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CASE REPORT

Behcet's Syndrome: A Case Report Review

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Abstract

This is the case report of a man who suffered from various problems; were eventually identified as features of Behcet's syndrome, which is uncommon in the clinical settings. Besides the known clinical symptoms of the disease, this patient has suffered intermittently from dysmenorrhoea and abdominal pain, which may well be features of this disorder which have not previously been documented. Early diagnosis and prompt treatment helped the multidisciplinary team to get positive response from the patient.

Keywords: Behcet's Syndrome; Chronic Morbidity; Thrombophlebitis; Colitis; Arthritis.

Introduction

Behcet's Disease is a chronic, relapsing, occlusive vasculitis affecting multiple organ systems in the body, characterized by oral and genital ulcerations, uveitis, skin rashes, arthritis, thrombophlebitis, colitis, and neurologic symptoms. Histologically, there is a combination of a perivascular neutrophilic or lymphocytic infiltration, endothelial cell damage or swelling, fibrinoid necrosis coupled with a pro-thrombotic tendency.

Prevalence and expression vary geographically, much higher in latitudes between 30° and 45° North, Around Mediterranean basin extending through Middle East and Orient Striking similarity of distribution to ancient Silk Road. Suggests that an inherited tendency to develop Behcet's Disease was spread by merchants who traveled these trading routes. Turkey has the highest prevalence of the disease (80 to 370 cases per 100,000 populations). Japan, Korea, China, Iran, and Saudi Arabia ranges from 13.5 to 20 cases per 100,000, much lower in Western countries. Prevalence of the disease in United States is 0.33 per 100,000.

Chronic morbidity is typical and leading cause is ophthalmic involvement, which can result in blindness. Effects of disease may be cumulative, especially with neurologic, vascular, and ocular involvement. Mortality rate is low, but death can occur from neurologic involvement, vascular disease, bowel perforation, cardiopulmonary disease, or as a complication of immunosuppressive therapy.

Case History

Mr. D. Singh, 32 year male with thin body built, belongs to a middle class family who works in a cultivating forms and constructions works on daily wages, admitted in male medicine wardwith the diagnosis of Behcet's syndrome. Chief complaints of the patient were fever from last 4 to 5 days, loose motions from last 4 to 5 days, Cough, cold and ulcers in mouth last 4 days, abdominal pain is present from

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last two days. The abdominal pain was sudden in onset, dull, continuous in nature, and non-radiating and pain rating was 5/10 on VAS. In addition to these complaints, he had complaints of breathlessness from last one week along with increase in respiratory rate while walking and become normal during sitting and relaxing. Cough was also present. Mr. D. Singh was unable to sleep adequately due to pain in abdomen, in mouth due to ulcer and fever from last 4 days. He had disturbed sleep and loss of appetite from last week

Mr. D Singh was apparently normal before 5 days of admission, and then he developed fever which was insidious in onset and is continuous in nature. On the same day, loose motion also occurred, 10 to 12 episode in a day, brown yellow in colour and watery in consistency. Next day morning he observed ulceration in mouth and started Cough with expectoration.Abdominal pain is also present which is sudden in onset and non-radiating in nature and 5/10 on VAS. He took symptomatic treatment for 3 days on the advice of local doctor. Symptoms did not relieved; then he came to hospital for further treatment.

In past, patient had a history of Behcet's syndrome before 4 years. The signs and symptoms were severe abdominal pain with diarrhoea. He had been treated for a week; once symptom relieved he was discharged. He could not follow any further check-ups and precautions thereafter. Furthermore he has history of Tuberculosis which was diagnosed since 3 years and is taking anti-tubercular drugs (Category 1). There is no evidence of Diabetes mellitus, hyper tension and other chronic diseases. In his family there is no history of any disease or similar signs and symptoms.

Onexamination patient was conscious and oriented to self, time, place and person. He was quiet and cooperative, well dressed and groomed. His height was 172cm and weights 58kg. Vital signs ware altered and was indicated signs of inflammation. Eyes, oral cavity, lips, respiratory system, digestive system, and musculoskeletal system were affected mainly. Blurring of vision, pain in the abdomen, diarrhea, and ulcerative erosions in the stomach, stomatitis, haemoptysis, angular stomatitis and pain in joints were the major health problems.

