Behcet’s Syndrome Associated Manifestations and its Treatment Response: A Case Report

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Behçet’s syndrome (BS) or Behcet’s disease (BD) is a chronic multisystem disorder of unknown etiology that is characterized mainly by recurrent mucocutaneous, articular, neurological, gastrointestinal, and ophthalmological lesions. A 20-year-old unmarried, Hindu, male patient presented with a history of recurrent oral and genital ulcers, large joint inflammatory oligoarthritis predominantly in right knee joint, folliculitis (dermatographia) on the legs, epididymitis, acneiform lesions over face, and panuveitis with associated constitutional symptoms. His Laboratory examinations were nonrevealing except for a moderate rise in erythrocyte sedimentation rate and C-reactive protein. This patient meets, in 1990, the International Study Group criteria for BD/BS. The main clinical manifestations of this disease are discussed and its early recognition and treatment response is emphasized.

Keywords: Arthritis, Behcets syndrome, Folliculitis, Orogenital ulcers, Panuveitis, Treatment

INTRODUCTION

Behçet’s syndrome (BS) or Behcet’s disease (BD) is a chronic multisystem disorder of unknown etiology that is characterized mainly by recurrent mucocutaneous, articular, neurological, gastrointestinal, and ophthalmological lesions.

BD was first described, in 1937, by a Turkish dermatologist Dr. Hulusi Behçet, as a triad of symptoms consisting of oral aphthae, genital ulcers and hypopyon related uveitis and erythema nodosum.¹

The underlying pathology includes an inflammatory process of arteries and veins which classified as a systemic vasculitis and it can involve both the arteries and veins of almost any organ system.²

The usual onset of BS occurs in the third decade. Onset is rare in people 50 years or older and before puberty. Both genders are affected equally, but men and the young have a more severe disease course.³

There is no definitive laboratory test to confirm BS; hence, diagnosis is based on clinical features.

A number of diagnostic criteria have been proposed for BS. The 1990 International Study Group (ISG) criteria⁴ mandate the presence of oral ulcers along with two of the following: Recurrent genital ulceration, eye lesions (anterior or posterior uveitis), skin lesions (erythema nodosum, pseudofolliculitis, papulopustular lesions, and acneiform nodules), and positive pathergy test.

ISG criteria of classification have been defined with a sensitivity of 85% and specificity of 96% (Table 1). The ISG Criteria for BS have a higher sensitivity and provide a weighted score to the various manifestations of BS.

According to International Criteria for BD (ICBD) Ocular lesions, oral aphthosis, and genital aphthosis are each assigned 2 points, while skin lesions, central nervous system (CNS) involvement, and vascular manifestations score 1 point each. The pathergy test, when used, scores 1 point. A score of at least 4 points is classified as BS.⁵

The present case was meets more than 4 points for diagnosis of BS.

CASE REPORT

A 20-year-old unmarried, Hindu, male patient was referred to us for evaluation of multiple complaints including
recurrent painful oral ulcers, 4 episodes in the past 1 year, and each time lasting for around 2 months. For the past 6 months, he complained of swelling and pain of right knee and both elbow joints. In addition to oral ulcers and arthritis, recurrent episode of folliculitis over the legs and feet, recurrent genital ulcers on the scrotum and shaft of penis were reported for last 6 months. He has right scrotal swelling since last 1 month. For the past month, he had right eye redness and pain on eye movement also. He also has a history of recurrent acneiform lesions over the face. He also had history of significant weight loss, night sweats, and loss of appetite. There was no history of diarrhea or blood mixed stools, abdominal pain, lymph node enlargement, recent onset hypertension, oliguria, hematuria, edema over feet or periorbital edema, headache, altered sensorium, stroke, and thrombosis in the past. There was no family history of stroke in young or deep venous thrombosis to suggest a hypercoagulable state. He had not received blood transfusions in the past or ever abused intravenous drugs. There was no history of contact with tuberculosis. He had received multiple courses of broad-spectrum antibiotics elsewhere without any relief. On examination, his pulse rate was 97/min in regular rhythm, all the peripheral pulses were palpable, blood pressure was 130/80 mm Hg in the right upper limb, and respiratory rate was 20/min. General physical examination revealed mild pallor, oral ulcers over the lateral surface of tongue and cheek, small, about 0.1 cm × 0.1 cm with clean base (Figure 1). A genital ulcer (Figure 2) measuring 1 cm × 1 cm was present over the penile shaft ventral surface with erythematous border and showing signs of healing with healthy granulation tissue at the base. Right scrotal swelling and tenderness were present. There were the presences of multiple follicular lesions over legs and healed rash (Figure 3). The presence of right knee (Figure 4) and elbow joint swelling and tenderness on examination was suggestive of inflammatory nature of arthritis. At admission, decreased visual acuity in the right eye (20/200) was observed. A slit lamp examination showed diffuse and painful episcleritis with injection in the superficial episcleral vessels; the fundus examination

