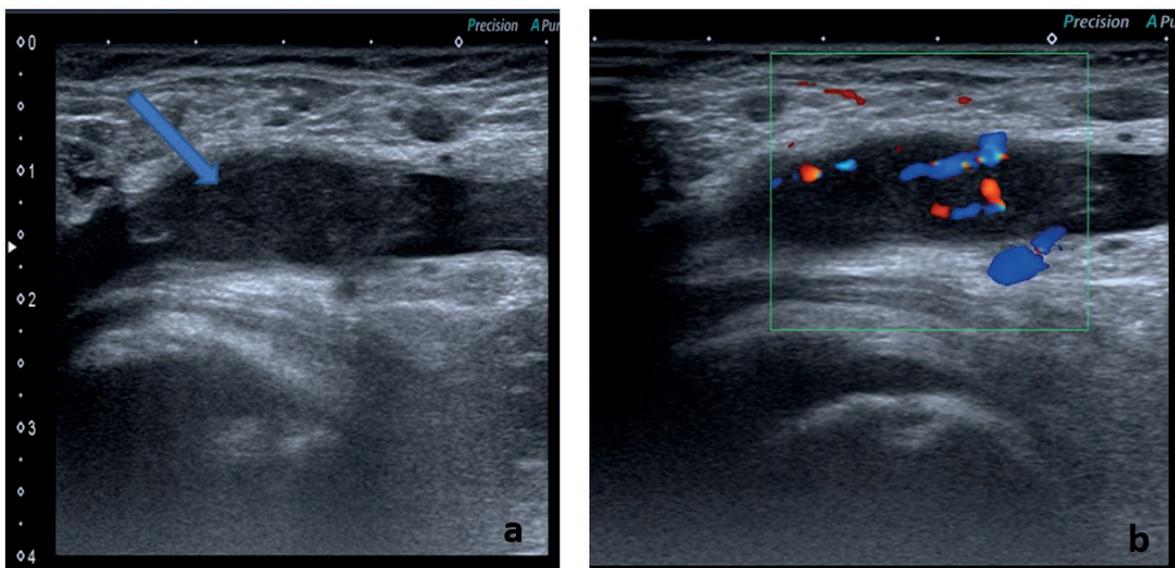


Fig. 1. Ultrasonography and Doppler ultrasonography findings of the patient. **a-** Mass located in the femoral vein on ultrasonography presented with arrow, **b-** Internal vascularization on Doppler ultrasonography

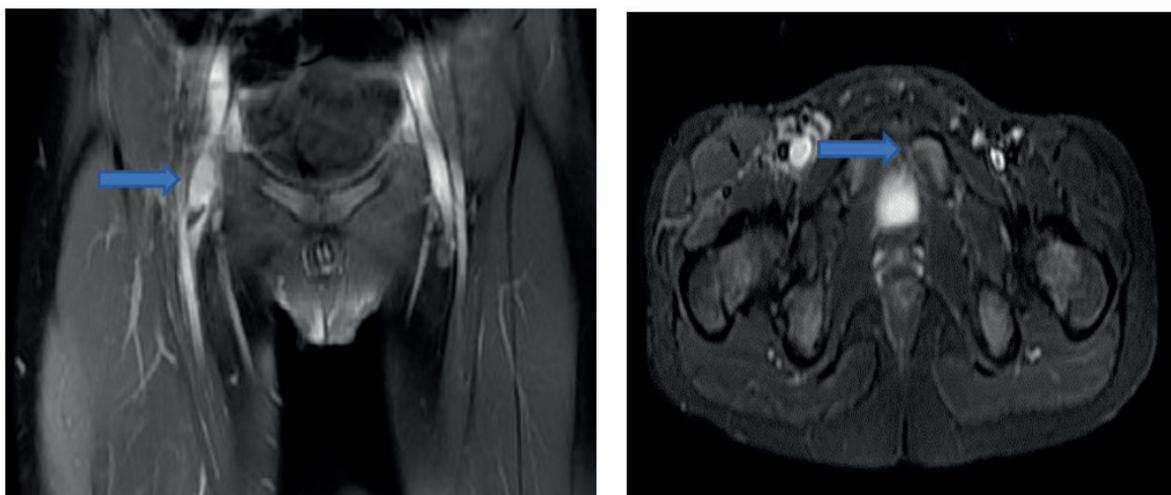


Fig. 2. Coronal (left) and axial (right) magnetic resonance images of the patient. Arrow indicates the mass of the patient

