Nonarteriosclerotic Vascular Disease

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KEYWORDS

• Thromboangiitis obliterans • Takayasu autoimmune arteritis • Giant cell arteritis
• Polyarteritis nodosa • Kawasaki disease • Small-vessel vasculitides
• Radiation arteritis • Raynaud phenomenon

KEY POINTS

- Thromboangiitis obliterans, or Buerger disease, is a chronic nonatherosclerotic endarteritis manifesting as inflammation and thrombosis of distal extremity small and medium-sized arteries resulting in relapsing episodes of distal extremity ischemia.
- Takayasu arteritis is a rare syndrome characterized by inflammation of the aortic arch, pulmonary, coronary, and cerebral vessels predominantly presenting with cerebrovascular symptoms, myocardial ischemia, or upper extremity claudication in young, often female, patients.
- Kawasaki disease is a small- and medium-vessel acute systemic vasculitis of young children, with morbidity and mortality stemming from coronary artery aneurysms.
- Microscopic polyangiitis, Churg-Strauss syndrome, and Wegener granulomatosis are systemic small-vessel vasculitides, affecting arterioles, capillary beds and venules, and each presenting with variable effects on the pulmonary, renal and gastrointestinal systems.

THROMBOANGIITIS OBLITERANS (BUERGER DISEASE)

Thromboangiitis obliterans (TAO), or Buerger disease, is a chronic nonatherosclerotic endarteritis manifesting as inflammation and thrombosis of distal extremity small and medium-sized arteries, resulting in relapsing episodes of distal extremity ischemia. It can often lead to digital and limb amputations. Since being described as a distinct entity by Leo Buerger in 1908, the evolving understanding of this disease has ascribed immunologic, genetic, and environmental factors to its cause.[1]

Pathogenesis

Buerger disease has a well-established association with smoking, and there is increasing evidence to suggest an immune-mediated vessel injury from tobacco

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Surg Clin N Am 93 (2013) 833–875
http://dx.doi.org/10.1016/j.suc.2013.04.003 **surgical.theclinics.com**
0039-6109/13/$ – see front matter

exposure. Both smokers and patients with TAO show increased levels of tobacco glycoprotein compared with nonsmokers, and it has been shown that patients with the disease have increased urinary levels of a tobacco metabolite.[2,3] Chewing tobacco may also trigger exacerbations of the disease.[4] Cellular immunity likely plays a role, because endothelial deposition of circulating complexes has been described in this population.[5] In addition, humoral immunity seems to play a role in TAO, because anti-endothelial antibodies, anticollagen antibodies, cytoplasmic antineutrophil cytoplasmic antibodies (c-ANCA), perinuclear antineutrophil cytoplasmic antibodies (p-ANCA) as well as anticardiolipin antibodies have been identified in patients with TAO.[6–9] More recently, endothelial injury with associated antiendothelial cell antibodies has been shown in patients with Buerger disease in clinically unaffected vasculature.[10] There is increasing evidence of a genetic component to the disease, because patients with certain HLA haplotypes have a predisposition to Buerger disease.[2,11,12] The result is a usually episodic acute endarteritis, leading to arterial and venous thrombosis.

Clinical Presentation

The classic presentation is that of distal limb ischemia and digital gangrene in the male smoker in his 30s to 40s, although an increase in women has been described and has been attributed to increased prevalence of female smokers.[11] Although the disease usually manifests as acute ischemia in vessels distal to the popliteal and brachial arteries, involvement of proximal limb, mesenteric, cerebral, coronary, and pulmonary vasculature has been described.[13,14] Patients present with exacerbations of ischemic limb pain and acute ischemia on a background of claudication, resulting in multiple minor and major amputations. Superficial thrombophlebitis and Raynaud syndrome may be found in up to 50% of patients with TAO.[15,16]

Diagnosis

A thorough history and physical examination should be the cornerstone of establishing or, more frequently, excluding a diagnosis of Buerger disease. Patients tend to be young, nondiabetic, and nonhypertensive smokers presenting with palpable proximal pulses but distal limb ischemia without significant atherosclerosis risk factors or history of trauma. Recurrence of disease correlating with failed attempt at smoking cessation may also be a strong diagnostic indicator.

Imaging studies

Noninvasive vascular testing with 4-limb digital plethysmography can confirm the degree of limb ischemia, and if the diagnosis remains unclear or the symptoms severe or limb-threatening, diagnostic arteriography may reveal segmental, abrupt arterial occlusion distal to normal-appearing vessels; infrageniculate vessels may be symmetrically diseased, with characteristic corkscrew collaterals (**Fig. 1**).[17]

Diagnostic criteria systems

Several systems of diagnostic criteria have been developed for TAO, extending the traditional 1983 Shionoya criteria, by which the following 5 findings are necessary for diagnosis: (1) smoking history, (2) onset before age 50 years, (3) infrapopliteal arterial occlusive lesions, (4) either upper limb involvement or phlebitis migrans, and (5) absence of atherosclerotic risk factors other than smoking.[18–20] Mills and Porter developed a set of criteria that include noninvasive vascular testing, and the Japanese Ministry of Health and Welfare Criteria mandates both noninvasive and angiographic findings.[15,21,22] Regardless, it is widely accepted that atherosclerosis and vasculitides of other causes as well as traumatic and embolic causes must be excluded to diagnose Buerger disease. Mills suggests as part of the diagnostic workup to consider a