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Case Report Open Access

Buergers Disease with Severe Eosinophilia and Acute Critical Ischemia

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Abstract

Buerger's disease usually presents with normal blood tests. We present a case with significant eosinophilia, a phenomenon very rarely reported.

Eosinophilia with acute onset Raynaud's and critical ischemia can be atypical early manifestation of Buerger's disease (Thromboangiitis obliterans).

Keywords: Buerger's disease; Superficial thrombophlebitis; Arterial upper limb involvement; Raynaud's phenomenon; Echocardiography

Introduction

Buerger's disease (Thromboangiitis obliterans-TAO)

This has been recognized as a form of vasculitis which has a strong evidence-based relation with smoking [1-15]. Classically occlusive segmental and often multiple inflammatory lesions of medium-sized and small arteries and superficial veins are observed. It accounts for roughly 0.5%–5% of patients hospitalized for arterial occlusive disease in Europe and up to 16% of such patients in Japan (higher prevalence in the world). Actual and definite prevalence worldwide is difficult because of the absence of general diagnostic criteria; however, it affects all races. Some data has been suggestive of a genetic predisposition (Higher incidence noted in Ashkenazi than in non-Ashkenazi Jews in Israel). In the acute phase presentation, all vessel walls are involved with inflammatory changes. Although it still is classified with vasculitis, unlikely a true form as disputed over that worldwide. TAO can be distinguished from other types of vasculitis by:

a. The tendency to occur in young males

b. Strong association with tobacco consumption

c. The rarity of systemic signs and symptoms

d. A highly cellular thrombus with relative sparing of the blood vessel wall

e. The absence of elevated acute-phase reactants

f. Absent immunological markers

A reasonable criterion which has helped in identifying patients with this rare and interesting form of the disease can be summarized as below.

Diagnostic criteria for Buerger's disease (Adar, Mozes, Fiessinger)

(Definitive diagnosis of Buerger's disease is retained in young smokers with distal lower limb ischaemia)

with at least two of the following three symptoms After the exclusion of the following

1) Superficfiafl ftThrombopThflebfiftfis – Dfiabeftes meflflfiftus

2) Arfterfiafl upper flfimb finvoflvemenft – AftTheromaftous flesfions

3) Raynaud's pThenomenon – Poftenftfiafl source off emboflfism

– Enftrapmenft syndrome

– Aufto-fimmune dfiseases

– Myefloproflfifferaftfive syndrome

– Hypercoaguflabfiflfifty sftaftes

The presence of any one of these symptoms renders the diagnosis probable.

Case Report

A case report of a 24-year-old male, smoker, presented acutely, complaining of cold, painful hands with discoloration for 2 weeks (whitish) worsening for 48 hours. On examination, apart from features of Raynaud's and tender fingers, he had good perfusion as shown on by Doppler testing. Blood tests showed eosinophilia; 13000/µL (Normal 0.04-0.40 × 10*9) and mild raised C-reactive protein 24(Normal range <10.0 mg/L). Chest radiograph and echocardiography showed no abnormalities. The results of vasculitic screening including Immunoglobulin's, complements, ANA, ANCA, RF, APLA and CCP were all either normal or negative. As there were no critical features of ischemia, the patient was discharged with a plan for close review. Smoking cessation was advised. He was re-admitted within a week with worsening of Raynaud's and features of critical ischemia with bluish nail and fingertip with some early features of mild digital necrosis. Treatment was commenced with intravenous heparin, aspirin and alprostadil. The patient's digital ischemia and intermittent claudication showed some improvement after 1 week, but eosinophilia persisted at 11000 with ruminant pain in digits. The importance of smoking cessation was re-emphasized. Magnetic Resonance Angiography showed no occlusion of the distal arteries of upper and lower extremities. Capillaroscopy showed micro hemorrhages (No CTD/Scleroderma pattern abnormality). Investigations for parasitic infections and malignancy were negative. HIV/Hepatitis B/C CMV/EBV/Parvovirus IgM was all undetected. Schistosoma testing was negative. He was tested for a rare possibility of the genetic mutation for Hypereosinophillic syndrome (PDGFRA and FIP1L1 genes) which was negative and underwent bone marrow biopsy which was also normal.

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He was started on oral steroids 1mg/kg. Within 2 weeks of initiation of steroids, his eosinophilia resolved to normal levels with no other worsening features of critical ischemia or Raynaud's. He was able to stop smoking with some help. His aspirin was continued along with allopurinol (some evidence of its use in preventing free radical ischemic injury).

He was improving in follow up with no relapse of any symptoms. His steroids were tapered down with a close follow up review. We have followed him up for another year and he has remained symptom-free with no ongoing concerns.

Symptoms

The main classic and well recognized initial presentation of such patients is Ischemic manifestations of the lower limbs. Claudication in the arch of the foot can be an early sign and is suggestive of, or sometimes even specific to TAO.

In 70%–80% of cases the diagnosis is not made until the critical ischemia phase, with rest pain and ischemic ulcerations of the toes or feet. Rest pain generally occurs on the forefoot, causing continuous pain and obliging patients to sleep with their legs dangling downwards. Super infection often occurs and the lesions progress towards necrosis and distal gangrene. Raynaud's phenomenon can be seen typically and is generally unilateral. It is observed in half the number of such patients. Interestingly evidence of superficial thrombophlebitis is also observed in 40-60 per cent of such cases. Deep Venous Thrombosis is generally not a hallmark of this disease and if present should warrant an alternative cause especially Behcets disease. Moreover, in another 10-12 per cent of patients features of palindromic arthritis has been seen.

As mentioned earlier smoking and constituents of tobacco have been found as the single most important causative agent for this and remain the cornerstone in considering stopping it in all patients to avoid treatment failures and risk of multiple amputations.

Supportive treatment involves lines of treatment for critical ischemia and to prevent complications related to that.

Discussion and Conclusion

Eosinophilia has only rarely been reported as a feature of the presentation of Buerger's disease. Its presence does not necessarily imply a systemic vasculitis or related syndrome. Although there is limited evidence for the use of steroids in this condition, the presence of inflammatory features such as eosinophilia could strengthen the rationale for using steroids in certain cases by halting the ongoing immune driven inflammatory cascade. The presence of eosinophilia might suggest alternative pathogenic mechanisms in certain cases of Buerger's.

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