

Adventitial Cystic Disease of the Left External Iliac Vein: A Case Report¹

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Adventitial cystic disease (ACD) is a rare, but well-characterized vascular disease. It is most commonly seen in the popliteal artery, but it has also been reported in the venous system. The most commonly involved segment has been the common femoral vein; the disease resulted in luminal compromise and extremity swelling. We report here on a case of adventitial cystic disease of the left external iliac vein that was initially misdiagnosed as deep vein thrombosis in a 68-year-old man who presented with a painless swelling of his left leg.

Index words : Veins, extremities

Veins, iliac

Veins, thrombosis

Adventitial cystic disease is a rare, but well-characterized, vascular disease. It is most commonly seen in the popliteal artery, but it has also been reported in the external iliac, femoral, ulnar and radial arteries. Very few reports of this disease have been described in the venous system such as the common femoral vein, the popliteal vein, the wrist area veins, the lesser saphenous vein and the external iliac vein (1). The most common presentation in the venous system is extremity swelling.

We report here on a case of adventitial cystic disease of the left external iliac vein that was initially misdiagnosed as deep vein thrombosis in a 68-year-old man who presented with a painless swelling of his left leg.

Case Report

A 68-year-old man presented with a 3 - 4 day history

of painless left leg swelling. He had no previous history of trauma, malignancy or deep vein thrombosis. The past medical history was unremarkable. On physical examination, his left leg was swollen and no tenderness, palpable masses or lymphadenopathy were noted. Deep vein thrombosis was suspected based only on the physical examination. Computed tomography (CT) revealed that a short segment of the left iliac vein was occluded by an unenhanced mass-like lesion (Fig. 1A, B). No enlarged lymph nodes or abnormal lesions were found around the vein (Fig. 1A). On 3D CT angiography, smooth luminal defects of the left external iliac vein were noted (Fig. 1C). We then performed sonography for further evaluation. This study showed a small rounded or oval, anechoic or hypoechoic mass that measured 19 × 13 × 11 mm in the left external iliac vein. Doppler sonography confirmed the absence of flow in the cystlike lesion (Fig. 1D). Marsupialization of the cystic mass was performed. The specimen consisted of 5 pieces of gray to yellowish membranous wall that measured 10 × 2 mm at the largest dimension (Fig. 1E). Histologic examination of the vein showed cystic change with mucoid degeneration, and these findings were compatible with the diagnosis of adventitial cystic

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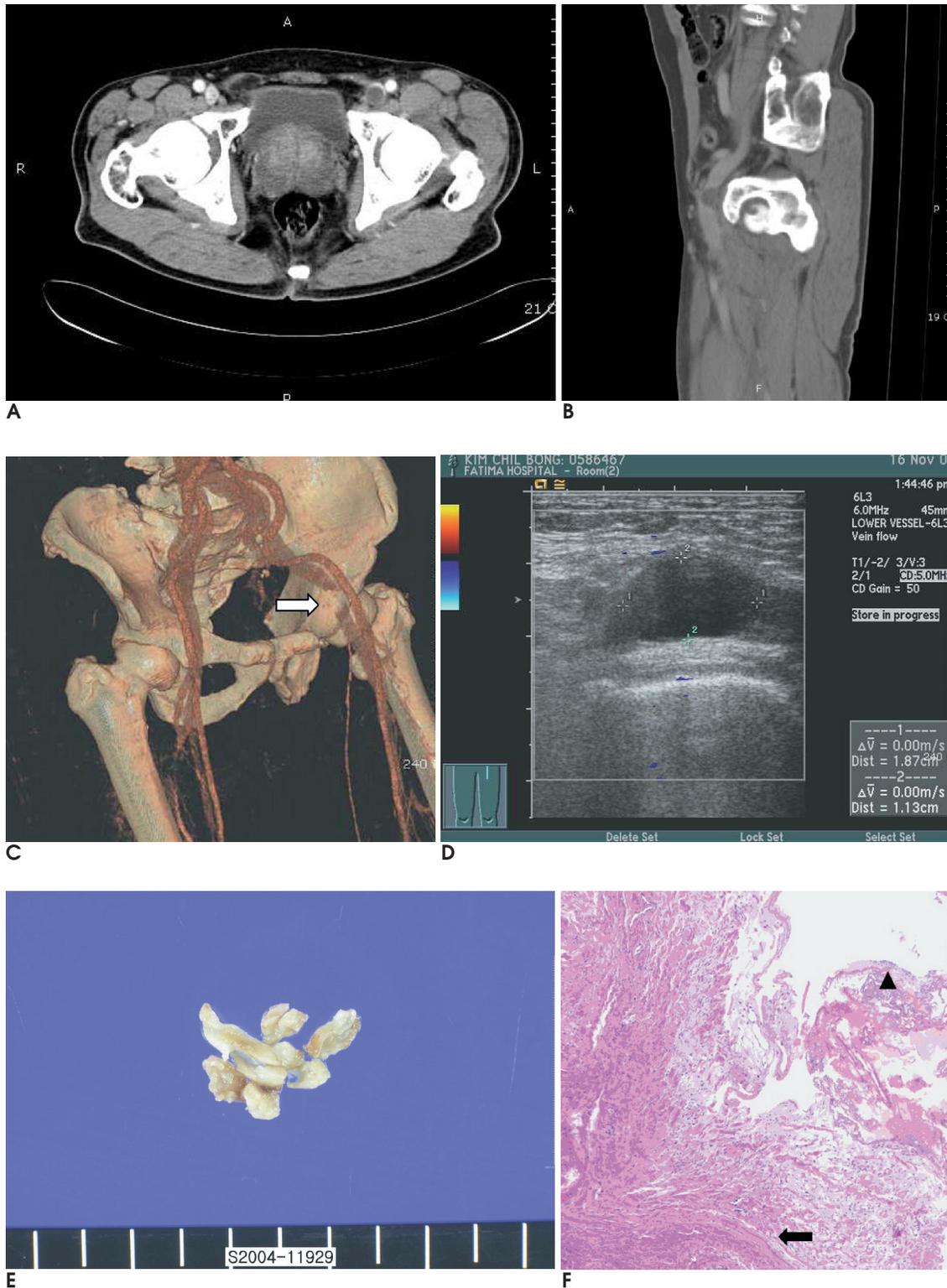


