

Picture Quiz: Answer

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Keywords

Behçet, neurological, venous, sinus, thrombosis.

1. The multiple, dusky, red lesions over the shins which were tender to palpation are typical of erythema nodosum. Common causes include sarcoidosis, post-streptococcal infection, tuberculosis and inflammatory bowel diseases.¹ Pregnancy, drugs and Behçet's disease are also recognised.¹
2. Superior sagittal sinus thrombosis (marked by arrows in Figure 3).
3. The likely diagnosis is Behçet's disease.
4. Diagnostic clues in this case include the patient's ethnicity, presentation with erythema nodosum, arthritis, systemic upset and mouth ulceration. However, it was actually the sagittal sinus thrombosis that prompted diagnosis, supported then by the development of unusual but characteristic pathergy lesions at venepuncture sites. Major clinical features of Behçet's disease also include genital ulcers, eye lesions (such as uveitis) and papulopustular skin eruption.

Case Outcome

The patient was anti-coagulated with low molecular weight heparin and then warfarin. He was also commenced on a reducing course of prednisolone. Symptoms and signs resolved within two weeks, and this was accompanied by normalisation of

inflammatory markers. He remains clinically well and is now on methotrexate.

Discussion

Behçet's disease is named after Helusi Behçet, a Turkish Dermatologist who described the triad of aphthous ulcers, genital ulceration and hypopyon uveitis in 1937.² Aetiology remains unknown, though epidemiological studies implicate environmental and genetic factors. HLA B51 gene carriage increases the relative risk of the condition by up to 6.7.³ There are also geographical patterns of prevalence: Behçet's has been known as 'The Silk Route' Disease as it is commonly diagnosed along the old trade routes extending from the Mediterranean to Indo-China. This may reflect dissemination of the HLA B51 gene by Silk traders in the past.

Prevalence varies from 4 per 100,000 in the UK⁴ up to 37 per 100,000 in parts of Turkey.⁵ Onset frequently occurs between the ages of 20-40⁶ and male to female ratio varies geographically, with 2:1 male preponderance in Turkish patients.⁷

Behçet's is a multi-system disorder which may present with rheumatological, ophthalmological, dermatological, neurological, gastroenterological, pulmonary and haematological complications.

The International Study Group for Behçet's Disease⁸ has defined criteria for diagnosis which include:

- Recurrent aphthous ulcers, at least three times in a twelve month period with at least two of:

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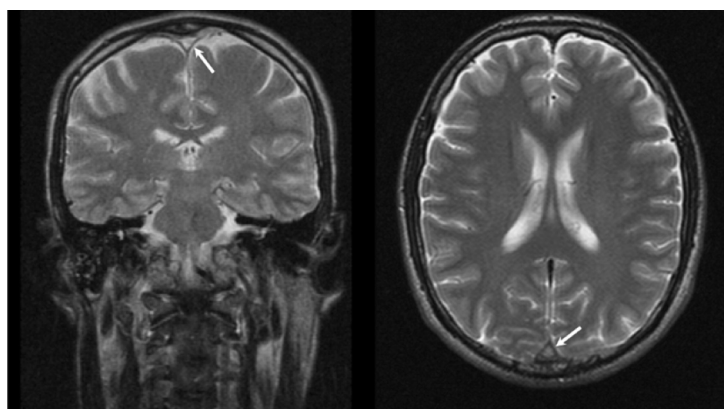


Figure 3. Coronal and axial T2-weighted magnetic resonance images showing sagittal sinus thrombosis (marked by arrows).

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- Genital ulceration
- Typical defined eye lesions such as uveitis or retinal vasculitis
- Typical defined skin lesions particularly erythema nodosum and papulopustular eruption
- Positive pathergy test
- Minor criteria to aid diagnosis include:
 - Arthritis
 - Deep venous thrombosis
 - Superficial thrombophlebitis
 - Epididymitis
 - Positive family history
 - Gastrointestinal involvement
 - Central nervous system involvement
 - Vascular involvement

Neurological manifestations occur in 5–10% of patients with Behçet's, are generally confined to the central nervous system (CNS) and are classified into two groups depending on CNS parenchymal involvement.^{9,10} These groups should be considered separately as they are clinically and prognostically distinct.¹⁰

The more common “parenchymal” group consists of vascular-inflammatory CNS disease, with focal or multifocal parenchymal involvement. It usually presents as a brainstem syndrome and may be accompanied by

sphincter disturbance and cognitive and behavioural changes.¹¹ More rarely it can cause a hemiparesis, isolated cranial nerve palsy or multiple sclerosis-like picture.¹¹ The clinical course usually consists of attacks and remissions with prognosis worsening with total number of attacks.¹⁰

The “non-parenchymal” group usually presents with venous sinus thrombosis and raised intracranial pressure.¹⁰ Brain MRI may identify the venous sinus thrombosis but is otherwise normal. The prognosis is significantly better than for “parenchymal” neuro-Behçet's.¹¹

Treatment of Behçet's is with steroids and other immunosuppressants such as azathioprine and methotrexate. There is a lack of evidence from randomised trials regarding the management of neuro-Behçet's¹² and the role of anticoagulation for venous sinus thrombosis in this context is controversial.^{10,12,13} We elected to anticoagulate our patient in view of the extensive nature of the sagittal sinus thrombosis.

In summary, we have presented the case of a patient who fulfilled the diagnostic criteria for Behçet's disease with non-parenchymal neurological involvement. This case demonstrates that clinical features may evolve, and that an initial diagnosis depends on a high index of suspicion and pattern recognition. The importance of genetic ethnic background and full history is emphasised, as the diagnosis of Behçet's is based on a constellation of disparate symptoms and signs.

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