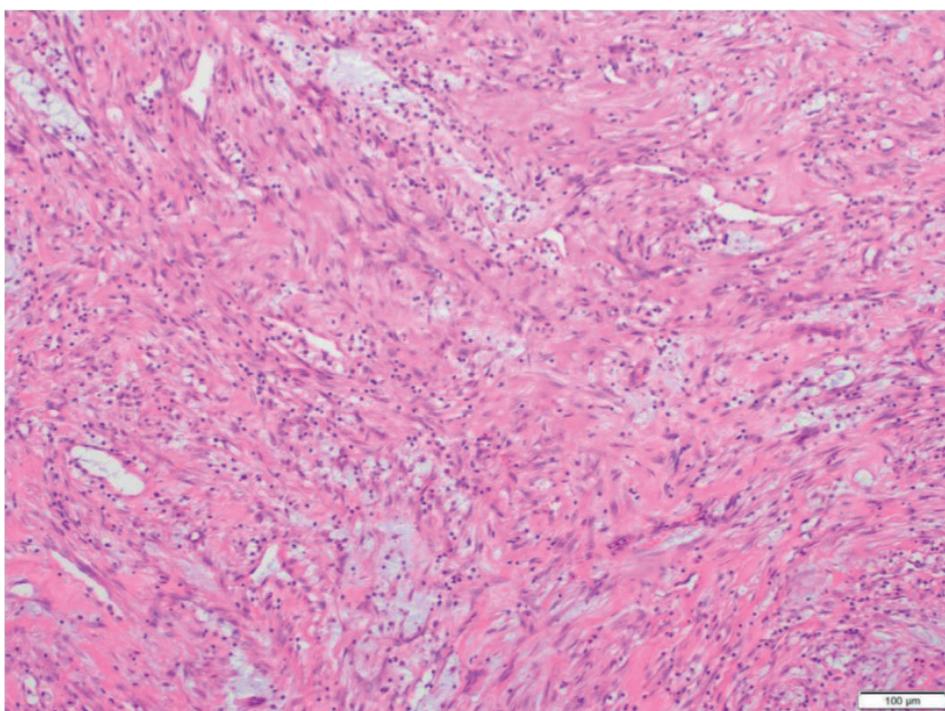


Fig. 3. Histopathologic features of the specimen. Sections show proliferation of benign spindle cells arranged in intersecting fascicles accompanying loose stroma and vascular areas. Scattered lymphocytes are also noted. (Hematoxylene-Eosin staining)

DVT. In this case, the laboratory analyses and ultrasonographic findings excluded DVT. A careful radiological evaluation is mandatory for IF to exclude DVT and other diagnoses.

The most common regions for IF are the upper extremities, head, and neck.¹ Among the 17 cases defined by Patchefsky and Enzinger, the most common location of the lesions was reported

as upper extremities (n:7, 41%), followed by head-neck (n:5, %29), lower extremities (n:3, 12%), and trunk (n:2, 12%).¹ In our case, IF was located at the lower extremity, which is a rare location. IF is mostly known to affect small/medium-sized arteries and veins and is rarely localized in large arteries. There are very few adult patients with IF involving large vessels that have been reported in the literature.^{2-4,11} To the best of our knowledge, our case is the first reported pediatric patient with IF originating from a large vessel (right femoral vein).

The clinical findings of IF may mimic DVT due to the narrowing of the lumen of large vessels. Clinicians should be aware of symptoms suggestive of IF, including non-tenderness, the development of superficial collateral veins that are indicative of a chronic process, and normal D-dimer levels.

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Ethical approval

Authors declare that they have written consent from the family.

Author contribution

Conception and design: YY; data collection: OE, ATY; analysis and interpretation of results: EÖ, HG; draft manuscript preparation: YY. All authors reviewed and approved the final version of the manuscript.

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Conflict of interest

The authors declare that there is no conflict of interest.

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