Fig. 1. A 68-year-old man with an Adventitial Cystic Disease (ACD) of the left external iliac vein. **A**(axial image), **B**(sagittal image). CT revealed that a short segment of the left external iliac vein was nearly occluded by an unenhanced mass. No enlarged lymph nodes or abnormal muscular structures were found around the vein. **C.** On 3D CT angiography, smooth luminal defects (arrow) of the left external iliac vein were noted. **D**(sagittal view). Color Doppler ultrasonography shows a 19 × 13 mm sized cystic mass in the lumen of the left external iliac vein. **E.** The specimen consisted of 5 pieces of gray to yellowish membranous wall that measured 10 × 2 mm at the largest dimension. **F.** The microscopic findings demonstrated that the vein (arrow) had cystic change with mucoid degeneration (arrowhead). The wall shows dissected collagen bundles via mucoid degeneration (Hematoxylin-eosin stain, × 100).

disease (2). The vessel wall showed dissected collagen bundles via mucoid degeneration (Fig. 1F). For the 8 month follow-up period, the patient has been asymptomatic and follow-up Doppler sonography showed no recurrence of the cyst.

Discussion

Adventitial cystic disease of the blood vessel wall occurs most commonly in the popliteal artery, with an estimated incidence of about 1 in 1200 cases of claudication. On the other hand, adventitial cystic disease of the vein is a relatively rare vascular condition. In 1998, Levien and Benn identified 323 confirmed cases of adventitial cystic disease of the blood vessels; however, only 17 of these cases (5.3%) involved a vein (3). In 1973, Mentha first described this condition in the tributaries of the lesser saphenous vein (4). Several isolated cases involving the venous system have also been reported in the common femoral vein, the popliteal vein and the wrist area veins.

Venous adventitial cystic disease is more commonly seen in middle-aged men. The most characteristic clinical presentation is gradually-appearing swelling in the lower extremities, so the problem is sometimes misdiagnosed and treated as deep vein thrombosis. The swelling may be accompanied by a painless, non-pulsatile inguinal mass of several months or years duration.

Doppler ultrasound and CT images can show a cystic mass associated with blood vessels.

The pathological findings generally reveal an intramural, subadventitial cyst in the vascular wall. In most of the cases, the cyst was lined with a single layer of cells having mesothelial-like features, and the contents of the cysts were rich in mucopolysaccharides. The contents have been variously described as a clear, dense, yellowish gel or a syrup-like fluid (5).

The clinical presentation of this disease may mimic deep vein thrombosis and the differential diagnosis should consider all disease that can cause unilateral edema of the extremities such as arterial aneurysm, ganglion cyst, lipoma, venous leiomyoma, malignancy or lymphadenopathy (6).

The exact diagnosis is based on the clinical characteristics and complementary explorations. Minimally invasive management has been reported with image-guided drainage of the adventitial cysts (7). Although percutaneous aspiration may initially be successful in reducing the mass effect, the disadvantage of this procedure is that with time, the cyst fluid is re-excreted by the cyst lining (8). Surgical treatments for lesions on the vein include transluminal fenestration, transadventitial evacuation and segmental resection (1). Resection with grafting or with patch angioplasty is necessary when the vessel is occluded.

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