Behcet’s Syndrome Presenting as FUO

Sir,

Behcet’s syndrome (BS) usually comes in the domain of dermatologist, where it presents with recurrent oral and/or genital ulcers.1 Its encounter with physicians is uncommon. Fever may be one of the symptoms but Behcet’s syndrome, presenting as prolonged pyrexia is rare. We report an interesting case of classic fever of unknown origin who was followed for 3 months without any diagnosis. She was hospitalized for more elaborate investigations and later developed full fledged manifestations of Behcet’s syndrome. The disease responded well to treatment, only to recur after two months due to drug default. She was again treated successfully.

A 16 years old girl presented in emergency with H/o moderate to high grade fever with chills and rigors for three months. She had consulted few local practitioners with partial relief and without any diagnosis. One week prior to admission she developed dysentery and vomiting with symmetric inflammatory arthritis.2 Examination revealed dehydration, aphthous ulcers and redness in left eye. She was investigated for the common causes of fever in that particular setting. Cultures were sent and bone marrow aspiration was done. Patient was given symptomatic treatment. The investigation reports remained inconclusive as the positive findings were elevated leucocytes, 13,000 per cmm, raised ESR-64mm and positive CRP. Later she developed itching and pain in the vulva. On examination there were multiple 2-3mm tender, inflammatory ulcers. Aphthous ulcers increased in number. She also developed erythema nodosum over the skin of tibia bilaterally 6-7 in number and painful to touch.2 Next day a red nodule appeared at the corneal margin in the left eye which was diagnosed as non-necrotizing, nodular scleritis. Pathergy test,1 a specific and sensitive test for diagnosis of BS was positive.

She was diagnosed as Behcet’s syndrome and was given dexamethasone, colchicine dapsone and local treatment for ulcers. Improvement started within 2 days. She was relieved in 2 weeks except for the scleral nodule. Patient had a recurrence after 2 months. She gave a history of drug default and was again treated on the same lines and was relieved.

Behcet’s syndrome is primarily a systemic vasculitis syndrome1,2 and is a rare cause of FUO. It is characterized by recurrent episodes of oral and genital ulcers, iritis and cutaneous lesions. Severe complications include major vessel thrombosis, CNS involvement and blindness.1,2 Diagnosis depends on typical clinical findings.1

BS follows a peculiar geographical distribution along the old silk route and there are only a few reports of the disease from northern India. Possibly because the disease is either under-diagnosed or underreported. These cases occur sporadically3 and can be detected if index of suspicion is high. The lesson learnt from this case is, that BS must be suspected in all the cases who present with orogenital mucosal ulcerations, scleritis/ uveitis and also considered in the differential diagnosis of FUO. Early diagnosis and recognition of Behcet’s syndrome should prevent the uncommon fatalities and sight threatening complications. Treatment in most patients of Behcet’s syndrome is easy and usually successful.

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References