

# An Unusual Case of Vasculitis

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A 48-year-old male was admitted due to peripheral ischemia. He had arthralgia during the previous 6 months. Otherwise, he had no diseases per medical history and was on no chronic medications. He presented with digital gangrene patches on his hands and feet and ischemic wounds on both feet [Figure 1]. His blood tests showed leukocytosis 21000/mm<sup>3</sup>, normal creatinine 0.9 mg/dl, elevated C-reactive protein 17 mg/dl (normal < 0.5), and positive cytoplasmic anti-neutrophil cytoplasmic antibody (C-ANCA, anti-PR3 > 200 IU/L). Anti-nuclear antibodies, anticardiolipin, anti-beta2-glycoprotein, cryoglobulins, hepatitis B virus, hepatitis C virus, and HIV serological tests were negative. C3 and C4 were within the normal range. Computed tomography angiography (CTA) demonstrated a beaded appearance and significant filling defect of the branches of the popliteal artery [Figure 2]. Chest, abdomen, and sinus CTA showed no significant findings, and there was no visceral artery involvement. Corticosteroids (solumedrol IV 1000 mg/day followed by prednisone PO 80 mg/day), iloprost, and cyclophosphamide were initiated with promptly normalized acute phase reactants and gradual improvement of limb appearance.

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## DISCUSSION

Polyarteritis nodosa (PAN) is a systemic necrotizing vasculitis that usually targets medium-small sized arteries, particularly in the visceral, renal and soft tissue vasculature [1]. The ANCA test is typically negative, and when ANCA is positive, another diagnosis should be considered [2]. Imaging shows multiple small aneurysms, which is an important diagnostic feature [3].

We presented a case of medium/large vessel vasculitis, associated with positive ANCA, without involvement visceral arteries. Despite the serological results and

the involvement of large vessels, the clinical and radiological findings were more consistent with a diagnosis of PAN; however, technically speaking this is an ANCA associated vasculitis. Our case emphasizes the complexity of the classic classification of vasculitis, which is usually based on vessel size and ANCA findings [4].

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**Figure 1.** Images of an unusual case of vasculitis [A] wound on the dorsal aspect of left foot and necrotic toes, [B] necrotic digits, [C] anterolateral, and [D] posterior view (reconstruction CTA) of the lower extremities show beading (string-of-beads sign) of branches of the popliteal arteries (black arrows). There is severe vascular disease with the absence of the proper artery filling in the lower part of the lower limbs.