Behct's Syndrome

Behcet's syndrome is one of the largest type autoimmune diseases, sometimes called Behcet's syndrome, MorbusBehcet's, Adamantiades syndrome or Silk Road disease and it is named after Turkish dermatologist HulusiBehcet. He described triple symptom complex of recurrent oral ulcer, genital and uveitis. As a systemic disease, it can also involve visceral organs such as gastrointestinal tract. Pulmonary, musculo-skeletal, cardiovascular and neurological system.

Definition: A chronic disease featuring inflammation of small blood vessels and characterized by a triad features ulcers in mouth, ulcer of the genitalia and inflammation of eye (uveitis)

Risk Factor

According to the Literature	According to the Patient
Age-commonly affects young males between 20-35 years **Sex-**it is often more seen in males as compared to females **Genetic:** having certain genes is associated with a higher risk of developing behcet's disease.	Patient's age is 32 year Male No such history of behcet's disease found in patient's family

Etiology

According to the Literature	According to the Patient
Cause is unknown Genetic cause a long with environmental factors	No history of Behcet'sdiasease in patient's family

The cause of Behcet's syndrome is unknown. However, combination of genetic and environmental factors likely plays a role. Several genes have been found to be associated with the disease.A broad intracellular signaling abnormality of a transcription factor, which lowers the threshold of inflammatory responses to external stimuli, as proposed for 'fever' with decreased pyrine expression of neutrophils may be present.

Pathophysiology

A large number of serological studies show a linkage between the disease and human leucocytes antigen HLA-B51. However B51 tends not to be found in disease when a certain SUMO4 gene variant is involved and symptoms appear to be milder when HLA-B27 is present. At the current time, a similar infection origin has not yet been confirmed that leads to Behcet's syndrome, but certain strains of

streptococcus sanguinis has been found to have a homologous antigenicity.

Finally both innate and adaptive immune systems are activated in Behcet's syndrome, with a pro-inflammatory and Th1 cytokine profile. Behcet's syndrome may be linked to a specific, primary immune abnormality with a genetic mutation.

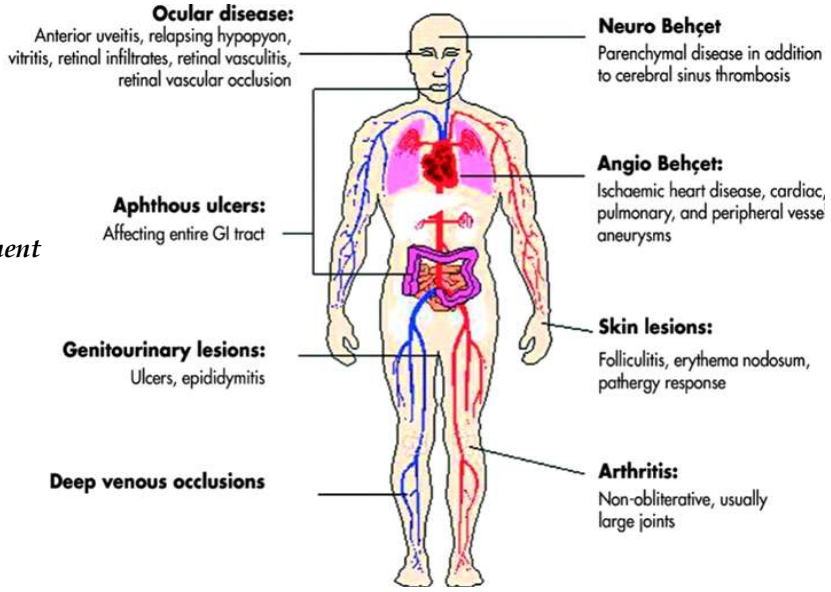
Clinical Manifestations

- Signs and symptoms may be recurrent, may precede onset of mucosal membrane ulcerations by 6 months to 5 years.
- Prior to onset of disease, patients may experience a variety of symptoms.
  - Malaise
  - Anorexia
  - Weight loss
  - Generalized weakness
  - Headache
  - Perspiration
  - Decreased or elevated temperature
  - Lymphadenopathy
  - Pain of the substernal and temporal regions
- A history of repeated sore throats, tonsillitis, myalgias, and migratory erythralgias without overt arthritis is common.

