Case Report

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Cystic adventitial disease of the common femoral vein: A case report

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Summary **Adventitial cystic disease (ACD) of the common femoral vein is a rare vascular disorder. It becomes more difficult to recognize preoperatively especially when the femoral vein is affected. We report the case of a 62-year-old female patient who presented with a one-month history of painless swelling in her right lower extremity. She had no specific past medical history and no history of trauma, and had a full coagulopathy profile that was negative for any hypercoagulable syndrome. On examination, her lower right leg was significantly swollen with a palpable mass in her right inguinal region. A computerized tomography (CT) with contrast was performed to provide more information and revealed an eccentric compression over the medial wall of the right common femoral vein. During surgical exploration, adventitial cystic mucinous disease was enucleated and the patient underwent femoral exploration, excision of the cysts and reconstruction of iliac femoral vein graft using an artificial blood vessel. The pathological examination confirmed the diagnosis. The patient continued to do well, and she had an unremarkable venous duplex evaluation at her 6-month follow-up. The presentation, investigation, treatment, and pathology of this condition are discussed with a literature review.**

Keywords: Adventitial cystic disease (ACD), femoral vein, 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 arterial variety of ACD has often been described in the popliteal artery (*1*). Patients with this disease will have severe swelling, tenderness, and pain (*2*). In this report, we discuss the case of a 62-year-old woman who presented with a swollen lower leg secondary to obstruction of the common femoral vein. We performed a computerized tomography (CT) scan and ultrasound and this led to excision of a cyst and reconstruction of an iliac femoral vein graft using an artificial blood vessel. As a result, the patient made a full recovery. We also discuss the pathology and the diagnostic methods for this condition.

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2. Case Report

A 62-year-old female was referred to our vascular unit with a one month history of right lower extremity painless swelling. She had no specific past medical history and no history of trauma and had a full coagulopathy profile that was negative for any hypercoagulable syndrome. On examination, her right lower leg was obviously swollen – 10 cm larger in circumference than the left side, with signs of palpable masses in the right inguinal region. No other abnormality was found on physical examination. Ultrasonography showed a cystic mass containing hypoechoic materials attached to the right common femoral vein. Because the diagnosis was uncertain, CT with contrast was performed to provide more information and to exclude other causes such as unusual tumors. A contrast-enhanced computed tomography (CECT) scan also showed the presence of an intraluminal low-attenuating mass lesion (3.2 × 2.1 cm)

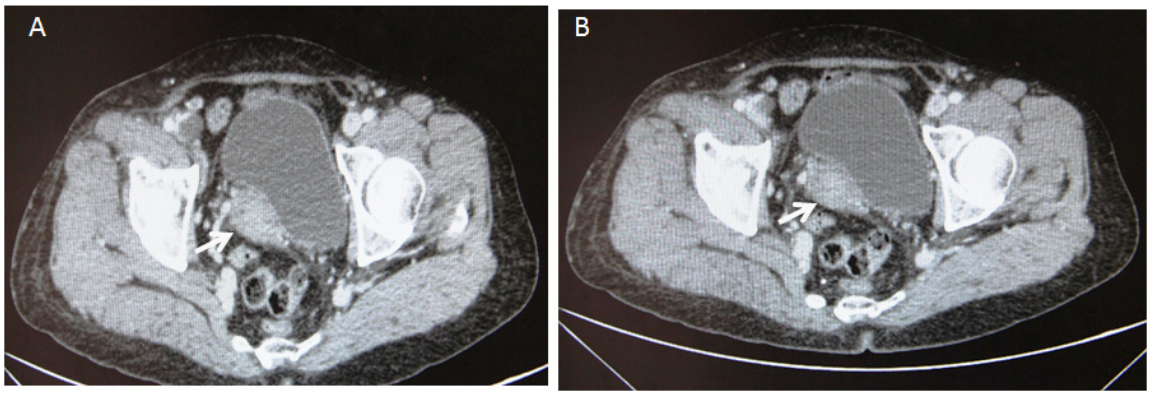

Figure 1. Contrast-enhanced computed tomography demonstrating distension of the left common femoral vein due to an intraluminal hypoattenuating mass lesion (arrow) attached to the bladder wall (A). The mass was presumed to be a deep vein thrombus (B).

