

Buerger's disease (thromboangiitis obliterans): clinical features and assessment by colour duplex ultrasound

Abstract

Buerger's disease is a progressive, non-atherosclerotic, segmental inflammatory vaso-occlusive disease of unknown aetiology¹⁻⁹, affecting the small and medium sized arteries, veins and nerves and is often bilateral⁵⁻⁷. The normal structure of the vessel wall is usually preserved, including the internal elastic lamina and media.

Buerger's disease predominantly affects male cigarette smokers^{1-3,8} with 11%–23% of disease distribution in females⁶⁻⁷.

Buerger's disease remains largely a clinicopathologic entity highlighting the importance of the sonographer's role to obtain a comprehensive clinical history and clinical examination. The combination of clinical assessment and the distinctive appearance of vessels affected by Buerger's disease on colour duplex ultrasound provide a useful tool for correct diagnosis.

Keywords: Buerger's disease, claudication, clinical, digits, ischaemia, smoking, ultrasound assessment.

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Introduction

Buerger's disease is a progressive, non-atherosclerotic, segmental inflammatory vaso-occlusive disease of unknown aetiology¹⁻⁹, affecting the small and medium sized arteries, veins and nerves and is often bilateral^{5,7} where inflammatory thrombi may affect both the arteries and veins⁵. This disorder has been described as an autoimmune response, however the precise triggering antigen has not been discovered^{10,11}. The normal structure of the vessel wall is usually preserved, including the internal elastic lamina and media. It is this feature that distinguishes Buerger's disease from arteriosclerosis and from other types of systemic vasculitis^{3,5} (Figures 1a, 1b).

Buerger's disease predominantly affects male cigarette smokers^{1-3,8} with 11%–23% of disease distribution in females^{6,7}, in studies of 850 and 112 subjects respectively^{2,6,11}.

The disease progression has a self-limiting process, with the progression of symptoms influenced by smoking^{4,10}, and an extremely strong association between heavy use of tobacco with Buerger's disease³⁻⁵. The disease has a high prevalence in people of low socioeconomic class who smoke bidis (homemade cigarettes with raw tobacco found in India)^{5,12}. More recently, cannabis use has been attributed to the development of a form of vasculitis with very similar features to Buerger's disease^{8,9}.

