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Case Reports

An Adolescent Chinese Boy with Behçet's Disease

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Abstract Behçet's disease is a rare childhood disease. Early establishment of the diagnosis is important both for the institution of appropriate treatment as well as for recognition of potentially disabling complications. We report this condition in an adolescent Chinese boy who presented with recurrent oral ulcers, uveitis, erythema nodosum as well as arthritis, epididymitis and superficial thrombophlebitis.

Key words Behçet's syndrome; Erythema nodosum; Oral ulcer

Case Report

A 12-year-old boy presented with prolonged fever for two weeks. He enjoyed good past health until 10 years of age when he noticed on and off painful red nodules over both upper and lower limbs. These nodules usually lasted for one to two weeks and resolved spontaneously. At the time of presentation, he also had painful nodules over his left leg. Accompanying the fever was right ankle swelling. Past history revealed recurrent acute left scrotal pain since he was 10 years old. During the first episode, left scrotal exploration revealed a normal left testis with an enlarged epididymal head.

The following investigations were performed including a normal complete blood count. Erythrocyte sedimentation rate was elevated to 93 mm in one hour. ASOT was 120 Todd units. Both rheumatoid factor and antinuclear antibody were negative. Rheumatic fever was suspected at the initial stage and patient was treated with a 10-day course of penicillin. However, he continued to have recurrent joint swelling involving the knees, ankles and elbows. Treatment with naproxen only provided transient relief of joint symptoms. At 13 years of age, he developed an acute red and painful eye and anterior uveitis was diagnosed by ophthalmologist and topical corticosteroid was applied. Multiple superficial aphthous ulcers were also noted within the oral cavity. These oral ulcers were recurrent with remissions and exacerbations several times a year and their occurrence preceded the administration of oral methotrexate as part of the treatment for the chronic arthritis.

Doppler ultrasonography over the lower limb skin nodules confirmed evidence of superficial thrombophlebitis. The diagnosis of Behçet's disease was established based on the clinical features of recurrent oral ulcers, anterior uveitis, erythema nodosum, as well as other manifestations of non-erosive arthritis, recurrent epididymitis and superficial thrombophlebitis. Pathergy test was negative. HLA typing revealed the presence of HLA-B51 allele. He was subsequently started on colchicine and aspirin for the recurrent superficial thrombophlebitis. He responded well to colchicine with marked improvement of oral ulceration and there was no further exacerbation of arthritis or skin lesions upon follow up.

Discussion

Behçet's disease[1-3] is a recurrent systemic vasculitis affecting various body organs. It is prevalent along the ancient Silk Road, the Mediterranean and in Japan. Turkey has the highest prevalence with 80-300 cases per 100,000

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population. It is rare in Western countries. The actual prevalence in Chinese population has not been documented.

The onset of the disease is typically in the third decade of life, although it can occur at any age. The aetiology of the disease is still unknown, but various causes have been attributed. Genetic influence plays a role, as the incidence of HLA-B51 is significantly higher among patients with Behçet's disease along the Silk Road region and in Japan. A significantly increase in the incidence of HLA-B51 is also observed among the Chinese population,[4] as was observed in our patient. An infective origin has also been implicated. Bacterial infection like *Streptococcus sanguis* and viral agent like Herpes simplex virus type one have been postulated as the possible causative agents, but none of them have been proven so far.

Vascular injuries, hyperfunction of neutrophils and autoimmune responses form an integral part in the pathogenesis of Behçet's disease. The inflammatory response is probably triggered by the hyperfunction of neutrophils resulting in enhanced chemotaxis and cytokines production which bring about tissue inflammation and vascular injuries.

