Intravascular fasciitis presenting as recurrent deep venous thrombosis

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ABSTRACT

Intravascular fasciitis is a rare variant of nodular fasciitis, a benign process that results from proliferation of myofibroblasts in the soft tissues. Nodular fasciitis occurs most often in the upper extremities but can also develop in the head, neck, trunk, and lower extremities of young patients. The intravascular variant occurs within small- and medium-size vessels. We have described a case of femoral vein intravascular fasciitis presenting as recurrent deep venous thrombosis. (J Vasc Surg Cases and Innovative Techniques 2020;6:609-11.)

Keywords: Deep venous thrombosis; Intravascular fasciitis; Nodular fasciitis

Intravascular fasciitis (IF) is a rare variant of nodular fasciitis, a benign process that results from proliferation of myofibroblasts in the soft tissues. Nodular fasciitis occurs most often in the upper extremities but can also develop in the head, neck, trunk, and lower extremities of young patients. The intravascular variant occurs within small- and medium-size vessels. In the present report, we have described a case of femoral vein IF that had presented as recurrent deep venous thrombosis (DVT). The patient provided written informed consent for her case and images to be used in our case report.

CASE REPORT

A 23-year-old white woman had presented to the hospital at 2 months postpartum with a chief complaint of left leg pain and swelling. Other than iron deficiency anemia, she was healthy. She had previously undergone tonsillectomy and excision of a back lipoma. She was a nonsmoker and denied the use of any drugs or alcohol. Her home medications consisted only of iron supplements. The physical examination findings were remarkable for a tender and edematous left leg. Her distal pulses were intact without any sensorimotor deficits. No skin discoloration to suggest phlegmasia was present. From the duplex ultrasound studies, left iliofemoral deep venous thrombosis was diagnosed. She subsequently underwent catheter-directed thrombolysis with good angiographic results. The completion venogram demonstrated residual thrombus within the common femoral vein that could not be declotted. The patient noted significant improvement in her symptoms and started therapeutic anticoagulation with subcutaneous enoxaparin, which was transitioned to warfarin. Despite medication compliance, the patient had developed recurrent left leg swelling within weeks. She was diagnosed with recurrent ipsilateral DVT and underwent repeat catheter-directed thrombolysis. The completion venogram demonstrated an abnormality of the common femoral vein similar to the findings from the initial venogram (Fig 1). A filling defect was also appreciated. However, it was unclear whether this represented an organized thrombus or residual debris. Thus, rather than performing venoplasty and stent placement, the patient was referred for surgical consultation.

On exploration of the left common femoral vein, the patient was found to have a soft, tan, ovoid mass at the confluence of the profunda vein and superficial femoral vein (Fig 2). This tumor was easily separated from the wall of the vein, and dissection along the posterior aspect revealed that it was protruding from the profunda vein. No back bleeding from the profunda vein occurred. The lumen of the profunda appeared to have been completely replaced by the same substance as the tumor. Bovine pericardial patch angioplasty was performed in standard fashion. Bovine pericardial patch angioplasty was chosen over a saphenous vein patch in case the patient experienced recurrent DVT postoperatively, in which case the superficial venous system would provide collateral outflow. A venogram and intravascular ultrasound examination were completed via patch puncture, with no identifiable anatomic abnormalities or outflow stenoses seen. The patient was discharged home on postoperative day 1 with an enoxaparin bridge to warfarin. The patient had undergone follow-up examinations at 3, 6, and 24 months, with surveillance duplex ultrasound scans demonstrating patency of the left common femoral vein.

The intraoperative frozen section analysis was significant for spindle cells without evidence of malignancy. The histologic examination revealed a spindle cell proliferation composed of short fascicles of myofibroblasts arranged in a storiform pattern

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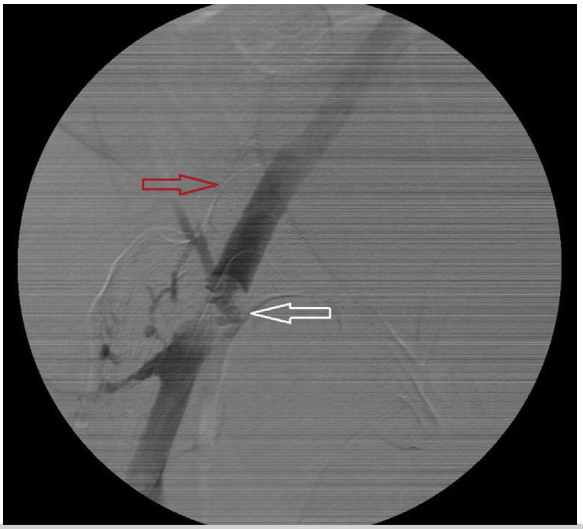

Fig 1. Left venogram of the patient in a prone position showing a filling defect in left common femoral vein (*white arrow*). *Red arrow* denotes the femoral head.

