Buerger’s disease (thromboangiitis obliterans): a reversible cause of upper limb digital infarcts

Sir, Buerger’s disease is a distinct pathological entity and disease progression can be reversed by cessation of smoking. Although classically affecting the lower limbs, patients with Buerger’s disease may present with non-specific rheumatic symptoms or isolated upper limb vasculitis. We report a case of Buerger’s disease with an atypical presentation, illustrating the management difficulties this may cause.

A 49-yr-old Caucasian man became unwell with a flu-like illness characterized by malaise, fevers and arthralgia affecting his wrists and knees. These symptoms settled after a 3-week period, but one evening he noted that his left middle finger was painful and purple. There was no obvious precipitant to this, no previous history of vascular spasm, no features of a generalized connective tissue disease and no significant past medical, drug or family history. He had smoked 15–20 cigarettes a day from the age of 18 yr.

On admission to his local hospital, worsening ischaemia of the right index and left middle fingers was apparent. A general examination revealed digital ischaemia with normal sensation, pulse 70 beats/min and regular, blood pressure 110/70 mm Hg bilaterally. All distal pulses were present, and the rest of the physical examination was unremarkable. Investigations were normal FBC/profile; a C-reactive protein (CRP) of 58 mg/l which fell to 4 mg/l 1 week later and remained normal throughout his illness; normal glucose and lipids; negative autoantibody screen (including ANCA); normal complement profile; negative cryoglobulins/hepatitis B and C serology; normal urine; normal echocardiogram/proximal arch aortogram. A presumptive diagnosis of a connective tissue disease with secondary vasculitis was made and prednisolone 30 mg/day commenced. Digital ischaemia progressed, and the prednisolone dose was increased to 60 mg o.d. and azathioprine 150 mg/aspirin 75 mg were added. Five months after presentation he was admitted to Hammersmith Hospital for re-evaluation. Necrotic areas affecting the left middle and ring fingers and large nail fold infarcts affecting the right index and middle fingers were noted. He was still smoking 10–20 cigarettes/day.

A clinical diagnosis of Buerger’s disease was considered and digital angiograms showed characteristic changes with multiple distal artery occlusions and corkscrew collaterals (Fig. 1). The patient was advised to stop smoking and immunosuppression slowly withdrawn. Four months later the patient was completely asymptomatic with healed digits.

The differential diagnosis of upper limb digital infarcts includes primary vasculitis; collagen vascular disease leading to secondary vasculitis or vasospasm; embolic disease; thoracic outlet syndrome; hyperviscosity syndromes; hypothenar hammer syndromes; ergot overuse and Buerger’s disease.

The first case of thromboangiitis obliterans (Buerger’s disease) was described by Felix Von Winiwater in 1879 [1], but it was Leo Buerger in 1908 who described the pathological changes in 11 amputated limbs [2]. Thromboangiitis obliterans is an inflammatory obliterative non-atherosclerotic disease of medium-to small-sized arteries and veins, and less frequently nerves. Initially involving distal vessels of legs or arms (less commonly cerebral/visceral vessels), pathological changes may then progress proximally in a contiguous or skip-like fashion [3]. An inflammatory lymphocytic infiltrate characteristically involves the vessel walls/vasa vasorum, and with time recanalization of thrombus and collateralization may occur [3].

Buerger’s disease predominantly affects males under 50 yr, most commonly those from Asia or the Orient [4]. Although its aetiology appears to revolve around tobacco use, the mechanisms behind this relationship remain unclear [5]. HLA associations have been reported, and physiologically vascular endothelial dysfunction is well described, with high levels of antibodies...
directed against the vascular endothelium detected in both active and quiescent disease states [6, 7].

Presentation is usually with ischaemia or claudication of the legs or less commonly the hands. Rest pain and digital ulceration are common and later sensory abnormalities, Raynaud’s phenomenon, superficial thrombophlebitis and secondary infection occur [5].

The diagnosis of Buerger’s disease is made in a typical patient with a history of exposure to tobacco smoke who presents with ischaemic digital lesions after exclusion of other possible causes.

Confirmatory evidence is then obtained by digital angiography [4]. Characteristically this reveals multiple, bilateral focal segments of stenosis or occlusion, affecting the distal arteries and associated with abundant ‘cork-screw’ collateral vessels (representing dilated vasa vasorum) [8].

The most important aspect of treatment is cessation of smoking (active and passive): the prognosis for those who succeed being generally very good [9]. Additional medical treatment options include the use of aspirin, iloprost and streptokinase, with iloprost (used intravenously), shown to be more effective than aspirin both in the short and long term [10].

Surgical treatment strategies include sympathectomy and vascular bypass procedures, although the use of the latter is limited by the distal nature of the disease. Amputation may be required for advanced cases, or where prior medical/surgical therapy has failed [4].

Consideration of atypical presentations in Buerger’s disease may facilitate early diagnosis and conservative management (abstinence from nicotine), avoiding the use of potentially toxic medical treatments, as in the case described, or surgery.

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