Behçet disease

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Abstract

In 1937, Professor Hulusi Behçet, a Turkish dermatologist, described two cases of relapsing ulceration of the mouth, eye and genitalia, a triple symptom complex that now characterizes the multisystem disorder that was named after him. The periodic relapses led Behçet to attribute these symptoms to a viral infection. This was subsequently found to be inaccurate and current evidence now favours a multifactorial aetiology with an immunological basis.

Keyword

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Introduction

In 1937, Professor Hulusi Behçet, a Turkish dermatologist, described two cases of relapsing ulceration of the mouth, eye and genitalia\cite{1,2}, a triple symptom complex that now characterizes the multisystem disorder that was named after him. The periodic relapses led Behçet to attribute these symptoms to a viral infection. This was subsequently found to be inaccurate and current evidence now favours a multifactorial aetiology with an immunological basis.

Behçet disease

Behçet correctly made the association between the oral, ocular, genital and skin lesions in the patients he described in his original paper. However, he attributed the periodic relapsing course of the disease to a viral infection, a theory that is now known to be incorrect. Although infection remains an important differential in our diagnosis of patients complaining of oral and genital ulceration with ocular involvement, with studies showing an increase in the streptococcal flora in this cohort of patients, current evidence has shown that the underlying pathogenesis of Behçet disease is complex and it is likely that an environmental stimulus triggers an abnormal immune response in a genetically susceptible host\cite{3}. The disease is more prevalent in those originating from areas surrounding the old silk trading routes, extending from the Middle East to China.
**Diagnosis**

Behçet’s observation that the ulceration in his patients was unresponsive to topical treatment correctly led him to conclude that the disorder had an underlying systemic basis. His report of non-specific findings from histological samples and haematological tests translates similarly to our findings today, with the diagnosis of Behçet disease largely being made on clinical grounds when associated symptoms meet the International Study Group Criteria\(^4\). In summary, in order to be diagnosed with Behçet disease, there must be at least three episodes of recurrent oral ulceration (either herpetiform or aphthous) in one year, as well as two of the following in the absence of other clinical explanations: recurrent genital ulceration, ocular inflammation, defined skin lesions or a positive pathergy reaction. There is no uniformly agreed diagnostic laboratory test for Behçet disease. Genetic studies have shown an association with HLA-B51 in 60% of patients\(^5\). Laboratory investigations reflect the inflammatory nature of the condition and include raised immunoglobulin levels particularly IgA and raised markers of inflammation erythrocyte sedimentation rate (ESR) and C-reactive protein. A positive pathergy reaction is characterized by the development of an erythematous papule or pustule at any site where the skin is traumatized. It is a feature rarely seen in patients originating from northern Europe or the United States, but is positive in up to 70% of those from Turkey or Japan\(^6\).

**Treatment**

In the first case described in his paper of 1937, Behçet reports a positive response to gold and trivalent arsenic. He explains that “thanks to this therapy the relapses become less frequent and the pains diminished”. There is currently no curative treatment for Behçet syndrome and treatment is directed against preventing irreversible organ damage. Mucocutaneous disease is usually treated with topical agents such as topical steroids. Systemic disease is controlled with immunosuppressive therapy. Examples include azathioprine, which has been shown to be beneficial in refractory cases, and has also been used in venous vessel disease, neurological and gastrointestinal disease. Colchicine is particularly useful for joint disease and erythema nodosum-like nodules\(^7\).

**Conclusions**

Since 1937, when Behçet disease was first described, we now know much more about the systemic nature of the condition. Unfortunately there is still no gold standard test to confirm the diagnosis and currently no curative treatment. Modern-day treatment aims to prevent irreversible organ damage and alleviate exacerbations of mucocutaneous, joint and constitutional symptoms that can significantly impair the patient’s quality of life.

**References**