CLINICAL IMAGE

Thrombosis and necrosis of the feet in the course of vasculitis associated with perinuclear anti-neutrophil cytoplasmic antibodies

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The diagnosis of small-vessel vasculitis can be difficult, mainly if typical symptoms and serological markers are not present at the onset of the disease. In severe cases, thrombotic and necrotic lesions are driven by the inflammatory etiology of the disease. Aggressive thrombolytic treatment might be ineffective then. The occupational exposure to heavy metals may play a role in disease induction and the stimulation of antinuclear cytoplasmic antibodies (ANCAs). Some studies have reported a possible relationship between metal exposure and the risk of ANCA-associated vasculitis.[1] In such cases, the differential diagnosis should include microscopic polyangiitis or eosinophilic granulomatosis with polyangiitis.[2] Antinuclear cytoplasmic antibodies are frequently detected in people with a history of asbestos exposure.[3] Patients with ANCA-associated vasculitis should be treated with intravenous glucocorticosteroids, followed by immunosuppressive therapy.[4]

A 61-year-old Caucasian man with massive necrosis of the toes was admitted to the angiology department. At the onset of the disease, Raynaud phenomenon and mild skin lesions were observed (petechiae on the fingertips and toes). The patient was treated with pentoxifylline and acetylsalicylic acid. Later, hemorrhagic and necrotic lesions appeared on the distal fingertips, which was associated with diffuse, intermittent joint pain in the knees and elbows (FIGURE 1A and 1B). Doppler ultrasound revealed multilevel, calcified atherosclerotic lesions in the arteries of the lower extremities. Severe changes were noted in the distal superficial femoral arteries (40% stenosis of the vessel lumen) and the anterior tibial arteries (trace flow was detected in the proximal part of the vessel, and no blood flow on color Doppler imaging of the distal part). On angiography, acute thrombosis in both anterior tibial arteries and digital arteries was detected.

Thrombolytic treatment was administered 3 times, but it did not stop aggressive necrosis. Biochemical and serological test results showed leukocytosis, thrombocytosis, high C-reactive protein levels, and the presence of antinuclear antibodies. The patient's medical history was remarkable for hypertension, 25 years of smoking, and occupational exposure to mercury,

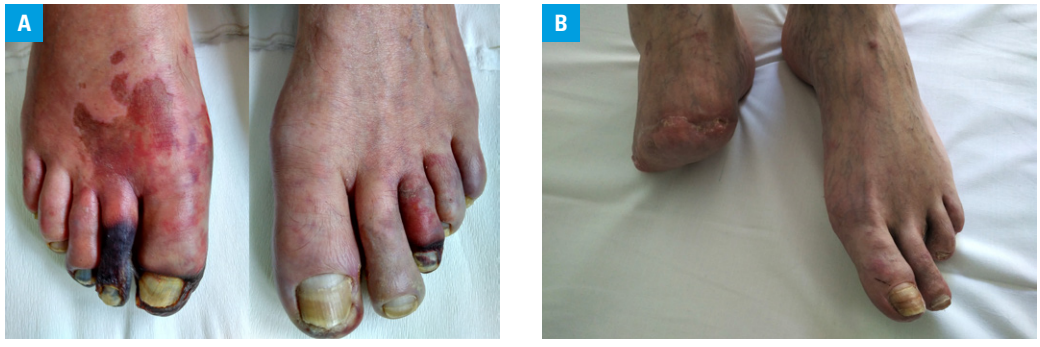

FIGURE 1 **A** – necrotic lesions in undifferentiated antinuclear cytoplasmic antibody (perinuclear pattern)–associated vasculitis before treatment; **B** – undifferentiated antinuclear cytoplasmic antibody (perinuclear pattern)–associated vasculitis: status post amputation

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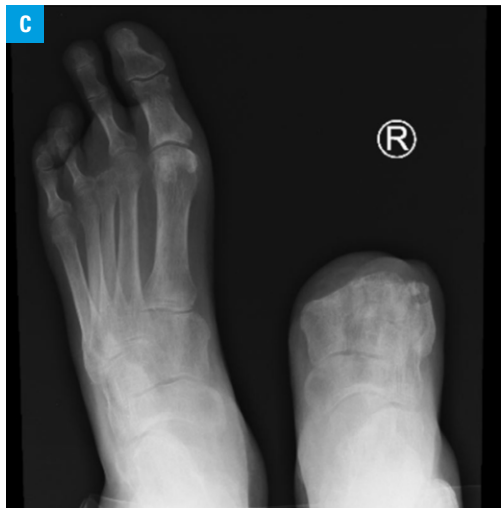

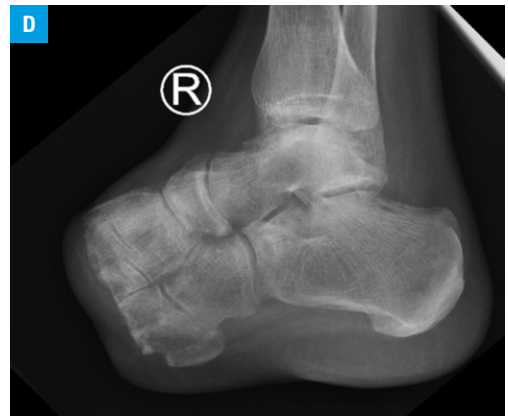

FIGURE 1 **C** – X-ray of the feet: status post amputation of the right foot and the third left toe; **D** – X-ray of the right foot after amputation

asbestos, and lead. Hypertension accompanied by advanced atherosclerosis and thrombus formation as well as long-term cigarette smoking are often observed in patients with small-vessel vasculitis; however, these conditions might also be related to mercury poisoning.

Since musculoskeletal symptoms and an increased number of antinuclear antibodies (titer, 1:160) were observed, the patient was admitted to the Department of Rheumatology, Rehabilitation and Internal Diseases for further diagnostic workup. An increased count of ANCAs with perinuclear staining on indirect immunofluorescence examination (p-ANCAs) was detected (titer, 1:640). Antiphospholipid antibodies and cryoglobulins were not found. Laboratory tests yielded negative results for hepatitis B and C virus, HIV, *Chlamydia trachomatis,* and *Yersinia enterocolitica* species. Due to the lack of organ involvement and the presence of p-ANCAs, the patient was diagnosed with undifferentiated p-ANCA–associated vasculitis. Since massive necrosis was present, ceftriaxone, intravenous immunoglobulin, and methylprednisolone treatment was also initiated. Three weeks later, the patient was hospitalized in the Department of Angiology and Vascular Surgery. Due to massive necrosis, amputation of the middle part of the foot of the right leg and the third toe of the left foot was performed (FIGURE 1C and 1D). The histological examination of the specimen revealed necrotic vasculitis without granuloma formation. Although the presence of clinical and serological manifestations in this case was suggestive of a systemic autoimmune disease, the classification criteria for a defined connective tissue disease or a specific type of vasculitis were not fully met. This case shows that p-ANCA–associated vasculitis limited to the skin, without organ involvement, might progress very quickly and aggressively, resulting in skin necrosis.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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