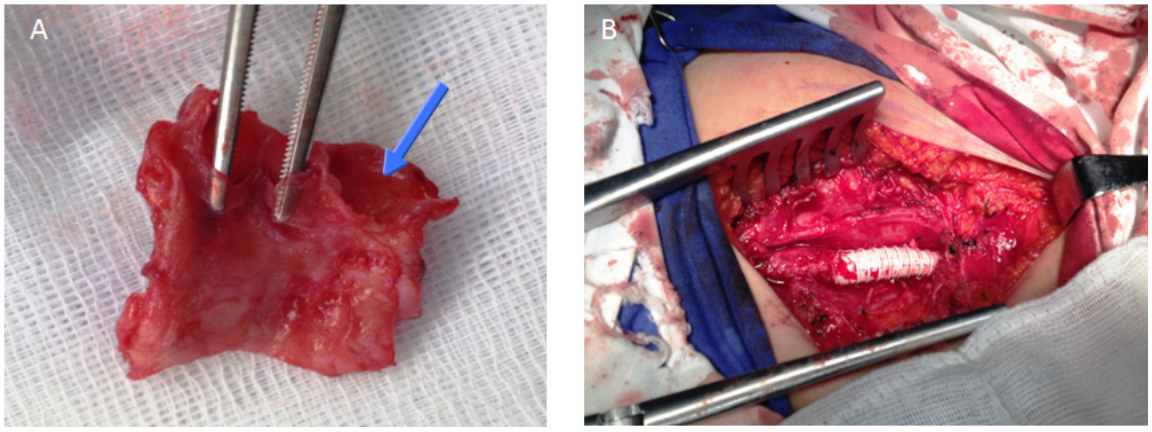

Figure 2. An adventitial cystic mass (Blue arrow) extends from the medial to the posterior surface of the right CFV (A). Operative image illustrates the excision of tumor and reconstruction of iliac femoral vein graft (B).

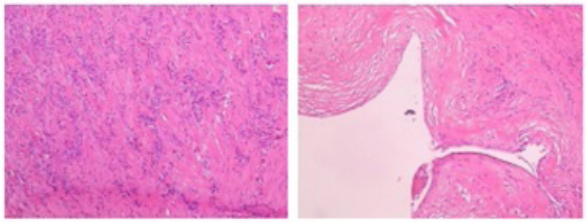

Figure 3. Postoperative photomicrograph indicated a cystic structure with layers of collagen separated by scanty elastic fibers and fibrosis in the wall of the vein and the excised tissue was connective tissue with infiltration of inflammatory cells.

involving the right CFV (Figure 1). The CT revealed obstruction of flow at the level of the CFV, with a presumed thrombus in the CFV and a long saphenous vein but no obvious extravascular mass. The clinical diagnosis was ACD.

A venotomy was made in the posterior wall to reveal thick gelatinous mucoid material lying within a cystic cavity formed by the vein wall. The cyst was excised, with reconstruction of the iliac femoral vein graft using an artificial blood vessel (Figure 2). Histology of the excised specimen revealed a cystic structure with layers of collagen separated by scanty elastic fibers, fibrosis in the wall of the vein and the excised tissue was connective tissue with infiltration of inflammatory cells (Figure 3). Postoperatively, the patient received anticoagulation therapy with warfarin and made an uneventful recovery. At the 6-month follow-up, the swelling in the leg had resolved, and the common femoral vein was patent on color duplex imaging, with no mass effect.