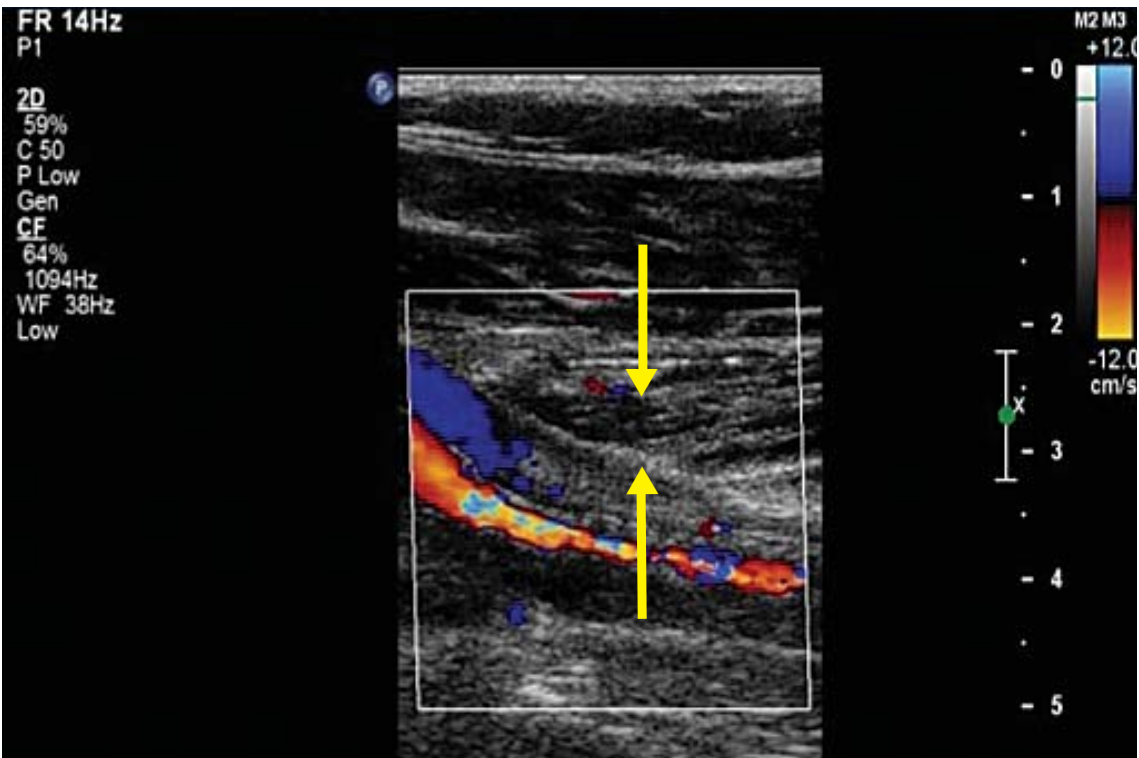
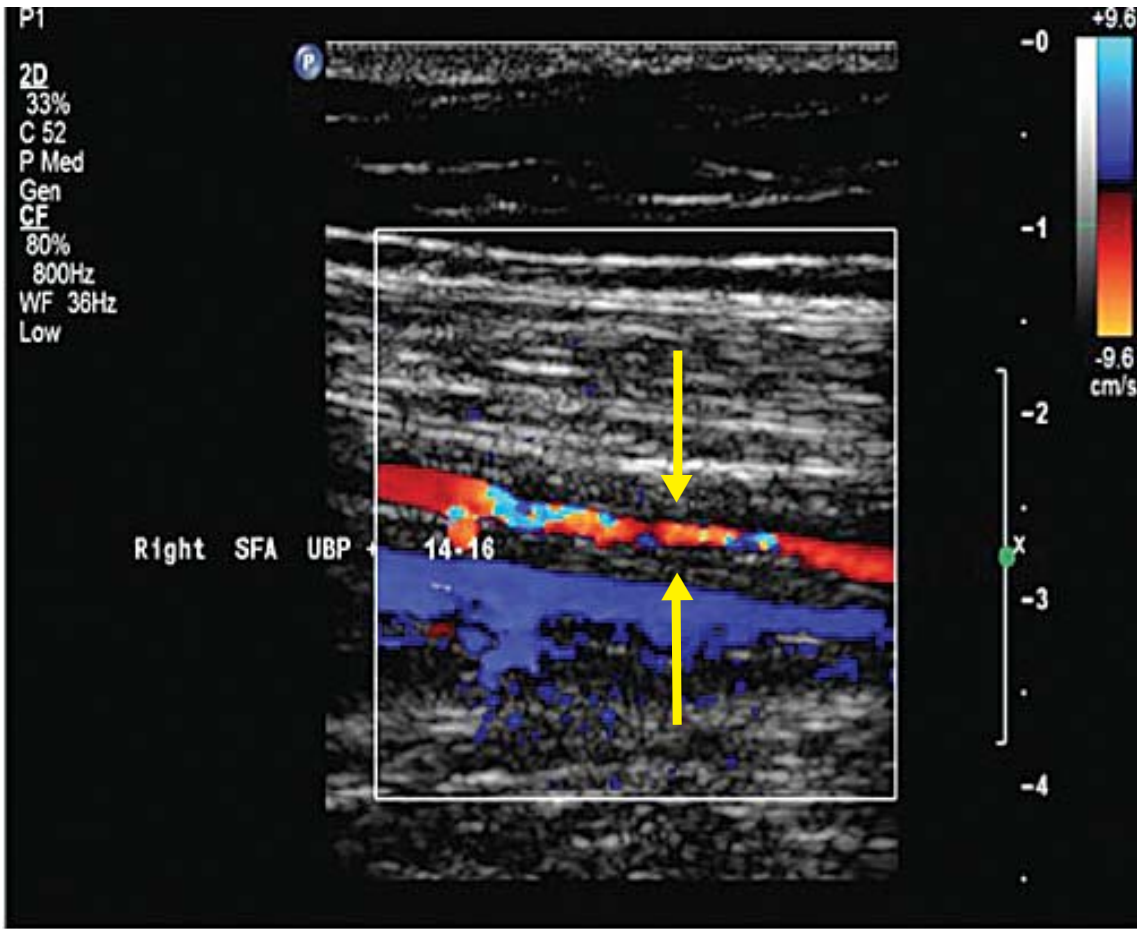
The majority of publications view current or past smoking to be a prerequisite for the diagnosis