According to the Literature According to the Patient

According to the literature	According to the Patient
Mouth ulcers: About 98% of people with Behçet's syndrome have frequent mouth ulcers. They can affect the mouth, tongue and throat, and are often painful. Sometimes there are many tiny ulcers clustered together.	Patient had numerous ulcers in the mouth and on tongue which were bleeding with minute manipulation.
Skin: Some people may develop acne like sores on their bodies. Others may develop red, raised and tender nodules on their skin, especially on the lower legs.	He did not get any skin lesions, but there was complaint of itching here and there on the body.
Genital ulcers: Women and girls may get ulcers on the vulva, in the vagina or on the cervix. Men and boys may get ulcers on the scrotum and the penis and may have pain or swelling in the testicles. Ulcers and boils may appear around the anus and in the groin.	**Nothing significant**
Eye inflammation: Inflammation within the eye is one of the most serious symptoms of Behçet's syndrome. The inflammation may be at the front or back of the eye, around the iris or next to the retina. Symptoms can include: • Floaters • Haziness or loss of vision • Pain • Redness in the eye.	Blurring of vision present, redness and irritation in the eye were present.
Fatigue: Extreme tiredness is a very common symptom.	Patient feels tired to complete his activities of daily living.
Joint problems: Persons may have aches, pains and swelling in various joints.	Pain in the numerous joints was present.
Digestive system: commonly signs and symptoms include abdominal pain, diarrhea and bleeding.	pain in the abdomen, diarrhea, and ulcerative erosions in the stomach, stomatitis, haemoptysis, angular stomatitis
Brain and nervous system: Behcet's disease may cause inflammation in the brain and nervous system that leads to headache, fever, disorientation, poor balance or stroke. Some people with Behçet's also experience depression.	**Nothing significant**
Vascular system: Inflammation in blood vessels may occur in Behcet's disease, causing redness, pain, and swelling in the arms or legs when a blood clots results. Inflammation in the large arteries can lead to complications, such as aneurysms and narrowing or blockage of the vessel.	Pain in different parts of the body was present.

Spectrum of organ involvement in Behçet's disease



Laboratory Investigations

Investigations Name	Patient's Finding	Normal Finding	Remark
Creatinine	0.34mg/dl	0.90-1.30mg/dl	decreased
Liver function test			
Total bilirubin	0.31mg/dl	0.30-1.30	Normal
Indirect bilirubin	0.15mg/dl	0.20-0.90	Decreased
Total protein	6.1gm/dl	6.4-8.3gm/dl	Decreased
Albumin	2.3gm/dl	3.5-5.0gm/dl	Hypoalbumenia
globulin	3.8gm/dl	1.8-3.6gm/dl	increased
Hematological			
Total red cell count	5.41million/ul	4.50-5.50	Normal
Hemoglobin	12.9gm/dl	13.0-17	Decreased
Mean cell volume	74fl	83.0-101.0	Decreased
Mean cell hemoglobin	23.9pg	27.0-32.0	Decreased
Total leukocyte count	10.5 10x 3ul	4.0-10.0	Slightly decreased
lympocytes	16%	20-45%	Decreased
Renal Function Test			
Urea	15.0mg/dl	15.0-39mg/dl	Normal
creatinine	0.70mg/dl	0.90-1.30	Decreased
Sodium	131.7mm/l	136-146	Hyponatremia
chloride	94mmol/l	102-109	Decreased

Diagnostic Test

According to the literature	
Complete blood count • Erythrocyte sedimentation rate (ESR) • C-reactive protein (CRP) Liver function test Urine analysis Barium studies MRI/CT Scan Chest x ray Endoscopy Genetic marker (HLA-B51) **Pathergy test:** This measures the increased sensitivity of the skin that occurs in Behçet's syndrome. A small pin-prick or injection; if a characteristic red spot appears on the skin around the pin-prick, and then the result is positive.	**No test can determine definitively whether or not about Behcet's syndrome. Criteria have been established for the diagnosis of the disease.**

Diagnostic criteria: (Behcet's syndrome research committee of Japan)

Diagnosing Behcet's can take some time. There's no test to confirm the diagnosis, and the symptoms can be confused with those of other, more common illnesses.