Table 1: Frequency of the clinical manifestations of BS and classification criteria of the ISG

<table>
<thead>
<tr>
<th>Manifestations</th>
<th>Frequency (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oral ulcers</td>
<td>97-99</td>
</tr>
<tr>
<td>Genital ulcers</td>
<td>=85</td>
</tr>
<tr>
<td>Papulopustular lesions</td>
<td>=85</td>
</tr>
<tr>
<td>Erythema nodosum</td>
<td>=50</td>
</tr>
<tr>
<td>Pathergy reaction</td>
<td>=60 (Mediterranean countries and Japan)</td>
</tr>
<tr>
<td>Uveitis</td>
<td>=50</td>
</tr>
<tr>
<td>Arthritis</td>
<td>=50</td>
</tr>
<tr>
<td>Subcutaneous thrombophlebitis</td>
<td>25</td>
</tr>
<tr>
<td>Deep vein thrombosis</td>
<td>=10</td>
</tr>
<tr>
<td>Arterial occlusion/aneurysm</td>
<td>=4</td>
</tr>
<tr>
<td>CNS involvement</td>
<td>10</td>
</tr>
<tr>
<td>Epididymitis</td>
<td>=10</td>
</tr>
<tr>
<td>Gastrointestinal lesions</td>
<td>1-30 (Most prevalent in Japan)</td>
</tr>
</tbody>
</table>

In the absence of other clinical explanations, patients must have the following: Recurrent oral ulceration (aphthous or herpetiform) observed by the physician or patient at least three times in one 12-month period plus two of the following: Recurrent genital ulceration. Eye lesions: Anterior uveitis, posterior uveitis, cells in the vitreous by slit-lamp examination, or retinal vasculitis observed by an ophthalmologist. Skin lesions: Erythema nodosum, pseudofolliculitis, papulopustular lesions, or acneiform nodules in postadolescent patients not treated with corticosteroids. Positive pathergy test read by a physician at 24-48 h. BS: Behcet’s syndrome, ISG: International Study Group, CNS: Central nervous system.
revealed a right hyperemic disc with blurred margins. Rest of the general physical and systemic examinations was unremarkable.

Hematological investigations revealed a normocytic normochromic anemia (hemoglobin 10 g/dL), total leukocytes count was 70,00/mm³ (neutrophils 72%, lymphocytes 28%), platelet count was 327,000/mm³, normal renal (blood urea 16 mg/dL, serum creatinine 1 mg/dL) and liver function tests (bilirubin 0.5 mg/dL, aspartate aminotransferase 24 U/L, alanine aminotransferase 09 U/L, alkaline phosphatase 155 U/L, serum protein 07 g/dL, and serum albumin 3.71 g/dL). He had markedly elevated acute phase reactants (erythrocyte sedimentation rate by Westergren’s method 80 mm/hr, C-reactive protein 10 mg/dL, normal <0.6 mg/dL). Serology for hepatitis B surface antigen, anti-hepatitis C virus, HIV-enzyme-linked immunoassay and Venereal Disease Research Laboratory test were negative. His tests for autoantibodies such as rheumatoid factor, antinuclear antibody, anticardiolipin, and antineutrophilic antibody were negative. His X-ray chest was normal, ultrasonography abdomen was suggestive of mild hepatosplenomegaly. Three blood cultures did not grow any microorganisms. His electrocardiogram and two-dimensional transthoracic echocardiography were suggestive of normal study. Mantoux (10 TU) and Pathergy tests were negative.

