Behcets Syndrome with a Rare Manifestation

Nirmal Taparia¹, Vitthal Dhadke ²

Abstract

We report a case of a 25 year old patient with history of recurrent oral aphthous ulcers and genital ulcers and phlyctenular conjunctivitis presenting to us with sudden onset of cough and difficulty in breathing diagnosed as Behcets disease with pulmonary embolism. A rapid and precise diagnosis and treatment of this patient led to good recovery of this patient. Pulmonary thromboembolism is a rare complication of Behcets disease.

Introduction

Behcets disease is a chronic, relapsing inflammatory disease characterized by recurrent oral and genital ulcers as well as lesions affecting the eyes, skin, nervous system, joints and vessels. It is classified among vasculitides. The diagnostic criteria are clinical and based on the Internationally agreed diagnostic criteria described in Table 1. It is a rare immune mediated small vessel systemic vasculitis. It is named after the Turkish dermatologist Hulusi Behcet who described the triple symptom complex in 1937. The specific etiology of Behcets disease remains elusive and it could be auto inflammatory or auto immune in origin. Behcets disease is a sporadic disease but carriers of HLA B51/HLA B5 have an increased risk of developing this disease than non carriers. Recent research has shown that multiple genes in the interleukin 23 signalling pathway contribute to this disorder. Polymorphism of JAK2 and STAT3 are associated with these disorders. Males and females are equally affected but males often have a more serious disease. Sex prevalence varies by country. Turkey has the highest prevalence of Behcets disease with 420 cases/1,00,000 population.

Here, we present a case of Behcets in a young male patient presenting in OPD with recurrent oral and genital ulcers and an episode of severe dyspnea and hemoptysis.

Case Report

A 25 years old male patient came to OPD with c/o oral ulcers and severe breathlessness and cough since 2 days. It was of sudden onset with no fever, chest pain, or trauma. He had no such past history. Patient gave a prior h/o asymptomatic hemoptysis.

Based on his clinical features of recurrent oral and genital ulcers and conjunctivitis, Pathergy test (Figures 6 and 7) and imaging studies a diagnosis of pulmonary embolism with Behcets disease was made. Pulmonary function test and ulcer biopsy were not done.

Patient was treated with IV fluids, Oxygen by nasal prongs to maintain SpO2 > 90%. IV steroids Methylprednisolone at a dose of 250mg twice daily for 5 days, Inj Low molecular weight heparin was given subcutaneously at a dose of 2500u twice a day for 5 days along with oral anti platelet agent aspirin 75 mg daily. Prophylactic antibiotic Inj. Cefotaxime 1 gm was given iv twice daily for 5 days.

Table 1: Diagnostic Criteria for Behcets syndrome

<table>
<thead>
<tr>
<th>Recurrent oral ulceration plus two of the following</th>
<th>Recurrent genital ulceration</th>
<th>Eye lesion</th>
<th>Skin lesion</th>
<th>Pathergy test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients value</td>
<td>Normal range</td>
<td>Patients value</td>
<td>Normal range</td>
<td>Patients value</td>
</tr>
<tr>
<td>Hemoglobin (g/dl)</td>
<td>11.0</td>
<td>12.0-15.0</td>
<td>WBC (x10^-3/ul)</td>
<td>9.5</td>
</tr>
<tr>
<td>Platelets (x10^-3/ul)</td>
<td>30</td>
<td>150-400</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Biochemistry</td>
<td>blood urea (mg/dl)</td>
<td>28</td>
<td>7-40</td>
<td>Sr. creatinine (mg/dl)</td>
</tr>
<tr>
<td>Blood Coagulation profile</td>
<td>Bleeding time</td>
<td>4min 55sec</td>
<td>&lt;7.1 min</td>
<td></td>
</tr>
<tr>
<td>Clotting time</td>
<td>5min 18sec</td>
<td>&lt;7.1 min</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Prothrombin time</td>
<td>14 sec</td>
<td>12.4-15.7sec</td>
<td></td>
<td></td>
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<tr>
<td>INR</td>
<td>1.15</td>
<td>0.8-1.2</td>
<td></td>
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</tbody>
</table>

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Discussion

Behcets disease is a benign condition with mucocutaneous and ocular manifestations of disease. There are 4 major case series in the world, Turkey with 2147 patients, Korea with 1155 patients, Morocco with 1034 patients, and UK with 419 patients.10

