Iliofemoral vein thrombosis due to an intravascular fasciitis

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Intravascular fasciitis is a rare intravascular benign tumor that is usually located in the head and neck, the upper extremities, and trunk. Here we report a unique case of intravascular fasciitis occurring in the common femoral vein and causing iliopelvical thrombosis in an otherwise healthy 29-year-old woman. (J Vasc Surg Cases 2015;1:73-6.)

Intravascular fasciitis is unusual variant of nodular fasciitis that refers to a well-established, benign, tumor-like lesion arising in the soft tissue.1 Intravascular fasciitis is a rare condition, and a total of 34 cases have been reported in the English literature since the first description of intravascular fasciitis by Patchefsky and Enzinger,2 and reviewed by Zheng et al.3 The lesion is commonly located in the small veins and arteries of the upper extremities and trunk as well as of the head and neck.1,2,3,6

Here we report the first case occurring in the common femoral vein (CFV) and causing iliopelvical vein thrombosis. The patient consented for her data, including images, to be published.

CASE REPORT

A 29-year-old woman presented with acutely developed pain and diffuse swelling of the left leg. She had no underlying medical disorder except taking drospirenone and ethinyl estradiol (Yasmin; Bayer Korea, Seoul, Korea) due to dysmenorrhea for 3 months. Magnetic resonance imaging at the local hospital (Fig 1, A) showed an enlarged left external iliac vein and CFV and intravascular T2 signal intensity with mild perivascular soft tissue infiltration, which were consistent with deep vein thrombosis. After placement of a vena cava filter through right internal jugular vein approach, an emergency mechanical thrombectomy was performed. This was followed by thrombolysis with 700,000 IU of urokinase into the region of residual thrombus for 7 hours but had no effect (Fig 1, B). An operation was performed to remove the thrombus through a small inguinal incision. There was inflammatory change around the CFV. After proximal and distal control, a 4-cm longitudinal venotomy over the CFV was made. The thrombus completely obliterated the lumen of the CFV and was free-floating within the lumen. The thrombus was deeply attached at the saphenofemoral vein junction and removed directly. The vein was primary repaired with 6-0 polypropylene suture. Because of the intimal damage at the saphenofemoral junction, the patient was treated with rivaroxaban. The follow-up duplex ultrasound imaging showed no residual or recurrent disease at 3 months after surgery, and the swelling of the leg was completely resolved.

The removed thrombus was 4.5 cm × 2.0 cm in size and looked like a fusiform mass, which has a smooth surface (Fig 2). Microscopically, the mass was composed of spindle cells with mild nuclear atypia and two mitoses per 10 high-power fields. The proliferative spindle cells were arranged in a swirling pattern. Extravasated red blood cells were frequently observed in the edematous stroma (Fig 3, A). Immunohistochemical studies (Fig 3, B and E) showed that the spindle cells were positive for smooth muscle actin and negative for desmin. The capillary network was clearly demonstrated with CD34 staining. Spindle cells positive for Ki-67 were frequently seen, and the labeling index averaged 7%. The overall morphologic and immunohistochemical findings were consistent with intravascular fasciitis.

DISCUSSION

Since the original report by Patchefsky and Enzinger,2 33 cases of intravascular fasciitis have been reported, and the most frequent anatomic locations are upper extremities and the head and neck region, where oral cavity and perioral soft tissue are preferentially involved.4-10 Intravascular fasciitis preferentially occurs in adolescent and young adults, who are generally healthy before the onset of the disease. Gender predilection has not been reported. Most intravascular fasciitis has been reported as palpable, painless, and non-tender subcutaneous nodules. The
involved vessels were arteries or veins, and the size of involved vessels varied greatly among reported cases, ranging from small intradermal to large subcutaneous vessels. Patchefsky and Enzinger originally hypothesized that intravascular fasciitis results from proliferation of myofibroblasts in the walls of arteries and veins, and therefore, the lesion extends into perivascular connective tissue and can be intramural, intraluminal, and extraluminal. Possible predisposing factors, such as preceding trauma and hormonal changes, were reported in a few cases.

Nodular fasciitis, which is the original disease entity, can resolve spontaneously, without the need for surgical excision. However, excision has been recommended as a treatment. The clinical behavior of intravascular fasciitis is also usually benign and cured by a simple local excision. However, a few recurrent cases after excision are included in the original cases series by Patchefsky and Enzinger, and follow-up should be done.

The growth characteristic with infiltrative border and histopathologic features, which include increased mitotic activity, may lead to a misdiagnosis of sarcoma. In the original case series by Patchefsky and Enzinger, a malignancy was diagnosed in six of 15 patients. Recognizing this disease entity of intravascular fasciitis is important to avoid a misdiagnosis, which inevitably leads to aggressive treatment. However, no imaging findings specific to intravascular fasciitis have been reported to date.

The lesion in our patient was located inside the CFV. To our knowledge, this patient is the first case in which the intravascular fasciitis was involved in a large and named deep vein. Furthermore, iliofemoral vein thrombosis caused by obstruction of the CFV resulting from intravascular fasciitis has not been described thus far.

The pathogenesis of intravascular fasciitis remains to be understood. The immunohistochemical features of the proliferative spindle cells, as defined by vimentin and α-smooth muscle actin expression, lacked desmin and CD34 expression, which suggests their myofibroblast origin. Certain stimuli, including hormonal change, trauma, or venous thrombosis, are assumed to give rise to proliferation of myofibroblasts in the vessel wall. In the present patient, it can be argued whether the venous thrombosis might have predisposed her to myofibroblast proliferation. However, the iliofemoral vein thrombosis was in an acute stage, and there was no vascular change.

Fig 1. A, T2-weighted magnetic resonance image showed the thrombus in the left common femoral vein (CFV). B, The initial venogram revealed thrombotic occlusion of distal external iliac vein and CFV. C, Mechanical thrombectomy and thrombolysis were attempted, but a short segment of thrombus was remained. D, After treatment for 1 month with rivaroxaban, an enhanced computed tomography (CT) scan showed remnant thrombus in the left CFV. E, The venogram showed fusiform mass in the CFV, and second course of thrombolysis did not have an effect.

Fig 2. The removed thrombus resembled a fusiform mass.
associated with chronic organized thrombus. Chronic venous thrombosis usually occurs as a cord-like fibrous thickening of the vein or a simple occlusive change with recanalization of vascular channel. There is no evidence that pre-existing venous thrombosis can cause intravascular myofibroblastic proliferation. Instead, a more plausible explanation is that the pre-existing intravascular fasciitis occluded the CFV and predisposed upstream acute venous thrombosis in our patient.

CONCLUSIONS

Our case suggests that intravascular fasciitis can occur in a large deep vein and can cause venous thrombosis by vein occlusion.

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