

Behçet's Disease: Different Systemic Manifestations at Different Ages

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ABSTRACT

Behçet's disease, an inflammatory condition, can involve various systems. The disease usually manifests with dermatologic and ocular signs but can also cause serious symptoms due to pulmonary or neurologic involvement. Although the onset may occur at any age, it typically emerges in the second to fourth decades of life. As in the case presented here, Behçet's disease can manifest with the central nervous system involvement early in life and pulmonary involvement in adulthood.

Keywords: Behçet's disease, central nervous system, pulmonary system.

Introduction

Behçet's disease is a rare inflammatory disease of unknown origin, characterized by recurrent oral and genital aphthous ulcers, uveitis, and skin lesions [1]. Although Behçet's disease is more likely to present with the aforementioned symptoms, it is not a chronic, persistent inflammatory disease but rather is caused by recurrent attacks of acute inflammation. Involvement of the gastrointestinal tract, the central nervous system, and large vessels can be life threatening but is less common [2, 3]. The etiology of Behçet's disease remains unclear, although environmental factors such as genetic predisposition (HLA-B5 alleles) and infectious agents are thought to play a role in the pathogenesis [4, 5].

Case Presentation

A 23-year-old male patient was admitted to our unit with complaints of shortness of breath, cough, chest pain, and hemoptysis. The patient written informed consent was obtained. His medical history included mild/moderate mental retardation and hemiplegia detected after birth, and 2 years ago he had been hospitalized four times at another center for symptoms of lung infection without bloody sputum. The patient's current complaints had started 20 days ago. Right hemiplegia and ulcerated lesions in the mouth and genital region were observed on physical examination. Sporadic inspiratory rales were heard on lung auscultation.

The patient was cooperative and oriented during follow-up examinations. He did not exhibit hemoptysis during follow-up. Pathergy tests conducted at 24 and 48 hours were positive. Chest computed tomography showed pulmonary artery aneurysms and pulmonary thrombosis considered consistent with Behçet's disease (Figure 1A, 1B). To evaluate neurological involvement, an Magnetic resonance imaging (MRI) was conducted in which the left half of the brain stem and left cerebral hemisphere appeared hemiatrophic. Furthermore, a porencephalic cyst formation near the left lateral ventricle was noted in addition to parenchymal cystic, encephalomalacia, and gliotic changes adjacent to the porencephalic cyst (Figure 1C). Three-dimensional time-of-flight (TOF) magnetic resonance angiography showed a marked reduction in the left medial cerebral artery (MCA) calibration compared to the right MCA calibration (Figure 1D).

The Rheumatology department evaluated the patient as "presumed Behçet's disease." The treatment was initiated with 1 g of methylprednisolone for 3 days, then continued with 60 mg of steroid as a maintenance therapy. An initial dose of endoxan therapy was given and subsequent doses were scheduled at 21-day intervals.

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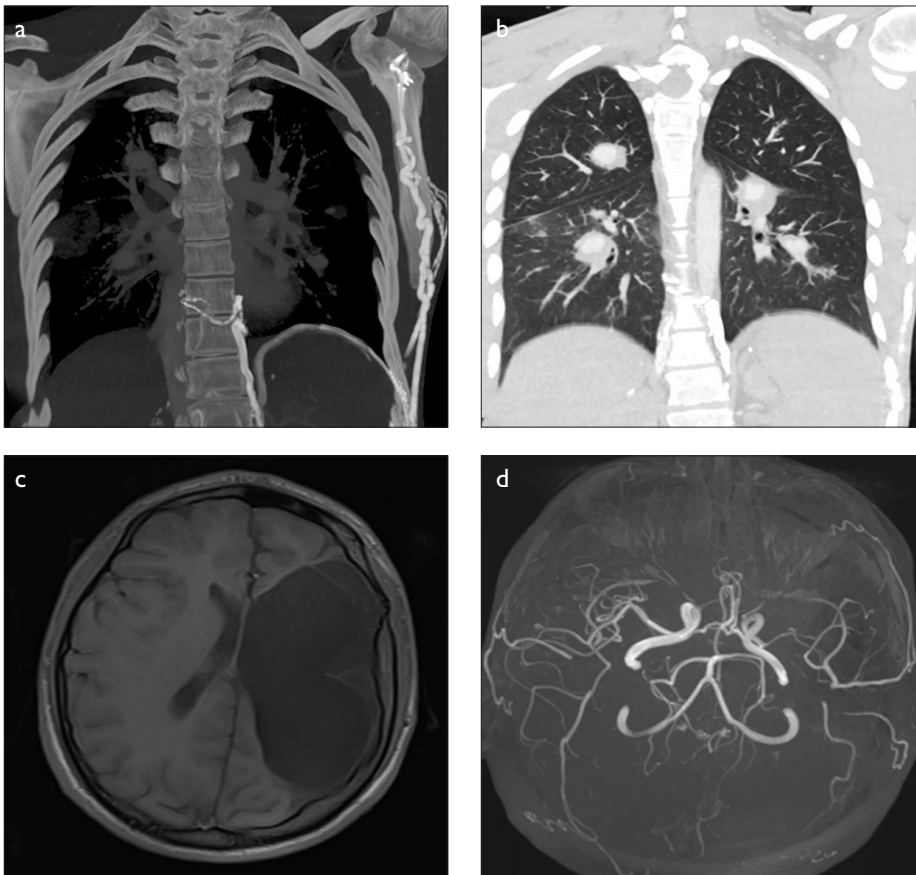