of the condition^{1,5-7,11}. A few investigators however believe that Buerger's disease can occur in non-smokers^{2,20}, although less than 5% of Buerger's disease patients have been reported as non-smokers²⁰. These cases might be triggered by cold, frostbite, trauma to the extremities or even abuse of sympathomimetic drugs¹⁷⁻²⁰.

Buerger's disease is a potential cause of lower extremity claudication in young patients, usually with the onset of symptoms occurring before the age of 40 or 45¹¹⁻¹⁵. Olin, *et al.* have reported that 7% of 112 patients were older than 60 years old, when first diagnosed⁷. The disease affects both arteries and veins, with typical disease distribution in the upper and lower limbs and, rarely, of the viscera⁵.

Patients may present with symptoms of calf or foot claudication and/or may present with ischaemic extremities⁵. Patients may experience foot claudication within the musculature of the forefoot. This pain may often be mistaken for an orthopaedic problem⁵. As the disease progresses, ischaemic pain at rest and ischemic ulcerations on the toes, feet or fingers may develop¹.

Shionoya, *et al.* have published clinical diagnostic criteria which are generally well accepted^{4,5,10}, although not universally^{4,10} since Buerger's disease remains largely a clinicopathologic entity and there has been a lack of unanimous diagnostic criteria⁴. The distal nature of the disease pattern in Buerger's disease and the involvement of the upper limbs are two major arguments for differentiating Buerger's disease from atherosclerotic arteriopathies²⁰.



Figures 1a and b: Vasculitis affecting the right mid superficial femoral artery (a) and tibiperoneal trunk (b) of a 52-year-old female. Note: Unlike Buerger's disease, the structure of the vessel wall has been affected with thickening of the vessel wall media (indicated by the arrow).



Figures 2 a, b and c: 42-year-old male with Buerger's disease. Note the digital ulceration and ischaemia of the fingers and toes.

