### CASE REPORTS

# BUERGER'S DISEASE – A CLINICAL CASE

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## Abstract

Buerger's disease is a distal segmental nonatherosclerotic vasculopathy that involves the inferior and superior limbs of smoker males younger than 45 years old.

This article aims to describe a clinical case and revise the literature about Buerger's disease.

A 45-year-old smoker male repeatedly visited the emergency department for refractory pain and inflammatory signs in the right hallux. After developing ulcers in the right foot, Doppler ultrasonography revealed segmental occlusion of distal arteries of that limb. It was also observed in arteriography "corkscrew" collaterals. Autoimmune, thrombophilic and cardiovascular diseases were excluded.

Analgesia, antibiotics and alprostadil were implemented. As a result, the patient stopped smoking and was submitted to minor amputation with complete healing, after which he remained asymptomatic.

Buerger's disease is a diagnosis of exclusion. Therefore, smoking cessation is the most effective treatment and is crucial to prevent disease progression.

Keywords: Buerger's disease, thromboangiitis obliterans, limb ischemia, peripheral artery disease

#### INTRODUCTION

Buerger's disease is a segmental nonatherosclerotic inflammatory condition involving small and medium size arteries and inferior and superior limbs veins. Its etiology is related to forming a highly cellular inflammatory occlusive thrombus with vascular wall preservation.<sup>1-6</sup> It is strongly associated with tobacco, affecting mainly smoker males younger than 45 years old. Its incidence is less than 6% in Western Europe, and it presents in most cases as a digital ischemic ulcer but also as peripheral neuropathy, migratory thrombophlebitis, Raynaud phenomenon and migratory arthritis.<sup>7</sup>

#### **CLINICAL CASE**

A 45-year-old smoker male repeatedly visited the emergency department for refractory pain and inflammatory signs in the right hallux last month (Figure 1). He denied upper limb complaints and a history of phlebitis. A peripheral arterial disease was not recognized; the patient was diagnosed with gout and medicated with anti hyperuricemic drugs without response. One month later, he developed ulcers in the first and fifth fingers of the right foot, and Doppler ultrasonography revealed: "Apparent occlusion of the posterior tibial artery. Extensive partial occlusion of the middle third of peroneal artery. Occlusion of the distal portion of anterior tibial and pedal arteries".

An arteriography was also performed: "Occlusion of the distal third of anterior tibial, posterior tibial and peroneal arteries, after their origin, without visible distal permeabilization. Observation of corkscrew collaterals.". Proximal arteries were preserved, and there were no skip lesions (Figure 2).

Laboratory studies excluded autoimmune, thrombophilic and cardiovascular diseases.

Analgesia and tight control of infectious processes with antibiotics (Amoxicillin/Clavulanate) were implemented. Microbiology identified Enterocococcus faecalis and







Occlusion of distal third of anterior tibial, posterior tibial and peroneal arteries, and observation of "Corkscrew" collaterals.



Citrobacter as resistant to penicillin, and antibiotics were replaced with Trimethoprim-sulfamethoxazole. In addition, a course of vasodilators with alprostadil was performed. After undergoing hallux amputation, he remained without other manifestations of the disease. Histological examination showed: "Arteries with intimal thickening, fibrosis, rupture and duplication of the internal elastic layer, with mononuclear inflammatory infiltrate. Lesions compatible with Buerger's disease.". After several attempts, the patient stopped smoking six months after arteriography (one month after amputation), with precise control of the underlying disease (Figure 3). No revascularization surgery was necessary. The patient was lost to follow-up eight months after arteriography.

#### DISCUSSION

Buerger's disease is a diagnosis of exclusion and should be considered in young male smokers with evidence of peripherical artery disease without other cardiovascular risk factors. Several sets of diagnostic criteria have been suggested, including the classic ones by Shionoya (smoking history, onset before age 50, infrapopliteal arterial occlusive lesions, either upper limb involvement or phlebitis migrans, and an absence of atherosclerotic risk factors other than smoking).<sup>7</sup>

These patients have a mean survival of 52 years and a 43% risk of amputation (12% for major amputation).<sup>8</sup> Although a minority succeeds, smoking cessation is the most effective treatment and the only way to stop the disease's progression, so the risk of amputation is eliminated eight years after quitting.<sup>9</sup> Vasodilators can be used in patients to help relieve symptoms. Pharmacologic agents predominantly investigated include prostacyclin analogues (cilostazol) and phosphodiesterase inhibitors (alprostadil).<sup>7</sup>

Endovascular treatments may not be feasible due to the distal vascular involvement of this disease. Nonetheless, it should be considered when there is superimposed peripheral artery disease or other options have failed.<sup>7,10</sup>

Investigational therapies are being developed (autologous bone marrow injection of mononuclear cells, intramuscular injection of growth factors, immunoadsorption, angiogenesis therapy with staminal cells, etc.). However, their effectiveness is still to be prove and should be evaluated on a case-by-case basis.<sup>7</sup>

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