Recurrent aphthous ulcers are a sine qua non for the diagnosis. The ulcers are usually painful, shallow or deep with a central yellowish necrotic base and are located anywhere in the oral cavity with 1 to 10 mm in diameter. They subside without leaving scars and are seen in 85% of patients.2 The localization in the order of frequency is lips, cheeks, tongue, gingival, palate, tonsils and pharynx.10

Non specific skin reaction to intradermal saline injections (pathergy test) or scratches is observed in 50% of patients.2

Genital ulcers are less common, in 73%, more specific, painful, do not affect the glans penis or urethra and produce scrotal scars. As shown in the figure in this case.

Eye involvement with bilateral panuveitis is the most dreaded complication. Eye disease occurring in 50% of patients may develop in few years as it did in our patient. Iritis, posterior uveitis, optic neuritis and retinal vein occlusion all can be seen.2

Non deforming arthritis or arthralgias are seen in 50% of patients and affect the knees and ankles.2 Neurological involvement seen in (7.3%) of cases classically manifests as meningoencephalitis, and GI cases (9%) presenting as ulceration of the ileo-cecal region, also occur.10

Pulmonary artery aneurysms (PAA) are the most common pulmonary lesions in Behcets disease and these are almost always associated with hemoptysis. Upto 78% patients with aneurysms have concomitant extrapulmonary venous thrombi or thrombophlebitis. Underlying pathology is pulmonary vasculitis which may result in thrombosis, infarction, hemorrhage or PAA formation.6,7 Immunosuppression is the mainstay therapy for treatment.8

Pulmonary disease can be divided into subtypes depending upon involvement viz9

1. PAA

2. Pulmonary parenchymal changes.


4. Radiologic abnormalities

5. Infection, immunosuppressive related presentation

6. Abnormal pulmonary function finding or COAD

Pulmonary emboli are a rare complication seen in 0.5% of cases of Behcets disease.9 Pulmonary artery vasculitis presenting with dyspnea, cough, chest pain, hemoptysis and infiltrates on chest x ray have been reported in 5% of patients.5

Our patient showed a thrombus in subsegmental branch of right pulmonary artery. The differential diagnosis could be acute coronary syndromes, acute pericarditis or pleuritis, pneumonia, hypersensitivity pneumonitis,
pneumothorax and anxiety neurosis to name a few. Most of them were ruled out clinically and by history. Patient was afebrile on presentation, WBC were normal, ECG, 2D Echo were normal. X-ray of chest showed left lower zone wedge shaped opacity and CT Thorax with Angiogram showed thrombus in left subsegmental pulmonary artery supplying lateral basal segment resulting in complete obliteration of the lumen with consolidation and mild pleural effusion.

Other forms of arteritis could present this way, but the classic triad of recurrent oral and genital ulcers with uveitis made a diagnosis of Behcets disease.

Our patient was treated with, aspirin 75 mg/day, low molecular weight heparin, local oral steroids and short course of systemic steroids. Patient responded well and was discharged home.

Other treatments available are Thalidomide 100 mg/day for severe mucous membrane involvement, Colchicine 1-2 mg/day orally for arthritis and severe ulcers, Azathioprine and Cyclophosphamide for sight threatening uveitis and CNS Behcets syndrome.25

A new drug Apremilast, a selective inhibitor of the enzyme phosphodiesterase 4, for recurrent painful oral ulcers was studied, with some beneficial effects.7

Use of interferon alpha (INF a2a and INF a2b) at a dose of 6-9 million units/day can be used for ocular lesions. Similarly drugs like Etanercept at doses of 25 mg subcutaneously twice a week for 4-6 weeks and Infliximab 10 mg/kg for ocular attacks have been reported in small studies with varied results.10

Conclusion

Behcets disease is a benign chronic relapsing disease with predominant mucocutaneous features. But in rare instance it may present with a life threatening complication like pulmonary embolism and/or panuveitis which should be recognized and treated promptly, which can result in significant morbidity, if delayed. Coronary / pulmonary arterial aneurysm rupture carries a high mortality rate.6,9

Treatment of Behcets disease is symptomatic. It is aimed at reducing symptoms and preventing complications. Prognosis varies based on the organ system involved.9

References