Major features	Minor features
➢ Recurrent aphthous ulceration of oral mucous membrane ➢ Skin lesions -Erythema nodosum – like lesions, subcutaneous thrombophlebitis, folliculitis (acnelike lesions), cutaneous hypersensitivity ➢ Eye lesions - Iridocyclitis, chorioretinitis, retinouveitis, definite history of chorioretinitis or retinouveitis Genital ulcers	➢ Arthritis without deformity and ankylosis ➢ Gastrointestinal lesions characterized by ileocecal ulcers ➢ Epididymitis ➢ Vascular lesions ➢ Central nervous system symptoms

Diagnosis

1. **Complete:** Four major features
2. **Incomplete:**
   a. 3 major features,
   b. 2 major and 2 minor features,
   c. Typical ocular symptom and 1 major or 2 minor features
3. **Possible:**
   a. 2 major features
   b. 1 major and 2 minor features

Revised International Criteria for Behcet's syndrome

Symptoms	Points
Ocular lesions(recurrent)	02
Oral ulcer(recurrent)	02
Genital ulcers(recurrent)	02
Skin lesions	01
CNS symptoms	01
Vascular manifestation	01
Pathergy test positive	01
Total	10

ICBD scoring; score >4 indicates Behcet's disease

Management

Currently no cure for Behçet's syndrome, evidence shows that there is an improved prognosis with early diagnosis and prompt treatment.

Bahcet's Disease needs multidisciplinary approach

- Dermatologist: For evaluation of mucocutaneous lesions (ie, oral ulcer, genital ulcer, skin lesions)
- Ophthalmologist: For evaluation of eye involvement
- Rheumatologist or orthopedic surgeon: For evaluation of joint involvement
- Neurologist or psychiatrist: For evaluation of CNS involvement
- Internal medicine specialist: For evaluation of gastrointestinal, pulmonary, renal, or endocrine involvement
- General surgeon: For evaluation of gastrointestinal involvement
- Chest surgeon or cardiologist: For evaluation of cardiovascular involvement
- Ear, nose, and throat specialist or dentist: For evaluation of oral cavity

Medication received by patient		
Medication	**Dose**	**Frequency**
Tab. Levocetrizine	1gm	BD
Tab. Paracetamol	800mg	SOS
Inj. Normal Saline	500mg	OD
Inj. Ringer Lactate	500mg	OD
Syp. Cough expectorant		TDS
Dualin Nebulisation	20mg	TDS
Azathioprine, dapson ointment for topical application for lesions		

Medication recommended according to the literature		
Drug	**Doses**	**Indication**
Methylprednisolone	40 mg/every 3 week lM	Erythema nodosum (but not orogenital ulcers)
Rebamipide	300 mg/day PO	Oral ulcers
Colchicine	1-2 mg/day PO	Erythema nodosum, arthritis, genital Ulcer s (oral ulcer in female)
Dapsone	100 mg/day PO	Orogenital ulcers, skin lesions, pathergy
Azathioprine	2.5 mg/kg/day	Recent onset ocular disease
lnterferon-alfa-2a	6x106 lU/3x/wk SC	Orogenital ulcers, papulopustular lesions
Thalidomide	100 mg/day	Orogenital ulcers, Papulopustular lesions
CyclosporinA	10 mg/kg/day PO	Ocular manifestations, skin lesions, orogenital ulcers
Etanercept	25 mg/2 x/wk P0	Oral ulcers, papulopustular lesions nodular lesions, (not pathergy test)

Patient's Education

Exercise

It's important to exercise joints and to keep up general level of fitness. Exercise such as yoga or Pilates may also help to reduce stress, which can sometimes trigger a flare-up of symptoms in some people.

Diet and Nutrition

Recommended a healthy, nutritious and balanced diet, with plenty of fruit and vegetables and water, and not too many fats and sugars to keep up general health.

Sex and Pregnancy

The genital ulcers associated with Behçet's can sometimes make sex uncomfortable or even painful. However, they're not sexually transmitted or contagious.

Some of the drugs used to treat Behçet's syndrome can affect sperm, eggs, fertility or even the baby, for example, thalidomide is known to be harmful to an unborn child.

Complications

Complications of Behcet's disease depend on signs and symptoms and the treatment. For instance, untreated uveitis can lead to decreased vision or even blindness. People with eye signs and symptoms of Behcet's disease need to visit an ophthalmologist regularly because treatment can help prevent this complication.Blindness, Paralysis, Embolism/ thrombosis, Aneurysm, Amyloidosis, Thrombic events (especially with positive anticardiolipin antibodies) and even Death may occur in untreated cases.

Conclusion

Behcet's syndromeis a chronic disease featuring inflammation of small blood vessels and characterized by a traid features; ulcers in mouth, ulcers of genitalia and inflammation of eyes. The signs and symptoms are oral lesions,urogenital lesions, cutaneous lesion, blurring of vision, abdominal pain, diarrhoea, gastrointestinal ulceration. Multidisciplinary approach is needed to treat the patients with Bahcet's disease and supportive care to build confidence in the patient and family.

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