Special Review

Peripheral Non-atherosclerotic Arterial Disorders: What Radiologists Need to Know

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Peripheral non-atherosclerotic arterial disorders (NAADs) are a heterogeneous group of rather uncommon conditions that tend to manifest in subjects without atherosclerosis. Each of these conditions has distinctive pathophysiology; however, there are some common clinical and radiological characteristics and in some cases a common treatment approach that unifies these conditions to a specific group, hence the NAADs. Clinicians and radiologists often fail to recognize NAADs, and there might be a delay in the management of such patients; this may result in seriously adverse outcomes that could otherwise have been avoided or minimized. Knowledge of these conditions and of their radiological appearances is therefore important to help establish a correct diagnosis to allow the prompt initiation of treatment.

The purpose of this pictorial review is to present a selection of NAADs cases and to discuss the radiological characteristics and the most common lines of therapeutic approaches.

Key Words: Non-atherosclerotic arterial disorders; vasculitis; entrapment; endofibrosis; cystic adventitial disease.

INTRODUCTION

Peripheral vascular disease (PVD) because of atherosclerosis is more prevalent in men older than 50 years, commonly presenting with intermittent claudication and is associated with morbidity (limb loss) and mortality (increased risk of myocardial infarction and stroke) (1–3). In a minority of cases, symptoms of intermittent claudication and mimics of PVD are precipitated by non-atherosclerotic arterial disorders (NAADs). The latter patients are often younger, have little or no risk factors for atherosclerosis, and have delayed diagnosis and sometimes, incorrect treatment. Knowledge of these uncommon conditions and their radiological appearance is essential for early diagnosis and initiation of treatment. NAADs are of a heterogeneous group and range from congenital anatomical variations external to the affected vessel to pathologic changes within the vessel wall (4).

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CONGENITAL ANATOMICAL VARIATION

Persistent Sciatic Artery (PSA)

A PSA is a remnant of the embryological vessel that serves as the main blood supply to the lower limb during early fetal development (5). The persistent vessel is a direct continuation of the iliac artery to the popliteal artery and is classified in complete and incomplete types. In the *complete* PSA, detected in the majority of cases, there is direct communication between the internal iliac artery and the popliteal artery. This is associated with a hypoplastic short segment of superficial femoral artery that ends high in the thigh. Patients with this condition are characteristically presented with a popliteal pulse in combination with the absence of a groin pulse. In the *incomplete* variant, alongside a normal femoral arterial system, a partially involuted sciatic artery remains, which communicates with the popliteal artery via collaterals.

Selective catheterization of the internal iliac artery is, therefore, essential to identify this anomalous vessel to avoid a misdiagnosis of a femoral artery occlusion. Clinical findings and symptoms are a result of aneurysm formation as the sciatic artery courses through the sciatic foramen, which can be complicated by thrombosis, embolization, and sciatic nerve compression (Fig 1) (6).

Popliteal Entrapment

Popliteal entrapment is a rare cause of claudication that results from an anomalous relationship between the popliteal artery

