ADAMANDIADIES-BEHÇET DISEASE: A CASE REPORT

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ABSTRACT
Adamandiades-Behçet disease is a rare clinical condition commonly found in males. It is a multisystem disorder of middle age characterized by oral, genital ulceration with opthalmic, gastrointestinal, neurological and joint involvement. We report here a case of middle age woman who had presented with multiple recurrent oral and genital ulceration over period of two years. Histopathological reports of genital biopsy confirmed Behçet’s disease. Ulcer reoccurrences are very common.

Keywords: Adamandiades-Behçet Disease, Aphthous Ulcers, Vasculitis

INTRODUCTUON
Adamandiades-Behçet disease was first reported in year 1931 and 1937 by a Greek ophthalmologist Benediktos Adamandiades and a Turkish dermatologist Hulus Behçet, respectively (Zoubulis, 2002). Common age of presentation being 20-30 year, this is a multisystem inflammatory disease of unknown etiology, classified as systemic vasculitis involving all types and size of blood vessels and characterized clinically by recurrent oral aphthous and genital ulcers, skin lesions, and iridocyclitis/posterior uveitis, occasionally accompanied by arthritis, vasculitis, gastrointestinal, neurologic, or other manifestations (McCarty, 2003; Suzuki, 2000).

CASES
A 45 year old female presented with the clinical history of multiple, recurrent, painful ulcers over the oral and genital mucosa for the past two years. Patient took several topical and systemic medications, for the same, with minimal symptomatic relief. Recurrence of ulcers every three monthly was reported. No history of same complaint was noted in the family members.

Examination of oral mucosa revealed multiple, sharply margined, ulcers ranging from 2 mm to 4 mm in diameter with surrounding erythema (Figure 1).

Genital examination showed solitary, erythematous, tender ulcer, 2X3 cm, covered with slough, extending from right labia majora, minora upto right perianal area (Figure 2).

Figure 1: Multiple oral aphthous ulcers

Figure 2: Genital ulcer
Investigations
Culture and sensitivity of the swab taken from the genital ulcer showed methicillin sensitive staphylococcus aureus. Pathergy test was negative. Systemic examination revealed no abnormal findings. Routine laboratory investigations were within normal limits. Histopathology of the genital lesion revealed the skin with mild acanthosis of the epidermis. The dermis showed moderate perivascular mononuclear cell infiltrate, with variable fibrin deposition. Few capillaries showed neutrophilic vascular infiltration suggestive of leukocytoclastic vasculitis (Figure 3). Therefore confirmed our clinical impression of Behcet’s disease.

Figure 3: Photomicrograph of skin biopsy. (a) Showing mild acanthosis and mononuclear cell vasculitis in the dermis. (H & E, x100). (b) Dense perivascular mononuclear cell infiltration, along with neutrophils infiltrating in the vessel wall (arrow) (H & E, x400).

Treatment
Patient was treated with oral colchicine 0.5mg once daily (OD) for one month, prednisolone 20mg for first two weeks then tapered to 10mg for next two weeks. Dapsone 100mg twice daily (BD) for one month and ciprofloxacin 400mg BD for seven days was also administered. Patient was advised topical triamcinolone acetonide 1% gel for the oral lesions and washing of genital lesion with normal saline. Both oral and genital lesions healed completely in one month. Maintenance dose of colchicine 0.5mg OD and prednisolone 10mg was given. No new lesion has appeared in three months and patient is currently under follow up.

RESULTS AND DISCUSSION
Behcet’s disease is a systemic autoimmune vasculopathy manifesting usually by recurrent oro-genital aphthous-like ulcerations and eye findings (Gul, 2000). There are no specific manifestations or special diagnostic tests for the disease. Aphthous oral ulcers are usually the first and most characteristic clinical feature. Genital ulcers resemble oral ulcers and may be single or multiple. Besides skin, vasculitic lesions can be seen in eyes, central nervous system, gastrointestinal system, bones, kidneys and large blood vessels.

The prevalence for a population of 100,000 is 13.4 in Japan, 14 in China, 16.7 in Iran, 20 in Saudi Arabia, 80 to 370 in Turkey and much less in other countries. There are only a few reports of Behcet's disease from India (Pande, 1995). In Indian population disease is milder in presentation, mostly affecting joints and mucocutaneous sites (Singal, 2013). Most cases of Behcet’s disease are sporadic and the parents of patients are usually unaffected (Gul, 2000). The unique geographic distribution might suggest an environmental link and an infectious cause has been suggested, including herpes simplex virus type1 and some Streptococcus species (Reynolds, 2008).

Significant association exists between the disease and human leukocyte antigen-B51, in Japan, the Middle East and the Mediterranean countries; however, this relationship is not as strong in Western countries (De Menthon, 2009).
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Treatment has become much more effective in recent years; BD is still associated with severe morbidity and considerable mortality. The main aim of the treatment should be the prevention of irreversible organ damage. Therefore, close monitoring, early and appropriate treatment is mandatory to reduce morbidity and mortality (Alpsoy, 2009). Colchicine may be useful for treating some of the manifestations of Behçet's syndrome, especially among women (Yurdakul, 2001). Dapsone also inhibits the enhanced chemotactic activity of neutrophils and can be used as an alternative compound to colchicine (Sharquie, 2011). Corticosteroids have been widely used almost for all lesions of the disease but most effective in controlling erythema nodosum lesions. It can also be given as monotherapy or in combination with other drugs such as colchicine, interferon (IFN)-α, cyclosporine, or azathioprine (Alpsoy, 2012).

Table 1: Revised International Criteria for Behçet Disease (International Team for the Revision of ICBD; coordinator: F.Davatchi)\(^a\)

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ocular lesions (recurrent)</td>
<td>2</td>
</tr>
<tr>
<td>Oral aphthosis (recurrent)</td>
<td>2</td>
</tr>
<tr>
<td>Genital aphthosis (recurrent)</td>
<td>2</td>
</tr>
<tr>
<td>Skin lesions (recurrent)</td>
<td>1</td>
</tr>
<tr>
<td>Central nervous system</td>
<td>1</td>
</tr>
<tr>
<td>Vascular manifestations</td>
<td>1</td>
</tr>
<tr>
<td>Positive pathergy test(^b)</td>
<td>1</td>
</tr>
</tbody>
</table>

\(^a\): BCD scoring: score ≥ 4 indicates Adamantiades-Behçet disease. \(^b\): Though the main scoring system does not include pathergy test, where pathergy testing is conducted, a positive result may be included for one extra point.

There are many criteria followed in the past for the diagnosis of Behcet’s disease most common being Behçet Disease Research Committee of Japan and International Criteria for Classification of Behçet Disease. But, there were many problems with these criteria so they were revised. Our case fulfills the essential criteria for the diagnosis of Behçet’s Disease, mentioned in Revised International Criteria for Behçet Disease, 2010 (Table 1) (Zouboulis, 2012).

We are reporting this case as; Behçet’s disease is rarely encountered. Proper evaluation of recurrent mucosal ulcerative disorder is required to arrive at diagnosis. Hence the clinicians should keep this possibility in mind when confronted with such clinical picture.

REFERENCES


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