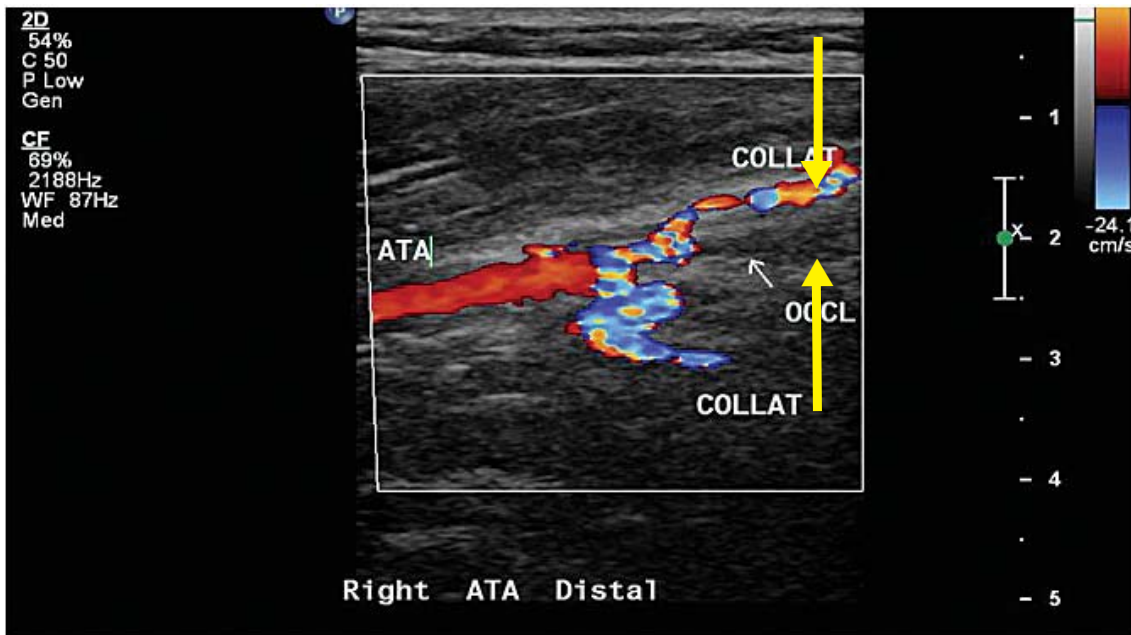


Figure 3a: CDU image of the anterior tibial artery in the distal calf just before it occludes with thrombus and a serpiginous course of adjacent collaterals ("corkscrew appearance").⁵ Unlike vasculitis, the occluded vessel walls remain unchanged (indicated by arrows).

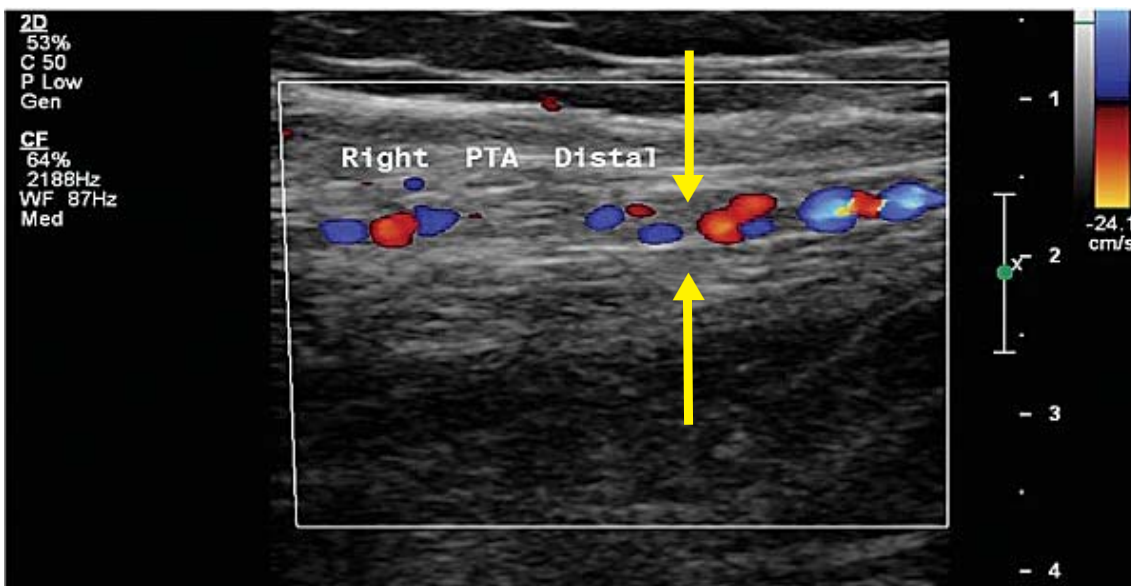


Figure 3b: CDU image of an occluded posterior tibial artery with thrombus and serpiginous, collateral flow in and around the vessel. ("corkscrew appearance").⁵ Unlike vasculitis, the occluded vessel walls remain unchanged (indicated by arrows).