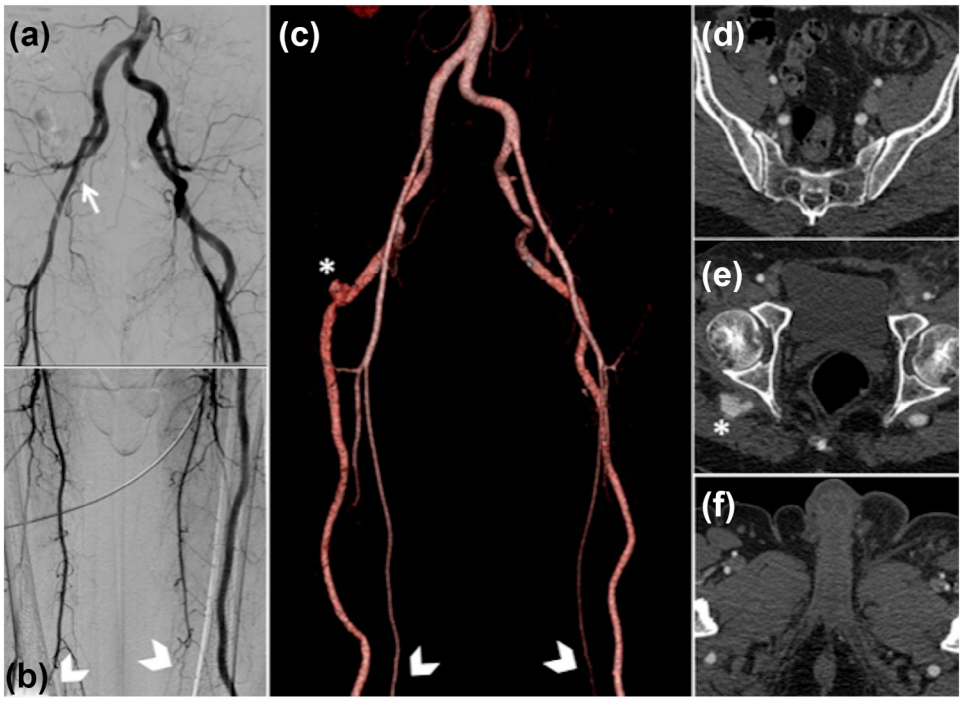

Figure 1. Persistent sciatic artery. **(a, b)** Digital subtraction angiography (DSA) showing bilateral persistent sciatic arteries with complete thrombosis of the persistent sciatic artery on the right (*white arrow*). Note the small distal superficial femoral arteries (*white arrowheads*). **(c)** Computerized tomography (CT) volume rendering technique (VRT) reconstructions performed following thrombolysis now showing both persistent sciatic arteries are patent unmasking the aneurysm of the right persistent sciatic artery, which was the cause of the thrombosis. Axial CT angiography images also following thrombolysis: **(d)** large internal iliac arteries in comparison to the external iliac arteries, the reverse of normal. **(e)** The aneurysm of the right persistent sciatic artery (*white asterisk*) and **(f)** the classic posterior location of the persistent sciatic arteries with small superficial femoral arteries.

and the muscles in the popliteal fossa. Five types have been described based on the developmental course of the popliteal artery in the popliteal fossa and its relationship with the medial head of the gastrocnemius (7). The types can be simplified below as follows:

- Type I: popliteal artery arises medial to the medial head of the gastrocnemius
- Type II: popliteal artery medially displaced to a lesser degree than in type I
- Type III: entrapment of the popliteal artery by a myotendinous band (mesodermal remnant from the medial head of the gastrocnemius)
- Type IV: entrapment of the popliteal artery by the popliteus muscle
- Type V: any of the above with entrapment of the popliteal vein.

The compression of the popliteal artery by the medial head of the gastrocnemius muscle classically manifests itself as claudication following exercise (8). Repeated trauma causes injury to the vessels with resultant stenosis, occlusion, or aneurysm formation (Fig 2). Vascular stenosis can be reproduced on angiography when the gastrocnemius is under tension and is considered the gold standard for diagnosis (Fig 3) (9). Treatment involves muscular decompression and may include arterial reconstruction (10).

VASCULOGENIC

Cystic Adventitial Disease

As the name indicates, the condition involves the development of cysts within the adventitial layer of the artery (usually the popliteal), resulting in luminal compression (10,11). Several theories postulate the pathogenesis of this condition; however, the exact etiology remains uncertain (12). Clinically, patients present with unilateral claudication owing to segmental stenosis or occlusion, which is demonstrated as a curvilinear filling defect on arteriography, also known as the "scimitar" sign or "hourglass" appearance (4,10). Ultrasound can reliably be used to identify cystic changes within the vessel; however, magnetic resonance imaging remains the gold standard as it interrogates the cyst as well as any associated extension to the adjacent joint (Fig 4) (13,14). Treatment options include surgical and endovascular repair with better outcomes demonstrated following the former (15–20). Care must be taken not to confuse this diagnosis with a popliteal aneurysm, particularly on computerized tomography.

Iliac Endofibrosis

This arterial disease typically affects endurance athletes, whereby they present with symptoms of claudication during maximal