

# Buerger's disease

## A case report and review of the literature

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### Summary

A case of thrombo-angiitis obliterans (Buerger's disease) in a 30-year-old man is reported. Irremediable gangrene necessitated amputation of both legs, and distal amputation of the fingers and thumb of the right hand.

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In 1908, Leo Buerger, a New York surgeon and pathologist, described a distinctive disease characterized by segmental, thrombotic, obliterative, acute and chronic inflammation of arteries and veins that occurred almost exclusively in male cigarette smokers.<sup>1-3</sup> For years the specificity of this entity came under severe challenge and at one stage Buerger's disease as an entity came into disrepute.<sup>4</sup> Today most workers agree that thrombo-angiitis obliterans is a distinct entity, but that other causes of peripheral vascular disease (e.g. arteriosclerosis) should be ruled out before a definite diagnosis is made.

We report on a patient with Buerger's disease and briefly review the literature.

### Case report

A 30-year-old White man presented in 1980 with sudden onset of intermittent claudication in the right calf and foot. The patient was a heavy smoker of cigarettes. Initially the claudication was mild, but within months it had become intolerable since he had also developed severe claudication in the left leg. The symptoms progressed rapidly and he developed intense ischaemic rest pain in the right foot.

The patient was normotensive with a normal haemoglobin concentration. Examination confirmed that the upper limb pulses were intact, but pulses in both legs distal to the popliteal artery were absent. There were no signs of carotid or coronary artery disease. A provisional diagnosis of Buerger's disease was entertained. The lipid profile, blood glucose level and results of glucose tolerance testing were normal and extensive studies failed to demonstrate an underlying auto-immune disease. Segmental pressures in both lower limbs, measured by the Doppler method, were markedly reduced, but were normal in the more proximal part of the lower limbs. An arteriogram revealed a normal aorta and normal iliac and femoral vessels. Multiple occluded segments were observed in the small and

medium arteries of both legs. An extensive reticular network of collaterals was seen around the occlusions. The classic 'corkscrew' appearance was not present.

The disease had a progressive course and the patient developed localized gangrene of the right foot. A bilateral sympathectomy and conservative amputation of the affected toes were performed. A temporary response was observed but deterioration in the patient's condition necessitated bilateral below-knee amputations within 3 months of his initial admission to hospital.

The patient stopped smoking and made good progress for 2 years. He was able to walk with below-knee prostheses.

He resumed smoking and presented in 1982 with severe rest pain in the right below-knee amputation stump which required an above-knee amputation. Healing of the stump was delayed for months (Fig. 1). Gangrene of the tips of the right thumb and first, second and third digits necessitated distal amputation. In recent months he has had signs of Raynaud's phenomenon in both hands. The patient has continued to smoke and healing of the amputated digits on the right hand is poor.



Fig. 1. A 30-year-old male amputee showing the characteristic result of thrombo-angiitis obliterans; note distal amputation of the thumb and three fingers of the right hand.

### Discussion

In our patient the typical features of Buerger's disease manifested, i.e. occlusive peripheral vascular disease. This occurs almost exclusively in male tobacco smokers,<sup>2,3,5</sup> although in recent years there seems to have been a relative increase in the incidence in women, which may be related to their increased use of tobacco. The disorder begins before the age of 35 years in most patients and before 20 years in some. When occlusive arterial disease of the extremities develops between the ages of 40 and 50 years, arteriosclerosis obliterans should be considered a greater possibility than thrombo-angiitis obliterans. In many patients Buerger's disease affects the arms as well as the legs, and

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demonstrates remission and relapse intimately related to cessation or resumption of smoking. The disease often causes excruciating pain in the extremities, out of all proportion to that found in other forms of peripheral vascular disorder. In many patients Buerger's disease is associated with migratory thrombophlebitis, but is not associated with diabetes mellitus, heart disease or hypercholesterolaemia.<sup>2,3</sup>

The cause and pathogenesis of Buerger's disease are unknown but a strong relationship with cigarette smoking exists.<sup>2,3</sup> Thrombo-angiitis obliterans has only rarely been reported in non-smokers. Some tobacco products (e.g. carbon monoxide) may be directly toxic to vessels, mainly endothelial cells; cigarette smoking may affect catecholamine metabolism and thus cause vasoconstriction, which predisposes to vascular injury. Carbon monoxide may lead to vascular ischaemia, and a hypercoagulable state may lead to thrombosis.<sup>2-4</sup>

It was initially reported that Buerger's disease was found only in Jews, but members of all racial groups are susceptible. A genetic predisposition is suggested by the increased prevalence of HLA A9 and HLA B5 in patients with Buerger's disease.<sup>6,7</sup>

The pathological features of thrombo-angiitis obliterans are distinct from those of other vascular lesions such as arteriosclerosis obliterans. The lesions of Buerger's disease are sharply segmental and predominantly affect the medium and small arteries and only occasionally the larger arteries.<sup>2,8</sup> Vessels of both upper and lower extremities are affected. Thrombo-angiitis obliterans has also been described in the heart, lungs, brain, gastro-intestinal tract and male genitalia.<sup>2,8</sup> The accompanying veins and adjacent nerves are often secondarily affected, leading to progressive fibrous encasement of these structures.<sup>3,9</sup> The arteries commonly involved are the posterior tibial, radial, ulnar, plantar, palmar and digital arteries.<sup>3</sup> The lesion is an inflammatory, non-suppurative panarteritis with associated thrombosis but without necrosis of the vessel wall.<sup>4</sup>

Arteriography plays an important part in diagnosis;<sup>8</sup> patients with Buerger's disease have multiple occluded segments in the small and medium arteries of the arms and legs. The absence of large artery involvement, which can be confirmed by arteriography, is a feature differentiating it from arteriosclerosis. The occlusive lesion shows an abrupt change from a normal to an involved segment; the artery proximal to the occlusion appears smooth and of even calibre. An extensive reticular network of collateral vessels is frequently seen around an occlusion.

The condition must be differentiated from arteriosclerosis obliterans, idiopathic arterial thrombosis, arterial embolism, scleroderma, Raynaud's disease and peri-arteritis nodosa.

The most vital factor in the management of Buerger's disease is cessation of smoking, since the condition does not progress

when this habit ceases. Arterial bypass has not been feasible for patients with Buerger's disease because the arteries involved are relatively small and in most instances are located distal to the knee or elbow.<sup>3,5</sup> There is no evidence to suggest that long-term anticoagulation therapy has any beneficial effect.<sup>3,5</sup> The effectiveness of vasodilating drugs, antiplatelet agents and steroids in Buerger's disease remains controversial. Sympathectomy produces vasodilation and may be of value in patients with thrombo-angiitis obliterans,<sup>3</sup> but the operation is obviously futile in patients with extensive gangrene of the foot or hand.<sup>3</sup> Prophylactic measures to prevent mechanical, chemical and thermal injury, even of minor degree, are essential. When amputation of a leg is necessary, an attempt should always be made to carry out a below-knee surgical procedure.

The onset of Buerger's disease is rapid and the progression of the disease always depends on the smoking habits of the patient. Although the improvement may not be immediate, the symptoms will be arrested and probably improve after cessation of smoking, except in cases of severe, irreversible ischaemia. If the patient resumes smoking, the symptoms will progress.

Once the disease is arrested, the life expectancy of patients with Buerger's disease is good and exceeds that of patients with atherosclerosis.<sup>2,3</sup>

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