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CASE REPORT

Multifocal Venous Thrombosis in Behcet's Disease

Lena Jafri¹, Nosheen Nasir² and Aysha Almas²

ABSTRACT
Behcet's disease is a multisystem inflammatory vascular disorder with a chronic course characterized by recurrent oral and genital ulcers, eye lesions, arthritis and skin lesions. It has a typically waxing and waning course. The cause and pathogenesis of the disease are unclear and specific treatment is not available. A 39 years old man presented with rash, ocular manifestation and left leg swelling. He was found to have deep venous thrombosis of left leg along with recurrent cerebral venous thrombosis. He was a known case of Behcet's disease since 3 years and had been on anticoagulants since then.

Key words: Behcet's disease. Venous thrombosis. Ulcer.

INTRODUCTION
Behcet's disease is a multi-system inflammatory disorder with a chronic relapsing course. It is prevalent along the ancient Silk Route.¹ There are no pathognomonic laboratory tests and the diagnosis requires recurrent oral ulceration accompanied by two of the following: genital ulceration, ocular disease, skin lesions or a positive skin pathergy reaction.²³ Despite the inclusive criteria set forth by the International Study Group, there are cases where the criteria cannot be met and a diagnosis is missed. It usually affects young adults 20 – 40 years of age.⁴⁵ We report, to the best of our knowledge, the first case from Pakistan regarding an aggressive case of Behcet's disease in a 39 years old man, with ocular manifestation and venous thrombosis at multiple sites despite being on anticoagulants.

CASE REPORT
A 39 years old man presented to the clinic with rash on the body for 3 days. He complained of low grade intermittent fever and left leg swelling since 4 – 5 days. He was unable to swallow due to painful mouth ulcers and hence was admitted. He was diagnosed to have Behcet's disease 3 years ago and had been on warfarin. In previous magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) repeated few months after cerebral venous thrombosis superior sagittal sinus was not clearly visualized in its posterior part, with significantly increased vascularity noted in the near vicinity of superior sagittal sinus. Features were suggestive of partial re-canalization of superior sagittal sinus following thrombosis (Figure 1). Magnetic resonance arteriography (MRA) images were unremarkable. He developed recurrent mouth ulcers progressively increasing in frequency with > 3 episodes in a year. He was also treated with steroids during this time. He gave a history of cigarette smoking 4 – 8/day. Patient denied photosensitivity, alopecia, weight loss and alcohol intake.

On examination, he was a lethargic man of good built, afebrile with a blood pressure of 135/89 mmHg, pulse rate 101 beats/minute and respiratory rate 19 breaths/minute. There were multiple papulo-pustular 1 – 2 cm lesions resembling acne on face, back, chest, groin and abdomen.
**Table I: Laboratory investigations after admission.**

<table>
<thead>
<tr>
<th>Laboratory parameter</th>
<th>Result</th>
<th>Laboratory parameter</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin (gm/dl)</td>
<td>14.9</td>
<td>Calcium (mg/dl)</td>
<td>9</td>
</tr>
<tr>
<td>Mean cell volume (FL)</td>
<td>82.7</td>
<td>PT (seconds)</td>
<td>13.8</td>
</tr>
<tr>
<td>White blood cell count</td>
<td>16.4</td>
<td>PT control (seconds)</td>
<td>11</td>
</tr>
<tr>
<td>Neutrophils (%)</td>
<td>80.7%</td>
<td>INR</td>
<td>1.31</td>
</tr>
<tr>
<td>Lymphocytes (%)</td>
<td>6.9%</td>
<td>APTT (seconds)</td>
<td>26%</td>
</tr>
<tr>
<td>Eosinophils (%)</td>
<td>0.1%</td>
<td>APTT control (seconds)</td>
<td>30%</td>
</tr>
<tr>
<td>Monocytes (%)</td>
<td>12.2%</td>
<td>Total Bilirubin (mg/dl)</td>
<td>0.8%</td>
</tr>
<tr>
<td>Basophils (%)</td>
<td>0.1%</td>
<td>Serum glutamic</td>
<td>66%</td>
</tr>
<tr>
<td>Platelets count (x10E9/L)</td>
<td>307</td>
<td>pyruvic transaminase (IU/L)</td>
<td>969</td>
</tr>
<tr>
<td>Blood Urea Nitrogen (mg/dl)</td>
<td>18</td>
<td>C-reactive protein (mg/dl)</td>
<td>10.6</td>
</tr>
<tr>
<td>Creatinine (mg/dl)</td>
<td>1.0</td>
<td>Erythrocyte sedimentation rate (mm/hr)</td>
<td>49</td>
</tr>
<tr>
<td>Sodium (mmol/L)</td>
<td>135</td>
<td>Lactate dehydrogenase (IU/L)</td>
<td>969</td>
</tr>
<tr>
<td>Potassium (mmol/L)</td>
<td>4.7</td>
<td>Malarial parasite</td>
<td>Not seen on peripheral film</td>
</tr>
<tr>
<td>Chloride (mmol/L)</td>
<td>104</td>
<td>ICT malaria</td>
<td>Negative</td>
</tr>
<tr>
<td>Bicarbonate (mmol/L)</td>
<td>19.3</td>
<td>Blood culture</td>
<td>No growth</td>
</tr>
<tr>
<td>Phosphate (mg/dl)</td>
<td>3.9</td>
<td>Urine detail report</td>
<td>Trace proteins and ketones</td>
</tr>
</tbody>
</table>

ANA, ASMA and AMA were negative.

**Abbreviations:**
- PT = Prothrombin time; INR = International normalized ratio; APTT = Activated partial thromboplastin time; ANA = Antinuclear antibody; ASMA = Anti smooth muscle antibody; AMA = Anti mitochondrial antibody.

medial aspect of thighs and few were on the scrotum.

**DISCUSSION**

We report this case of Behcet's disease with extensive deep venous thrombosis along with thrombosis of the cortical venous sinuses, a rare manifestation of this disease from our part of the world. Arterial and venous involvement is one of the characteristics of Behcet's disease. Vascular involvement was seen in 8.3% cases in Iran while 16.8% and 18% cases have been reported from Turkey and Saudi Arabia respectively.

We report a fourteen-fold increase risk of venous thrombosis in Behcet's patients. Venous involvement is common in men especially those with ocular involvement and a positive pathergy test.

It is observed that men are more likely affected by Behcet's disease. Age and gender distribution in India shows a male to female ratio of 1.8 and the age of onset is 33 years. These findings are corroborated in the presently reported case and conforms to the data available from the rest of the world. However the clinical presentation reported from India shows involvement of oral and aphthous ulceration followed by joint and skin involvement as opposed to clinical symptoms reported from China and Iran where there was no significant joint involvement.

To conclude, ocular and cerebrovascular thrombosis are important complications of Behcet's disease. These can occur despite preemptive anticoagulation as provided in the present case. Hence, there is a need for regular follow-up of such cases.

**REFERENCES**


