

An Unusual Cause of Fever

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Abstract

Behcet syndrome is an autoimmune multisystem disorder often affecting young adults and its pathological origin is unclear. Here we present a case report of a 24 year old male who presented with high grade fever with orogenital ulcers. On evaluation, diagnosis of Behcet syndrome was made based on clinical presentation, positive pathergy test and HLA B51.

Case Report

A 24 year old male presented with complaints of high grade intermittent fever for 45 days duration. He also gives history of development of painful ulcers in the groin region which initially started as a papule and later burst open to form an ulcer. He also had multiple painful oral ulcers for 10 days duration. He reported to have significant weight loss of ~ 15 kg in one month. He was treated outside with multiple antibiotics for fever. There was also history of similar illness 1 year back which resolved with treatment and treatment details were not available. There was no significant family history. He is a smoker and a social drinker too. On examination he was febrile-101.3°F, tachycardic with pulse rate of 102beats/min, BP was 110/70mm of Hg and systemic examination was unremarkable. Examination of oral cavity revealed multiple apthous ulcers



Fig. 1: Ulcers in the oral cavity

(Figure 1).

He also had three well circumscribed ulcers, one in the root of the right scrotum and other two similar ulcers in left inner thigh (PANEL B) with minimal serous discharge without lymphadenopathy.

Routine blood investigations revealed elevated total counts of 16,530 cells/cu.mm with neutrophilic predominance, Hb-19.9g/dl, PCV-53.7%, RBC count-6.14million cells/cu.mm and platelet counts of 3.71lakhs/cu.mm. Repeat haemoglobin showed decreasing trend. Renal parameters were normal. Blood and urine cultures were sterile. Routine fever work up such as MPQBC, typhoid IgM, Scrub typhus IgM, leptospira IgM were negative. HRCT Chest and CT abdomen



Fig. 2: Well circumscribed ulcers in the scrotum and groin

were normal. HIV ELISA, VDRL and TPHA were negative. ANA and Rheumatoid factor were also negative. Echo showed no regional wall motion abnormality, normal LV systolic and diastolic function with LV EF-66%, no evidence of vegetation. Edge biopsy of the scrotal ulcer was done revealed severe subacute inflammation.

Pathergy test was done which was positive –pustules were formed at the site of needle prick (PANEL C).

HLA B51 was positive which confirmed the diagnosis of BEHCET syndrome. Consultant physician of infectious diseases opinion was obtained and concurred with the treatment.

He was treated with IV fluids, antipyretic, oral antibiotics and later started on oral colchicine. Regular follow up showed resolving oral and genital ulcers and patient remained afebrile.

Two months later, he presented with acute onset of headache and blurring of vision in the right eye for one week duration. On examination of the central nervous system revealed right lateral rectus palsy with bilateral



Fig. 3: Positive Pathergy test

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papilledema. Perimetry showed visual field defects in the right eye. MRI brain with MRV showed thrombosis of superior sagittal sinus and medial aspect of transverse sinuses with bilateral papilledema. He was then initiated on oral anticoagulation and therapeutic INR was maintained. Consultant Rheumatologist concurred with the same line of management. He showed gradual improvement in his neurological signs in subsequent follow ups.

He later developed swelling of left lower limb which he left untreated. He was non compliant with oral anticoagulation therapy. He presented with complaints of painful swelling of the right lower limb. Doppler study of the venous system of both the lower limbs showed acute DVT of the right lower limb and chronic DVT of the left lower limb. His haematological evaluation showed persistently elevated haemoglobin levels with increased PCV for which consultant Haematologist opinion was obtained. He advised venesection for persistent polycythaemia (Likely secondary polycythaemia). Coagulation profile was normal. His thrombophilia workup was negative for antiphospholipid and anticardiolipin antibodies with increased serum homocysteine levels (21.29micromol/L) while evaluation of other thrombophilia parameters were planned later. He was restarted on oral anticoagulation therapy along with B complex and folic acid therapy.

Discussion

Behcet's disease is an inflammatory disorder of unknown etiology, characterized by recurrent oral aphthous ulcers, genital ulcers, uveitis

and skin lesions.¹ Initial cases of Behcet's syndrome were more commonly seen along the Mediterranean region, middle east and far east region along the ancient silk route, also called as silk road disease, with highest incidence of cases in Turkey.^{2,3} It commonly affects the people between 20 to 40 years of age; both the sexes were equally affected with more severe form of disease in males. The aetiology of Behcet's syndrome remains unknown. It is believed to be the combined effects of immune mechanisms and environmental factors resulting in secretion of cytokines that stimulate inflammation-inducing immune cells.^{3,4} HLA B 51(B510101) is the most common genome associated with Behcet's disease occurring in 60% of the patients.^{4,5}

Revised International Criteria for Behcet's Disease^{4,5}

Diagnosis of Behcet's syndrome was made based on the above criteria if a score of 3 points is obtained based on the parameters below:

1. Oral aphthosis: 1 point
2. Skin manifestations (pseudofolliculitis, skin aphthous): 1 point
3. Vascular lesions (Phlebitis, large vein thrombosis, aneurysm, arterial thrombosis): 1 point
4. Positive Pathergy test: 1 point
5. Genital aphthosis: 2 points
6. Ocular lesions: 3 points

The hallmark of Behcet's syndrome was the occurrence of recurrent oral and genital ulcers which were the most common presentation (97% cases).² Multiple painful mucosal ulcerations appear on the nonkeratinised oral

mucosa in the tongue, pharynx, buccal and labial mucosal membranes, usually covered with a yellowish pseudomembrane and heals without scarring and/or lymphnode swelling.^{4,6} Genital ulcers are usually localised on the scrotum in males and vulva and vagina in females. Pathergy test is positive in the patients with Behcet's disease which signifies the neutrophilic vascular reaction (appearance of erythematous papules or pustules) at the site of needle prick after 24 to 48 hours constitutes one of the diagnostic criteria.^{4,5} Severe manifestations of the disease involve the eyes (most common), the CNS, the main large vessels and the gastrointestinal tract causing major morbidity. Corticosteroids are used to control inflammation while immunosuppressive agents were used for second line therapy.^{4,5} Newer treatment modalities are targeted against cytokines such as TNF-alpha, IL-21 and IL-17.^{4,5} A strong clinical suspicion, early diagnosis and stepwise approach helps to control the disease process and helps to reduce the disease related as well as treatment related morbidity and mortality.

References

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