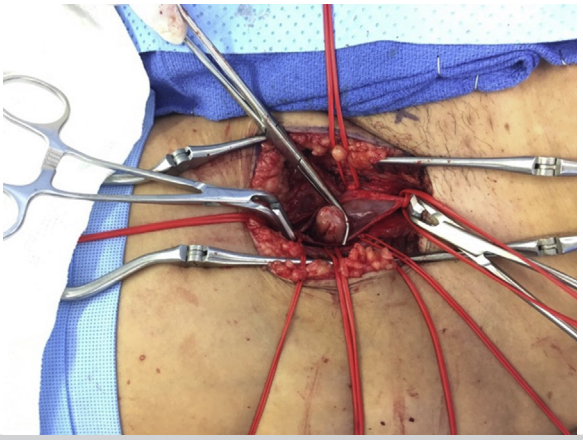

Fig 2. Intraoperative image of the left common femoral vein lesion in situ.

(Fig 3). Multinucleated giant cells were scattered among the fibroblasts, along with lymphocytic cells and few extravasated red blood cells. The spindle cells showed minimal atypia with rare normal mitoses. Elastic staining revealed a remnant of elastic fibers at the periphery of the lesion. The immunohistochemical profile was consistent with nodular fasciitis and showed diffuse positivity in the spindle cells for smooth muscle actin and vimentin. Staining for desmin and S-100 was negative. Beta-catenin exhibited cytoplasmic staining.

DISCUSSION

IF represents a variant of nodular fasciitis, which is thought to be a reactive proliferation of certain lines of mesenchymal cells, such as myofibroblasts. Best characterized as benign tumors, nodular fasciitis lesions are typically found in proximity to the superficial fascia. Histologically, these lesions will demonstrate a reactive,

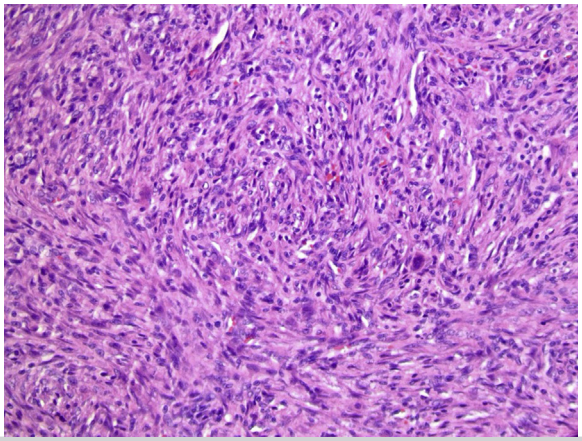

Fig 3. Histologic staining of the lesion showing proliferation of myofibroblastic spindle cells arranged in bundles and fascicles with a dense reticulin network.

inflammatory stroma, hence the term "fasciitis." The intravascular variant, as previously described by Patchefsky and Enzinger[1] in the largest known series, involves the wall of small- and medium-size blood vessels. In their series of 17 cases, the most common presentation was a painless, enlarging mass.[1] Although IF has been reported in older patients, most patients (82% in the series reported by Patchefsky and Enzinger[1]) have been <30 years old.[2,3] These lesions have not shown any gender predisposition and have generally been small, with a mean diameter of only 1.5 cm. Since the report of their large series >35 years ago, multiple individual cases have been reported. Zheng et al[4] provided a comprehensive summary of these reports through 2014. Of the 16 additional cases, 7 (44%) had been lesions found in the head and neck. In contrast, head and neck lesions had accounted for only 29% of the lesions reported by Patchefsky and Enzinger.[1] Although benign, two patients in the series reported by Patchefsky and Enzinger[1] had developed local recurrence.

To the best of our knowledge, the present case is only the third reported case of an intravascular fasciitis presenting as DVT. Lee et al[5] described the case of a young female patient who had presented with left leg swelling. Unlike our patient, however, axial imaging studies were available for their patient. A computed tomography angiogram had revealed a mass in the left common femoral vein with heterogeneous enhancement in the arterial phase that was even more pronounced in the delayed phase.[5] These findings were strongly suggestive of a tumor, which prompted surgical exploration. The femoral vein was resected and repaired with an interposition prosthetic graft, in contrast to excision with patch angioplasty, which was used for our patient. Pan et al[6] described the case of a young male patient who had presented with lower extremity pain and swelling with a negative D-dimer level that had been misdiagnosed as DVT.[6] The patient had subsequently been diagnosed

with IF after an ascending venogram had demonstrated a mass in the common femoral vein that was eventually resected and found to be consistent with IF.

CONCLUSION

When young, healthy patients with a low venous thromboembolic risk present with aggressive or recurrent disease in the absence of inciting factors, consideration should be given to axial imaging studies and/or surgical exploration. Venography and adjuncts such as intravascular ultrasonography could also be helpful. Although rare, the differential diagnosis should include vascular tumors such as IF.

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