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CASE REPORT

Adventitial cystic disease of the common femoral vein presenting as deep vein thrombosis

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Summary Adventitial cystic disease of the common femoral vein is a rare condition. We herein report the case of a 50-year-old woman who presented with painless swelling in her left lower leg that resembled deep vein thrombosis. She underwent femoral exploration and excision of the cystic wall. The presentation, investigation, treatment, and pathology of this condition are discussed with a literature review.

1. Introduction

Adventitial cystic disease (ACD) of the veins is a rare condition with an uncertain etiology in which a mucin-containing cyst is formed in the walls of the veins. The disease may be difficult to diagnose because its incidence is very low and the initial presentation is similar to that of deep vein thrombosis (DVT).[1–3] We herein report the case of ACD of the common femoral vein (CFV), which showed characteristic features similar to that of DVT, in a 50-year-old woman who presented with painless swelling in her lower leg. This case report was approved by the investigational review board (Research No. KC11ZISE0635).

2. Case report

A 50-year-old woman presented with a 1-month history of unexplained edema in the left lower leg. The circumference

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of the right thigh was 3 cm larger than that of the contralateral side at the mid-thigh level. On the day of admission, her serum D-dimer level was normal (0.26 mcg/mL), but ultrasound examination revealed the presence of an anechoic focal mass, measuring approximately 3.2 cm × 1.7 cm, in the left CFV without compressibility (Fig. 1). A contrast-enhanced computed tomography (CT) scan also showed the presence of an intraluminal low-attenuating mass lesion (2.7 cm × 1.8 cm) involving the left CFV (Fig. 2).

In view of presumed DVT, the patient was anticoagulated with low-molecular-weight heparin; a retrievable inferior vena cava (IVC) filter was placed at the infrarenal segment by the right internal jugular venous approach, after which surgery was done for exploration. While performing the surgery, the common femoral artery was exposed and retracted laterally to facilitate dissection of the CFV. The CFV, deep femoral vein, and great saphenous vein (GSV) were dissected. When a transverse venotomy was performed on the CFV, a 3.0 cm × 1.7 cm adventitial cystic mass extending from the medial to the posterior surface of the left CFV and the vein lumen was found in a compressed state (Fig. 3). After the gelatinous mucoid substance was evacuated from the cyst, the lumen of the CFV showed no persistent stenosis and pathologic scarring. We resected the cyst wall, except the parts attached to the CFV, with a simple closure of the venotomy site in a transverse fashion using GOR-TEX CV number 7 suture (W. L. Gore & Associates, Inc., Flagstaff, AZ, USA). Pathological results identified the mass as an adventitial cyst.

An improvement was observed in the edema of the patient's leg after surgery and the IVC filter was retrieved percutaneously the next day. The patient was discharged on postoperative Day 2 without being prescribed any anticoagulants.

3. Discussion

ACD is characterized by the accumulation of a gelatinous fluid containing mucoproteins and mucopolysaccharides within the adventitial layer of the blood vessel.[4] ACD of the venous system is a very rare condition, with only about 27 cases described in the worldwide literature. Maldonado-Fernández et al[5] summarized 18 cases of venous ACD in 2004, which were all reported prior to 2001. In this report, we summarized an additional nine cases, which were reported after 2001 (Table 1).

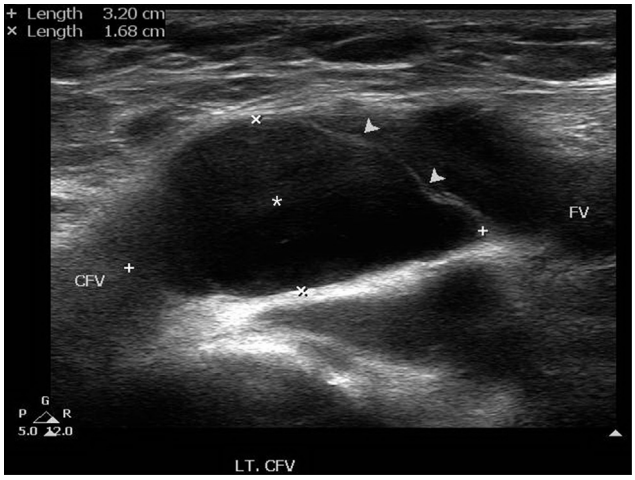

Figure 1 Ultrasound longitudinal scan showing the presence of a 3.2 cm × 1.7 cm, noncompressible, anechoic mass lesion (asterisk) involving the left common femoral vein, and compressing the venous lumen.

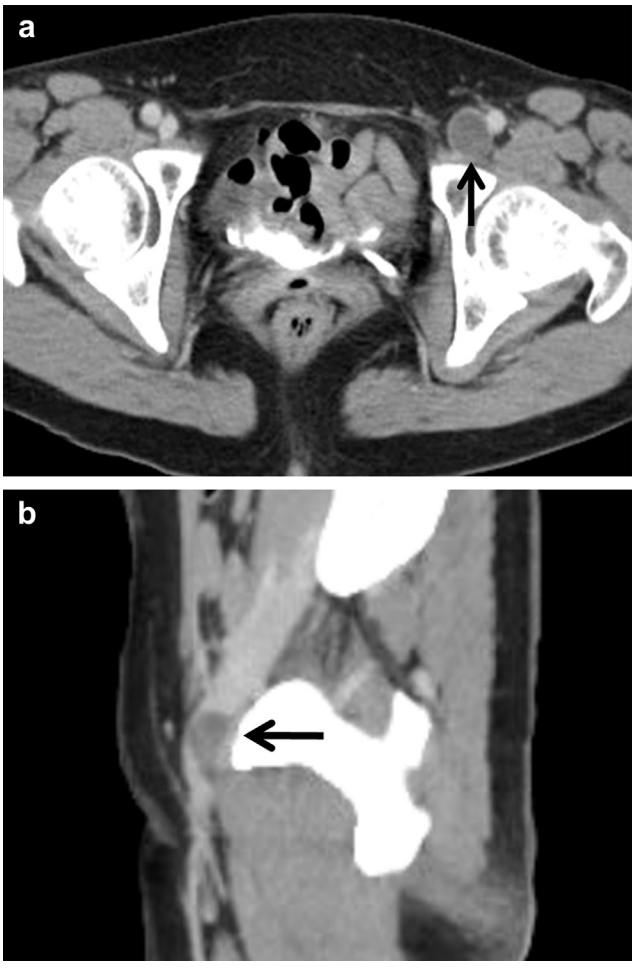

Figure 2 Contrast-enhanced computed tomography demonstrating distension of the left common femoral vein due to an intraluminal hypoattenuating mass lesion (arrow) attached to the posterior wall. The mass was presumed to be a deep vein thrombus.

The exact etiology and pathogenesis of venous ACD still remain uncertain. However, it can be explained in similar terms as that of arterial ACD: (1) the developmental theory (mesenchymal cells from nearby joints implant into the adventitia of the vessel during embryological development); (2) the repeated trauma theory (the adventitia undergoes cystic degeneration as a result of stretching and distortion near the joints); (3) the systemic disorder theory (degeneration of the adventitia as a result of connective tissue diseases); and (4) the ganglion theory (synovial cells implant into the adventitia near the joints).[4,6]

According to the review of 27 cases (Table 1), ACD of the vein tends to develop at a later stage in life (23—75 years; mean: 46.7 years). Among the 27 patients, 16 were men and