Case Report

Buerger’s disease: a case report
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Abstract
Buerger’s disease is an occlusive vasculopathy affecting small- to medium-sized blood vessels. The disease usually affects smokers. Clinically, it is characterized by features related to ischemia. If untreated gangrene and amputation may supervene. A wide variety of therapeutic options ranging from vasodilators to sympathectomy are useful. We, herein, report an adult female with this disease, who underwent sympathectomy for relief of her symptoms.

Key words
Buerger’s disease, sympathectomy.

Introduction
Buerger’s disease (thromboangiitis obliterans) is an inflammatory occlusive disease affecting medium and small arteries and veins. Most patients are men between 25 and 45 years of age. It is an episodic and segmental inflammatory process with thrombosis of peripheral vessels. The cause is not clearly known. Smoking is found to be a major risk factor.

The prevalence is higher in Asians and individuals of Eastern European descent. In the initial stages, polymorphonuclear leukocytes infiltrate the walls of small and medium-sized arteries and veins. The internal elastic lamina is preserved, and thrombus may develop in vascular lumen. As the disease progresses, mononuclear cells, fibroblasts, and giant cells replace the neutrophils.

Buerger’s disease is characterized by occlusion of distal arteries producing claudication, pain, and tissue necrosis. In the presence of severe digital ischemia, trophic nail changes, painful ulcerations, and gangrene may develop at tips of fingers or toes.

There is no definite treatment modality available for Buerger’s disease. Analgesics and vasodilators can provide symptomatic relief to the patients. In refractory conditions, like persistent pain and non-healing ulcers, sympathectomy is a useful treatment modality.

Case history
A young female aged 20 years, presented in skin outpatient with multiple necrotic ulcers on her both feet for four years. Her complaints started with pain in her both legs when she used to walk for few hundred yards. Gradually pain aggravated and she felt it even at rest. There was no history of fever, joint pains, GI tract, respiratory or any other systemic symptoms. Along with intermittent claudication, there were attacks
of numbness and bluish discoloration, more prominent in her left foot, which used to get worse in cold weather.

Almost one year after the onset of symptoms, she developed small painful punched out ulcers on dorsum of left foot and tips of left toes. Ulcers showed poor response to antibiotics, local dressings and analgesics. There was no history of intake of drugs like beta-blockers, methysergide etc., which could be related to her symptoms. She smoked 2-3 cigarettes per day for the last five years.

Cutaneous examination revealed bluish red, diffuse dusky erythema on dorsum of both feet extending up to tips of toes. It was more prominent on left foot. There were multiple, small punched out necrotic ulcers of 1 to 1.5 cm size, present on tips of big toe, 4th and 5th toes of left foot. Pulse examination revealed absence of dorsalis pedis and posterior tibial pulses in left foot. Pulses were feeble in her right foot. Her blood counts, urine examination, blood urea nitrogen, serum creatinine, liver function tests and ANA were in normal limits. ECG and echocardiography revealed no abnormality. Angiography was performed and showed distal anterior and posterior tibial arteries narrowing with almost complete obliteration in the distal third of left leg. There were typical corkscrew configurations of collateral vessels originating from occluded vessels. She was treated with analgesics, calcium channel blockers (nifedipine) and regular dressings for ulcers. Symptoms didn’t improve significantly after one month of treatment.

Later an alternative treatment with sympathectomy was considered and Surgical Unit in Jinnah Hospital, Lahore was consulted. She was shifted to surgery unit and sympathectomy was done. A transverse incision made from mid axillary line to lateral border of left rectus muscle. After division of external oblique, internal oblique and transversus abdominis, peritoneum was reflected medially. Sympathetic chains with its ganglia were identified and four ganglia from L1 to L4 were completely excised along with connecting chains.

Three weeks later, there was significant relief in pain and almost complete healing of ulcers with atrophic scars, she had no further attacks related to Raynaud’s phenomenon. She was followed for six months with no further vasospastic attacks and painful ulcers in her feet.

Discussion

The pathogenesis of Buerger’s disease is still not clear, whether, it is an inflammatory or occlusive disease, as it combines features of both. Infections, hormonal influences, cold weather and genetic predisposition have all been blamed.³

Arteriosclerosis, endothelial proliferation and thrombosis leading to ischemic ulcerations and necrosis, are the pathogenic features. Fewer than 20% of patients are women, with smoking as an aggravating factor in both males and females. Cessation of smoking is imperative and the disease can remain active with as little as a single cigarette a day.⁴
Diminished sensation from ischemic neuropathy is present in over 70% of patients at presentation. Proximal pulses are normal while distal pulses are diminished or absent. Ulcers are present in 75% of patients.

Various differentials are considered in a patient of Buerger’s disease. Arteriosclerosis is similar, but with greater involvement of larger vessels and with associated internal disease. In addition, it usually becomes apparent at a later age, usually above fifty years. Damage is far less extensive regarding ischemic ulcers on feet in Raynaud’s disease and frequent systemic involvement in association with connective tissue disease. Arteriography is still an important investigation in suggesting the diagnosis of Buerger’s disease, showing very tortuous or corkscrew-like collateral vessels, somewhat similar to diabetic collateral vessels.

Non-ulcerated skin should be kept moisturized. Warm pads must be used judiciously, as burns are possible in these patients with diminished sensation. Pain is extreme and analgesics are usually given along with vasodilators, like calcium-channel blockers to prevent vasospastic attacks. The disease shows resistance to most of the treatment modalities. Sympathectomy may at least reduce the vasospastic manifestations of disease and aid in the establishment of collateral circulation to the skin. It is indicated for relief of intractable rest pain and healing of ulcers refractory to other treatment. Sympathectomy is classically carried out by supraclavicular method. Sympathetic trunk is traced downwards and divided below the third thoracic ganglion. All rami communicate associated with the second and third ganglia, are meticulously divided. Buerger’s disease is a progressive problem with no definite and effective therapy. Prognosis is dependant on the success of smoking cessation. Though Buerger’s disease afflicts more commonly males, yet it can affect females also, with smoking found to be a common factor in both.

References