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Behçet's disease and late progressive myelopathy in a patient with severe kyphotic deformity following childhood spinal trauma: is there any connection?

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Abstract A highly unusual case is presented of a patient affected by Behçet's disease and severe kyphosis in the thoracic spine, who had a history of traumatic spinal cord injury during childhood. A review of the literature is also presented. Magnetic resonance imaging showed spinal cord compression at the level of the apex of the kyphosis in the upper thoracic spine. Although there is no information indicating that the coexistence of post-traumatic spinal deformity with Behçet's disease is not just a coincidental finding, the possibility that it was the presence of Behçet's disease concomitant with the antecedent spinal trauma that may have triggered the severe kyphotic deformity cannot be ruled out.

Keywords Spinal cord injury · MRI · Kyphosis · Behçet's disease

Introduction

Post-traumatic spinal deformity is a well-known complication of spinal injury occurring in patients with dislocations or fractures of the spine [2, 4, 5, 9, 10]. However, to my knowledge, there is rarely a deterioration of spinal cord function in patients with kyphosis, and late progressive myelopathy has been reported only in only a few cases [3]. Neurological problems can, on the other hand, be associated with Behçet's disease – a multisystem disease first described by the Turkish dermatologist Hulusi Behçet in 1937, with ocular, oral and genital manifestations due to a systemic vasculitis [1, 7]. Neurological findings of Behçet's disease include intracranial hypertension, meningoencephalitis, and symptoms due to involvement of the peripheral or central nervous system [1, 7, 8]. In this study, an interesting case of a patient affected by Behçet's dis-

ease and severe kyphosis in the thoracic spine, with a history of traumatic event during childhood, is reported.

Case report

A 44-year-old man presented with a 4-month history of sphincter disturbances, associated with gait disturbance and numbness in the lower extremities that had been gradually increasing over the preceding 2 years. He had suffered from progressive kyphosis since experiencing a significant spinal trauma in the thoracic region at the age of 2 years, and he had no history of tuberculosis or other infection affecting spine. The patient had been taking 1.0 mg per day of colchicine for a diagnosis of Behçet's disease.

The general physical examination was normal, except that the patient had a kyphotic trunk. Neurological examination revealed a grade 3 spastic paraparesis and bilateral Babinski sign together with hypesthesia below the waist with preservation of joint position sense and increased deep tendon reflexes in the lower extremities. The results of a laboratory evaluation were within normal

Fig. 1 T1-weighted sagittal (*left*) and axial (*right*) magnetic resonance (MR) images display a 150° kyphotic deformity in the upper thoracic spine that displaces medulla spinalis anteriorly, causing severe compression over the apex of the kyphos

Fig. 2 T2-weighted sagittal (*left*) and axial (*right*) MR images show the compression of the spinal cord from the anterior aspect. Note the absence of any delayed syrinx formation in the spinal cord above and below

limits. Complete radiographic evaluation of the thoracic spine was performed, including plain radiographs, computed tomography (CT), and magnetic resonance (MR) imaging. The radiographic examination revealed a severe thoracic kyphosis of 150°, the magnitude of which had remained stable over the preceding 2 years. MR imaging of the thoracic spine confirmed a severe kyphotic deformity in the upper thoracic region, producing anterior cord compression over the apex of the kyphosis (Fig. 1, Fig. 2).

The patient required decompression of the spinal cord, correction of the kyphosis and spinal fusion to obtain stabilisation. After a thorough review of the neuroradiologic studies, and long discussion with the patient regarding the risks and benefits of the various treatment options, however, he refused surgical intervention. The patient received further conservative management consisting of oral steroids, but no improvement in his condition was achieved.

Discussion

International study group criteria for a diagnosis of Behçet's disease (oculo-oral-genital syndrome) consists of recurrent oral aphthous ulcerations recurring at least three times in a 1-year period in the presence of two of the following:

genital ulceration, retinal vasculitis or uveitis (anterior or posterior), cutaneous lesions (erythema nodosum, acneiform nodules, papulopustular lesions), or a positive patergy test [6]. From a neurological point of view, it is a systemic vasculitis that is known to cause some manifestations due to cerebral, cerebellar, brainstem, spinal cord and cranial or peripheral nerve involvement [1, 7, 8]. Neurological involvement in this disease, which has a benign chronic course with relapses or fluctuations, occurs in 5–50% of the patients [8]. While the causative role of Behçet's disease in our patient with progressive myelopathy and severe kyphotic deformity caused by spinal trauma is questionable, the mechanism of the late neurological involvement is probably "vascular" secondary to Behçet's disease rather than "mechanical" secondary to kyphosis.

Untreated, the deformity may progress, producing rarely pain and/or neurological problems. Therefore, it is imperative to diagnose and treat the condition at an early phase. It is known that factors that may influence the onset of cord compression include the age of the patient, the number of segments involved, the angle of kyphosis, the presence of syrinx formation, and any impairment of vasculature of the cord [3, 4]. It is hypothesized that a patient with Behçet's disease is more prone to myelopathy in post-traumatic kyphosis, although the absence of any response to the treatment by medication other than the colchicine that the patient was already taking would exclude the Behçet's disease as contributing to the myelopathy. Furthermore, Behçet's disease was thought to be the cause of the myelopathy because the kyphotic deformity had been stable for the previous 2 years, and if the apex of the kyphos had been at a different level, the mechanical decompression would probably have little or no effect. Although the precise reason for the late progressive myelopathy developing in this patient can not be definitely stated, the presence of Behçet's disease concomitant with the antecedent spinal trauma could possibly be a triggering factor. Since I am not aware of any reported similar case, I thought it worth reporting this coexistence.

Conclusion

This report is concerned with a patient with a severe post-traumatic kyphotic deformity of the thoracic spine and concomitant Behçet's disease. In the case presented, it is unclear whether the presence of both post-traumatic spinal deformity and Behçet's disease is just a coincidental finding or whether there is a relationship between the two, because it is impossible to find out whether the underlying disease has anything to do with progressive kyphosis or whether the extent of the trauma that had occurred during childhood could by itself adequately explain the kyphotic deformity.

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