Figure 1. a-d. Coronal MPR MIP (a) and coronal MPR parenchymal window (b), chest CTA, T1 TSE transverse (c), and TOF 3D multislab MIP transverse brain MRI (d)

Laboratory tests indicated increased sedimentation and elevated C-reactive protein level. The patient was determined to be HLA-B5-positive in Deoksiribo Nükleik asit DNA typing analysis. After a marked reduction in his complaints, the patient was discharged.

Discussion

This case is noteworthy because although Behçet's disease usually does not cause symptoms in young patients, in rare cases it can cause vascular occlusion and give rise to neurological symptoms from birth.

Behçet's disease is a systemic inflammatory disease with a protracted course featuring recurrent attacks that can involve multiple organs [6]. In addition to skin and mucosal symptoms, patients may develop ocular, articular, vascular, gastrointestinal, cardiac, pulmonary, and neurological involvement. The disease affects both sexes approximately equally and typically emerges when patients are 20-40 years of age. Male gender and early onset are considered indicators of poor prognosis [7]. Though more common in young adults, Behçet's disease can also be seen in neonates. Intrauterine growth retardation and transient Behçet's syndrome have been reported

as early manifestations [8-10]. The central nervous system involvement has also been reported in children, but is rare [11].

The disease is a major cause of morbidity, both with skin and mucosal involvement and in some cases with other organs and systems involvement. The involvement of the large vessels is also associated with a higher mortality rate. Death from Behçet's disease usually occurs in case of large vessel involvement such as pulmonary artery aneurysm, gastrointestinal involvement, and neurological involvement [12].

Neurological involvement occurs in 2.2-50% of patients with Behçet's disease [13]. The prevalence of neurological involvement is twice as high in male patients than in female patients. Neurological involvement is often described as brain stem or corticospinal tract syndromes, increased intracranial pressure often associated with venous sinus thrombosis or aseptic meningitis, behavioral disorders, or isolated headaches. Less common signs of neurological manifestations of Behçet's disease include intracerebral hemorrhage due to ruptured aneurysm, peripheral neuropathy, isolated optic neuritis, and Parkinsonian syndromes. These symptoms

are usually seen in adulthood and are rarely observed in children [11, 14].

Intrathoracic findings of Behçet's disease include vena cava superior syndrome, thromboembolism of mediastinal veins, aortic and pulmonary artery aneurysms, pulmonary infarction and hemorrhage, pleural effusion, and more rarely myocardial and pericardial involvement, cor pulmonale, and mediastinal or hilar lymphadenopathy. Episodes of dyspnea, cough, chest pain, and hemoptysis may occur as a result. The simultaneous formation of thrombus in the pulmonary artery and right ventricle has been reported in numerous studies [15].

In contrast to typical neurological manifestations of Behçet's disease, our patient exhibited hemiatrophy of the left half of the brain stem and the left cerebral hemisphere, which was likely congenital. In addition, he exhibited a porencephalic cystic formation near the left lateral ventricle, as well as parenchymal cystic, encephalomalacia, and gliotic changes adjacent to this cystic formation. Angiographic examination revealed substantially reduced left MCA calibration compared to the right MCA. Our patient also had extensive aneurysms in the pulmonary arteries and thrombus in some aneurysms and the right ventricle, consistent with other cases reported in the literature.

The onset of Behçet's disease may occur at any age and is most likely at 20-40 years of age; however, some manifestations cause symptoms from birth. As in this case, Behçet's disease may affect the central nervous system development, affect the pulmonary system in young adulthood, and cause involvement of various systems emerging at different ages.

Informed Consent: Informed consent was obtained from the patient included in the study.

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