Radial Artery Occlusion, A Rare Presentation of Behcet’s Disease

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Abstract
Behçet’s disease (BD) is a multi-system inflammatory disorder which presents with recurrent orogenital ulceration, uveitis, and erythema nodosum. Medium vessel vasculitis of upper limb is extremely rare and it is only reported in patients with Behcet’s disease on long follow up. Mean duration from diagnosis of disease to development of vasculitis is 5.8 years. We present a patient who presented with gangrene of fingers with absent radial pulse and during course of his illness he developed features of Behcet’s disease. Diagnosis was established by clinical features and histopathology and patient was treated with steroids and colchicine.

INTRODUCTION
Behçet’s disease (BD) is a multi-system inflammatory disorder that presents with recurrent oral ulcers, genital ulcers, uveitis, and erythema nodosum. The cause of Behcet’s disease remains unknown, although an autoimmune reaction triggered by an infectious agent in a genetically predisposed individual has been suggested. Vasculitic lesions in Behcet’s disease affect arteries and veins of various sizes. Upper limb arteritis is extremely rare; in various series, a total of nine patients have been reported with upper limb artery involvement.¹⁻⁴ We report a case of Behcet’s disease with radial artery occlusion which is not a known common site of vascular involvement.¹⁻⁴

CASE REPORT
A 47 year old male, electrical engineer by profession presented with numbness, throbbing pain and blackish discoloration of left thumb, index and middle finger for 4 days. This discoloration started from distal part of digits and was progressing proximally. He also had history of giddiness 10 days prior to this presentation. History of recurrent oral ulcers was present. There was no history of skin rash, edema, genital ulcers, abdominal pain, diarrhea, bleeding from any site, neurological deficit or psychiatric symptoms. There was no other significant past history. There was no history of alcohol consumption and smoking. On examination the left radial artery was not palpable. Left dorsalis pedis was weakly palpable and an area of prominent pulsation was present in right post tibial artery territory (? Small aneurysm).

Hematological values were normal except TLC: 24500/mm³. Renal, hepatic, biochemical parameters and serum lipid profile were normal. Rheumatoid factor, anti nuclear antibodies and anti ds DNA were negative. USG of abdomen was normal. Arterial Doppler of upper limb revealed marked intimo medial thickening & narrowing of left radial artery with only trickle of intra luminal flow with collaterals along course of artery and good velocity flow in left ulnar artery.

Possibility of Takayasu’s arteritis was thought of and the patient was started on steroids. Subsequent MR angiography showed abrupt cut off of left radial artery 2 cm distal to its origin. The findings excluded Takayasus’s arteritis. As MR angiography was inconclusive, angiography of all four limbs was done. It showed that the left radial artery was cut off after its origin and formed collaterals thereafter (Fig. 1).

Angiography of the lower limbs revealed that both left and right anterior tibial arteries were cut off after their origin without any collaterals. Radial artery biopsy was advised but patient refused for same.

On follow up after 3 weeks of initial presentation patient developed aceneform lesions and recurrent large penile ulcer which were healing with scar formation. Finally patient agreed for radial artery biopsy and histopathology (Fig. 2) revealed lumen completely occluded by a thrombus containing lymphomononuclear cells and occasional giant cells. The thrombus was undergoing organization with recanalisation. There was no fragmentation of internal elastic lamina. Lymphomononuclear infiltration of vessel wall was present. No fibrinoid necrosis was seen in multiple sections examined. Pethargy test was negative, but he was on steroids that time. Diagnosis of probable Behcet’s
disease was made on the basis of oral and genital ulcers. Patient received colchicine in addition to steroids and anticoagulants. There is no further progression of disease on follow up of 6 months.

**DISCUSSION**

Behçet disease (BD) is a multisystem disease characterized by skin and mucosal lesions, musculoskeletal, ocular, gastrointestinal, neurological and vessel involvement. Usual presentation of a Behcet’s patient is orogenital ulcers, uveitis and skin manifestations. Vessel involvement in Behcet’s is well established, the mechanism of which is not known. Incidence of vascular involvement in Behcet’s disease is 7-29%. Arterial involvement can occur in the form of aneurysm formation (common), pseudo aneurysm or occlusion (rare). Arterial occlusion can cause infarction, organ failure, and weakness of legs and arms while rupture of aneurysm may be fatal. Small vessel vasculitis (veins & capillaries) is common and accounts for most of the pathologic process in Behcet’s disease. Majority of arterial complications show mean delay of 5.8 years from disease onset with predominant involvement of lower limb arteries. Behcet’s disease affects large arteries like aorta or femoral arteries. Upper limb vessels (subclavian/axillary) involvement is rarely reported. The most frequently affected vessels, in decreasing order, are pulmonary, femoral, popliteal, subclavian, and carotid arteries. In all previously published series vasculitis developed in the patients with a long history of the disease but our patient presented with arterial occlusion that too of radial artery which is an extremely rare presentation of Behcet’s disease.

Histopathological studies in Behcet’s disease demonstrate nonspecific vasculitis with mononuclear cell or neutrophilic infiltrates, endothelial proliferation, destruction of elastic lamina, fibrinoid necrosis and thrombus formation.

Combined treatment with glucocorticoids, anticoagulants, antiplatelets and immunosuppressants are effective in arterial occlusions. Aneurismal lesions sometime require surgical correction. For all patients presenting with digital gangrene or compromised peripheral circulation Behcet's disease should be considered in differential diagnosis.

**REFERENCES**