Behçet's disease has a wide spectrum of clinical presentation and there are no pathognomonic laboratory findings. The diagnostic criteria was proposed by the International Study Group for Behçet's disease in 1990 (Table 1).[5] According to the criteria, the diagnosis of Behçet's disease is established in the presence of recurrent oral ulceration plus at least two of the following: recurrent genital ulceration, eye lesions, skin lesions and a positive pathergy test. The pathergy test consists of pricking a sterile needle into the patient's forearm. The result is considered positive if a sterile erythematous papule of more than 2 mm developed between 24 to 48 hours. The positive response indicates mononuclear cells accumulation causing the skin hyperreactivity.

Oral ulceration is the most common symptom in patients with Behçet's disease and may present as the initial symptom. It takes the form of recurrent painful oral ulcers. The typical lesion is round with sharp, erythematous border with the surface covered by a pseudomembrane. Lesions usually heal within two weeks without scarring, but sometimes it may be confused with oral mucositis caused by methotrexate toxicity.

Genital ulcers are also frequent and occur on the scrotum and penis in men and on the vulva in women. They are painful and heal with scars. Epididymitis is a recognized feature which presents as an acute scrotal swelling and recurs occasionally. Erythema nodosum typically occurs in the shin area and is painful. They usually resolve spontaneously, leaving a hyperpigmented area.

Ocular lesions can occur both in the uvea and retina, with the latter being potentially sight threatening. Arthritis typically non-erosive and non-destructive is also common. Vasculitis can involve vessels of all sizes. Venous involvement includes superficial thrombophlebitis and deep vein thrombosis. Arterial involvement with aneurysms and occlusions are less common, but can cause significant morbidity, especially with pulmonary arterial aneurysm a potentially lethal complication.

Another potentially serious disabling complication is central nervous system involvement with either acute meningoencephalitis or slowly evolving pyramidal and brainstem signs resulting in progressive irreversible disability.

The choice of treatment in Behçet's disease depends on the severity of symptoms and the affected body parts. The mucocutaneous lesions are most common and topical corticosteroids are useful for both oral and genital ulcers. Colchicine has been shown to be effective in treatment of

Table 1 Criteria for the diagnosis of Behçet's disease

Recurrent oral ulceration	Minor aphthous, major aphthous, or herpetiform ulceration observed by physician or patient, which recurred at least three times in one 12-month period
Plus 2 of:	
Recurrent genital ulceration	Aphthous ulceration or scarring, observed by physician or patient
Eye lesions	Anterior uveitis, posterior uveitis, or cells in vitreous on slit lamp examination; or Retinal vasculitis observed by ophthalmologist
Skin lesions	Erythema nodosum observed by physician or patient, pseudofolliculitis, or papulopustular lesions; or Acneiform nodules observed by physician in postadolescent patients not on corticosteroid treatment
Positive pathergy test	Read by physician at 24 to 48 hours

mucocutaneous lesions presumably by inhibition of neutrophil function, especially chemotaxis.[6] Successful use of thalidomide has been documented in the treatment for oral and genital ulcers,[7] but its potential adverse effects limit its widespread use. The devastating ocular, central nervous system manifestation and potentially lethal pulmonary artery aneurysm warrant consideration of systemic corticosteroids and other immunosuppressive agents like cyclosporin, azathioprine and cyclophosphamide. Antiplatelet agents such as aspirin and dipyridamole are indicated in arteritis and venous thrombosis.

Behçet's disease usually runs a remission and exacerbation course. Significant morbidity occurs in patients with ocular and neurological involvement. Vascular complication, especially pulmonary artery aneurysms, in general have a bad prognosis. A 10-year survey of mortality in patients with Behçet's disease revealed that the observed mortality in the 15 to 24-year-old age group was significantly above that expected in the general population,[8] indicating that Behçet's disease is a cause of increased mortality in young patients.

In conclusion, Behçet's disease though a rare condition in the paediatric population, shares many similar clinical features as the adult patient.[9,10] They often pose a diagnostic problem because of the many overlapping features with other autoimmune diseases. However, early recognition of this rare disease in children is important for prompt institution of appropriate treatment as well as for surveillance of potentially disabling complications.

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