Case Report

Behcet's Syndrome With Coronary Arteritis

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Introduction:
Behcet's Syndrome is a systemic inflammatory disease having a chronic and prolonged course with appearance of 4 major symptoms: oral and genital ulceration, eye disease and cutaneous manifestation as well as other multi system involvement such as large joint arthropathies, central nervous system involvement, gastrointestinal ulceration and thrombosis of major vessels[1,2].

The vascular system is often involved, most frequently on venous side, with disorders such as superficial thrombophlebitis and deep vein thrombosis[2,3]. Arterial involvement is rare complication in Behcet's Syndrome.

Case Report:
Forty nine years old African American Female with Past medical h/o HTN, Behcet's Syndrome since 1988 with recent flare up of disease with oral and genital ulcers, headache, arthralgia and gastrointestinal symptoms was referred to cardiology clinic with c/o chest pain for 4 days, reterosternal, non radiating,4/10 in intensity, no aggravation on exertion, relieved by SL nitroglycerine associated with mild SOB, nausea and diaphorersis. Patient denied tobacco, alcohol and illicit drugs. Never had similar chest pain before. Review of system was unremarkable except for Behcet's Syndrome flare up.

Her medications were tenolol, Valsartan, Indocin, ASA, Lortab, Zoloft, Premarin, Triamatrene and Tagamet. Her family history was negative for coronary Artery disease.

On physical exam pt was afbrile, heart rate 52/min,blood pressure 140/90,respiratory rate 16/min average built female 3-4 oral mucosal ulcers. Eye exam was unremarkable. Cardiac exam showed normal S1 and S2,no added sounds. Abdomen was soft, non tender and no hepatosplenomegaly. Her Right knee and both ankle joints were mildly tender without signs of effusion. Her genital exam revealed 2-3 small painful ulcers on labia minora. Rest of systemic exam was unremarkable.

Course of Illness:
Laboratory data showed normal basic metabolic profile and cell count with differential. EKG showed normal sinus rhythm with no acute T-wave or ST changes. Myocardial Perfusion study was done which showed Stress induced ischemia at the apex, extending a short distance proximally along the anterior wall. EF was 65%. Coronary Angiography showed normal coronaries and normal LVSF. Patient was started on steroid for about 2 weeks and her chest pain resolved along with remission of behcet's syndrome flare up. Myocardial perfusion study was repeated about 3 month after of resolution of chest pain which turned out to be negative for ischemia.

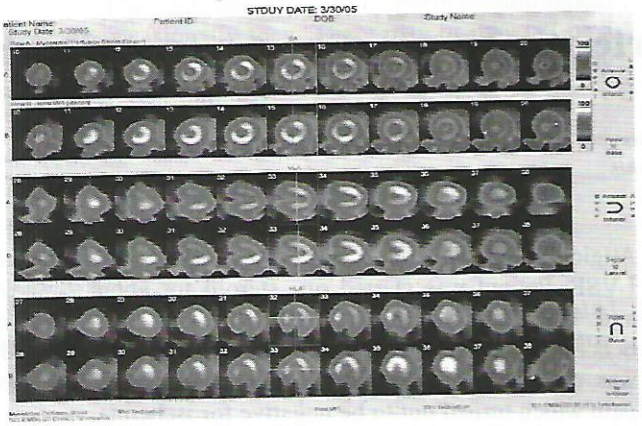
Discussion:
In my patient with about 15 years history of behcet's syndrome presented with her typical flare -up with new symptom of chest pain. Diagnosis of coronary arteritis was made for following reasons:

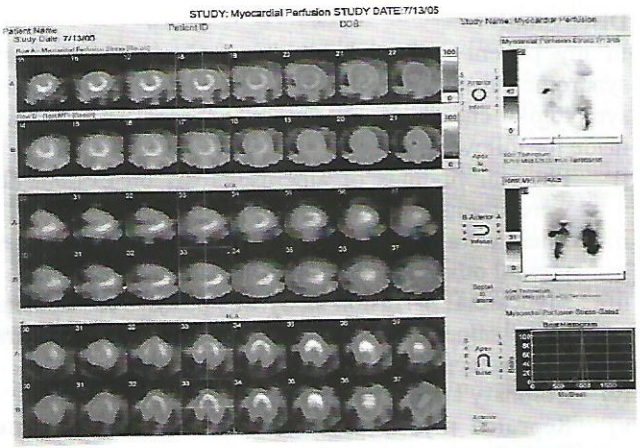
i) Physical findings of atherosclerosis on examination were absent.
ii) There were no risk factors for coronary artery disease present other than the HTN.
iii) Myocardial perfusion study was positive for ischemia at the same time with exacerbation of other systemic and mucocutaneous manifestations.
iv) Coronary angiography was normal.
v) Repeated myocardial perfusion study was negative for ischemia after resolution of acute flare up.

Myocardial infarction is rare in behcet's syndrome with only 10 cases reported so far in English literature since 1980[4,11]; these 10 patients developed myocardial infarction at a relatively young age(22-39 years) and 6 had no eye involvement as in my case. In the vascular lesions, aneurysm was found in 5 patients[5,7,8,10,11].

Coronary artery occlusion inn Behcet's syndrome is possibly due to vasculitis of coronary arteries[4,6,7,9,10]. There has been a report that histopathologically confirmed destructive changes of the coronary artery consistent with arteritis[4]. Other investigators have also estimated that coagulation disorder or abnormal response of vascular endothelium involved in cardiovascular manifestation of Behect's syndrome[12]. In the present case, prompt resolution of the symptoms provided by corticosteroid therapy, absence of atherosclerotic changes in coronary angiography and negative repeat myocardial perfusion study support arteritis as the possible etiology of chest pain.

In conclusion, Anginal symptoms and myocardial infarction are rare in Behcet's syndrome, but should be regarded as a clinically significant complication because it often leads to poor prognosis.





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