Thromboangiitis Obliterans

Definition

Thromboangiitis obliterans is inflammatory thrombosis of small and medium-sized arteries and some superficial veins, causing arterial ischaemia in distal extremities and superficial thrombophlebitis. Tobacco use is the primary risk factor.

Symptoms and signs

Includes

1. claudication,
2. non healing foot ulcers,
3. rest pain,
4. gangrene.

Diagnosis: is by clinical findings, noninvasive vascular testing, angiography, and exclusion of other causes. Treatment is cessation of tobacco use. Prognosis is excellent when tobacco use is stopped, but when it is not, the disorder inevitably progresses, often requiring amputation.

Thromboangiitis obliterans occurs almost exclusively in tobacco users (nearly all of them smokers) and predominantly affects men aged 20 to 40; it rarely occurs in women. It occurs more commonly in people with HLA-A9 and HLA-B5 genotypes. Prevalence is highest in Asia and the Middle East.

Pathophysiology

Thromboangiitis obliterans produces segmental inflammation in small and medium-sized arteries and, frequently, in superficial veins of the extremities. In acute thromboangiitis obliterans, occlusive thrombi accompany neutrophilic and lymphocytic infiltration of the intima; endothelial cells proliferate, but the internal elastic lamina remains intact. In an intermediate phase, thrombi organize and recanalize incompletely; the media is preserved but may be infiltrated with fibroblasts. In older lesions, periarterial fibrosis may occur, sometimes affecting the adjacent vein and nerve.

Causes

The cause is unknown, although cigarette smoking is a primary risk factor. The mechanism may involve delayed hypersensitivity or toxic angiitis. According to another theory, thromboangiitis obliterans may be an autoimmune disorder caused by cell-mediated sensitivity to types I and III human collagen, which are constituents of blood vessels.

Symptoms and Signs

Symptoms and signs are those of arterial ischemia and superficial venous thrombosis. Some patients have a history of migratory phlebitis, usually in the superficial veins of a foot or leg. Onset is gradual, starting in the most distal vessels of the upper and lower extremities with coldness, numbness, tingling, or burning. These symptoms may develop before objective evidence of disease. Raynaud syndrome is common. Intermittent claudication occurs in the affected extremity (usually in the arch of the foot or in the leg; rarely in the hand, arm, or thigh) and may progress to rest pain. Frequently, if pain is severe and persistent, the affected leg feels cold, sweats excessively, and becomes cyanotic, probably because of sympathetic nerve over activity. Later, ischemic ulcers develop in most patients and may progress to gangrene.

Pulses are impaired or absent in one or more pedal arteries and often at the wrist. In young men who smoke and have extremity ulcers, a positive Allen test (the hand remains pale after the examiner simultaneously compresses the radial and ulnar arteries, and then alternately releases them) suggests the disorder. Pallor with elevation and rubor with dependency frequently occur in affected hands, feet, or digits. Ischemic ulceration and gangrene, usually of one or more digits, may occur early in the disorder but not acutely. Noninvasive tests show greatly decreased blood flow and pressure in the affected toes, feet, and fingers.

Diagnosis

- Other causes of ischaemia excluded by testing
- Angiography

History and physical examination suggest the diagnosis. It is confirmed when

- The ankle-brachial index (ratio of ankle to arm systolic BP) for legs or segmental pressures for arms indicates distal ischemia
- Echocardiography excludes cardiac emboli
- Blood tests (eg, measurement of antinuclear antibody, rheumatoid factor, complement, anticentromere antibody, anti-SCL-70 antibody) exclude vasculitis
- Tests for antiphospholipid antibodies exclude antiphospholipid antibody syndrome (although these levels may be slightly elevated in thromboangiitis obliterans)
- Angiography shows characteristic findings (segmental occlusions of the distal arteries in the hands and feet, tortuous, corkscrew collateral vessels around occlusions, and no atherosclerosis)

Treatment

- Smoking cessation
- Local measures
- Sometimes drug therapy

Treatment is cessation of tobacco use. Continuing to use tobacco inevitably leads to disease progression and severe ischemia, often requiring amputation.
Other measures include avoiding cold; avoiding drugs that can cause vasoconstriction; and avoiding thermal, chemical, and mechanical injury, especially that due to poorly fitting footwear. For patients in the first phase of smoking cessation, iloprost 0.5 to 3 ng/kg/min IV infusion over 6 h may help prevent amputation.
Pentoxifylline, calcium channel blockers, and thromboxane inhibitors may be tried empirically, but no data support their use. Use of antiendothelial cell antibody measurements to follow the course of disease is being studied. When these options fail, lumbar sympathetic chemical ablation or surgical sympathectomy can alleviate ischemic pain and enhance ulcer healing in about 70% of patients with an ankle-brachial pressure index ≥ 0.35 and no diabetes mellitus.

Key Points

Thromboangiitis obliterans is inflammatory thrombosis of small and medium-sized arteries and sometimes superficial veins in distal upper and lower extremities.

It occurs almost exclusively in male smokers aged 20 to 40.

Claudication may occur, and patients may develop ischaemic ulceration and gangrene of one or more digits.

Diagnose clinically but exclude other causes of ischemia by testing.

Smoking cessation is essential; iloprost infusion may help prevent amputation, but there is little evidence supporting use of other drugs.