3. Discussion

ACD is characterized by the accumulation of a gelatinous fluid containing mucoproteins and mucopolysaccharides within the adventitial layer of a blood vessel (*1*). ACD was first reported in 1947 by Atkins and Key (*3*), but ACD of the arteries is more frequent in men, it is predominantly located in the popliteal artery and it clinically presents with intermittent claudication (*2*). However, ACD of the venous system is a very rare condition, with fewer than 20 cases reported in the worldwide literature (*4-8*). One of the earliest reports of venous ACD was in the short saphenous vein, and in contrast to arterial ACD, venous

ACD rarely affects the popliteal segment. The venous variety occurs with an equal frequency in both sexes and it most often involves the common femoral vein and causes swelling of the affected limb (*2*).

The exact etiology of ACD remains unclear, but it can be explained in similar terms as that of arterial ACD (*2,9-12*): *i*) The repeated trauma theory (the adventitia undergoes cystic degeneration as a result of stretching and distortion near the joints); *ii*) Ectopic aganglionosis (synovial cells implant into adventitia usually related to arterial ACD near joints); *iii*) the systemic disorder theory (degeneration of the adventitia as a result of connective tissue diseases); and *iv*) the developmental theory (mesenchymal cells from nearby joints implant into the adventitia of the vessel during embryological development).

Histopathologically, the cyst may be uniloculated or multiloculated. The disease process produces an expanding cyst that destroys the elastic tissue between the medium and the adventitia of the vessel wall, and the elastic tissue is replaced with fibrous connective tissue. There is usually no acute or chronic inflammation. The cyst is lined with fibrous connective tissue and the cyst contains an eosinophilic mucoid gel that consists of mucoproteins and mucopolysaccharides (*1,5*).

The diagnosis of ACD of the vein can be suspected on the basis of patient history, results of a physical examination and image findings. As was the case in our patient, the diagnosis is rarely made before surgery owing to the rarity of the condition. First-line investigation should probably involve duplex ultrasound imaging, which is cheap, available in most centers, and noninvasive, to exclude aneurysms and synovial cysts and to localize the cyst to the vessel wall. Ultrasound imaging may show the presence of a typical, anechoic mass with a posterior acoustic window and may allow ultrasound-guided treatment (*13,14*). CT and magnetic resonance imaging (MRI) have also been advocated to localize the pathology to the vessel wall, exclude other pathologies (such as Baker's cyst), and allow guided drainage.6 MRI can reveal high-signal-intensity cysts with extrinsic compression of the vessel lumen. Venography in venous ACD may reveal the site and extent of obstruction to flow and may show a classic scalloped appearance or hourglass narrowing caused by extrinsic compression of the vessel lumen. CT venography is superior to traditional venography for making the diagnosis because the cystic mass can be directly observed regardless of the degree of obstruction. A CT venography can be successfully used in imaging ACD of the vein. When compared with venography, it has the advantage of a noninvasive examination that can directly image the surrounding parenchyma and aid in surgical or percutaneous treatment planning. Whatever imaging is used, it will be necessary to have a high index of suspicion to correctly diagnose this rare condition preoperatively (*15-17*).

Owing to the small number of reported cases, the ideal treatment is unknown and there are three options for venous ACD treatment: *i*) Most authors advocate transadventitial or transluminal evacuation of the cyst, with removal of the cyst wall to prevent recurrence, as in the case we have described. *ii*) Minimally invasive management has been reported with image-guided drainage of adventitial cysts, but incomplete evacuation of cysts secondary to high viscosity has resulted in high recurrence rates. *iii*) The use of needle aspiration of the fluid, guided by ultrasound or CT, has been tried successfully in some cases, although the fluid has a tendency to reaccumulate because the mucin-secreting mesenchymal cells are left in situ (*1,4,13*).

In summary, ACD of the vein is a rare malady, but it should be suspected for patients with symptoms of deep vein thrombosis, and especially when the diagnostic investigation indicates an extrinsic mass. Thus, ACD of the vein needs to be considered in the differential diagnosis of unexplained leg swelling. Furthermore, to ensure a successful outcome, close follow-up of the patient is necessary.

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