Our patient fulfilled both ISG and ICBD criteria, (i.e., orogenital ulcers, arthritis, skin lesions, and vascular manifestations-hence scoring more than 8 points, fulfilling ICBD criteria for the classification of BS), hence was diagnosed as having BS.

In view of the fact that he had been extensively evaluated before presenting to us and no exact cause of orogenital ulcers, arthritis and folliculitis identified in spite of multiple prior hospital visits and admissions, he was labeled as fever of unknown origin, subsequently identified to be due to BS vasculitis and its manifestations.

For management of his disease manifestations, we have started treatment with oral colchicine - 0.5 mg daily in two divided dose, oral Prednisolone as 1 mg/kg daily tapering dose and local steroid cream for orogenital ulcers, dexamethasone eye drops six times daily in the right eye, tablet Etoricoxib 90 mg single daily dose for arthritis and joints pain, antibiotics coverage for folliculitis with local care of skin and other supportive therapy.

With therapy, all his symptoms improved in a week (Figures 5-7) and he was discharged in stable condition. He was asymptomatic when contacted over the phone at 20 days later and was due to the tapering of steroid dose and maintenance therapy.

DISCUSSION

The present case was a male and it is also supported by the Indian study of Arulrajamurugan et al., they have also concluded in their study that median age at onset of BS was 32 years which is 1 year later compared to other studies,
musculoskeletal manifestations were common in Indians and eye involvement was less.

Oral aphthae were the initial manifestation in almost all the Indian patients in study done by Arulrajamurugan et al.,7 and it same like other studies. It may be the forme fruste of the disease hence all the patients with complex aphthosis should be monitored for the development of features to fulfill the diagnostic criteria as this may help in diagnosing many cases of BS earlier.

Genital ulcers were also observed in all the patients in study by Arulrajamurugan et al.,7 and the Libyan study population but it was reported less in German and Turkish population.8 A meta-analysis showed that the most frequent association is with HLA-B51/B5, which has been validated in various ethnic groups.9

The pathergy reaction represents hyperreactivity of the skin to trauma. Typically, it is characterized by erythematous papule or pustule formation at a skin site 24-48 h after insertion of the needle. This phenomenon is quite specific for BS. However, wound healing after biopsy-induced trauma is normal.

In present case, Pathergy reaction was negative which is often negative in Indian patients as suggested by many studied in India.7,10 Pathergy reaction considered highly sensitive and specific for BS in patients originating from Turkey, the Middle East, Japan and Korea.11

Seen in up to 50% of patients, the typical features of joint involvement include a nondeforming, nonerosive peripheral monoarthritis or oligoarthritis that lasts not more than a few weeks. The knees are involved most frequently, followed by the ankles, wrists and elbows.12

Ocular involvement was in the form of panuveitis and retinal vasculitis and was less in Indian patients as suggested by Arulrajamurugan et al., in their study and which were also similar to the other Indian studies from Delhi.13

The Indian study was done by Arulrajamurugan et al., and was suggestive of that cutaneous manifestations such as erythema nodosum, papulopustular lesions, acneiform lesions, pyoderma gangrenosum and Sweet’s syndrome can occur in BS. In their study, they also demonstrated that patient’s with BS erythema nodosum and vesiculopustular lesions were present equal to the percentage quoted in German and Turkish population and are higher than the Libyan patients.14

**CONCLUSION**

There are only a very few case reports available from India indicating that this disease may be uncommon in our country. However, it is common in Japan and Mediterranean countries, and this variation may be because of environmental and genetic difference. The diagnosis should be clinical and ISG or ICBD classification criteria are also helpful for the same. Treatment is symptomatic using steroids and immunomodulatory therapy. It is efficient depending on the rapidity of initiation, the compliance, and the duration of therapy. The systemic prognosis is good in patients with BD in the absence of CNS involvement or involvement of the major vessels.

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