Case study

Figures 2 a, b and c demonstrate the clinical features of a 42-year-old man with known Buerger's disease who had had previous partial right hand digit amputation. He had reduced his smoking but had not ceased completely. He presented to the laboratory

with a painful, ischaemic second toe, calf claudication and impotence. He had right resting ankle brachial index of 0.42 and left ankle brachial index of 0.47. Colour duplex ultrasound (CDU) demonstrated patent iliac, femoral and popliteal arteries bilaterally. He had bilateral occlusion of the posterior tibial,

Buerger's disease: CDU characteristics
Occlusion of distal calf or pedal arteries
Occlusion of forearm, palmar arch or digital arteries
Normal appearing arteries proximal to the occlusive lesions
Serpiginous or corkscrew collateral development at the level of occlusion
Intact vessel wall in the area of thrombotic occlusion, often free of calcification

Table 1

Buerger's disease: clinical diagnostic criteria
Smoking history
Onset before the age of 50 years
Infrapopliteal artery occlusions
Either upper limb involvement or phlebitis migrans
Absence of atherosclerotic risk factors other than smoking

Table 2

peroneal and anterior tibial arteries from approximately the mid calf. Figures 3 a and 3b.

Colour duplex ultrasound and clinical assessment of Buerger's disease

Because of the unusual distribution of vascular lesions in Buerger's disease, it is important to combine a clinical assessment of the patient with the CDU findings.

Obtaining a comprehensive clinical history and clinical examination is of paramount importance, since the early stages of this disease may only affect the digits of the hands or feet. Therefore, generally speaking, performing a standard upper or lower extremity arterial duplex alone may not be diagnostic when the disease is in its early stages, because most standard imaging protocols stop at the ankle level in the lower extremity or at the wrists in the upper extremity.

The major CDU abnormalities that may be detected are shown in Table 1. The clinical factors are presented in Table 2, and include the patient's smoking history and careful examination of the upper and lower limbs of the patient.

Ankle brachial index measurements are recorded at rest and post-exercise in accordance to most standard lower extremity CDU imaging protocols. Patients with Buerger's disease will usually have a reduced ankle brachial index with a further reduction following exercise, unless the disease is distal to the level of cuff placement, which is frequently the case. If the disease is distal to the cuff placement, the ankle brachial index may be normal (0.92–1.0) and it will require digital pressure recordings or photoplethysmographic waveform analysis of the toes. A reduction in pressure and/or a dampened or absent waveform may be demonstrated on one or several digits in the presence of digital ischemia related to the disease.

Buerger's disease in the upper extremity is assessed similarly with a wrist to brachial index measurement followed by digital pressure recordings or photoplethysmographic waveform analysis of the fingers and then CDU surveillance of the upper extremity arteries.

A standard upper or lower extremity CDU of the arteries

in the affected limb should be performed in all cases. When Buerger's disease is present, CDU may demonstrate thrombotic occlusion of the calf or pedal arteries when assessing the lower limb. In the upper extremity, the radial, ulnar, palmar arch or digital arteries may be occluded. In both instances, the vessels proximal to the diseased segment appear normal. The diseased segment will demonstrate thrombotic occlusion and local collateralisation will often appear serpiginous or corkscrewed⁵ in appearance. Other authors have described the angiographic findings as "spiders legs" or "tree roots"¹⁰.

Conclusion

Buerger's disease is a complex entity and the diagnosis of the condition is dependent upon the fulfillment of clinicopathologic criteria which is generally accepted^{4,5,10} but there has been a lack of unanimous diagnostic criteria⁴. The distal nature of the disease pattern and the involvement of the upper limbs are two major arguments for differentiating Buerger's disease from atherosclerotic arteriopathies²⁰.

CDU is a particularly useful imaging tool and can readily identify the distinctively characteristic appearance of vessels affected by Buerger's disease, although the entire clinical picture is of utmost importance.

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