Anterior Spinal Cord Syndrome in a Patient with Behçet’s Disease

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Abstract

Although neurological involvement in Behçet’s disease is not so uncommon, isolated spinal cord disease is quite rare and reported to be observed in about 2% of all cases with neurological involvement. Here we report a Behçet’s patient with spinal cord disease presented with anterior spinal cord syndrome. This rare syndrome is caused by hypoperfusion of the anterior spinal artery and to our knowledge has not been previously reported in patients with Behçet’s disease. This report defines the characteristic clinical features of this entity and emphasizes the importance of early immunosuppressive treatment and initiation of rehabilitation.

Behçet’s disease (BD) is a chronic vasculitis of unknown etiology, which may involve many organs and systems. Although mucocutaneous lesions are more common, eye, vessels of any size, gastrointestinal system, and central nervous system can also be affected.1 Neurological involvement of BD—known as neuro-Behçet disease (NBD)—was first described in 1940s, and the incidence is being reported ranging from 5% to 50%.2,3 Various forms of neurological problems can be seen in BD, and can be classified in two major groups: The first, central nervous system (CNS) parenchymal involvement with a predilection to brainstem-diencephalic regions, is seen in the majority of patients with NBD. The second form is cerebral venous sinus thrombosis (CVST), which is seen in up to 20% of cases with neurological involvement.4 Although subacute meningoencephalitis accounts for 75% of cases with parenchymal involvement, different syndromes might be encountered during the course of parenchymal NBD. One such presentation of parenchymal disease can be seen in the form of spinal cord involvement. Involvement of the spinal cord as part of the diffuse type of parenchymal NBD pattern was reported in about 10% of NBD cases in some series, whereas isolated spinal cord disease is an uncommon presentation observed in about 2% of patients with neurological involvement.5,6 In patients with spinal cord disease, most common presentation is transverse myelitis, affecting the cervical and dorsal areas. Here we report a BD patient with spinal cord disease that is notable because he was presented with anterior spinal cord syndrome (ASCS). This rare syndrome is caused by hypoperfusion of the anterior spinal artery, leading to ischemia in the anterior two thirds of the spinal cord, and to our knowledge has not been previously reported in patients with BD.

Case report

A 22-year-old man with multiple oral aphthous lesions, giant genital ulcers, and paraplegia was admitted to the intensive care unit. He had been in good health and had no complaints related with BD until 5 days before his admission. Five days earlier, he had abrupt onset of oral and genital lesions and severe leg pain accompanying to his symptoms. He had been hospitalized in another hospital. Three days after the beginning of these symptoms, he suddenly developed weakness in both legs.
and therefore was referred to our hospital. In his initial physical examination, in addition to high fever (38.2 to 38.8°C), multiple major oral and genital ulcers and flask paralysis in both legs was detected. In sensorial examination, pinprick test as well as temperature sensation were disturbed below T11 level on the right and T6 level on the left. His superficial sensation, proprioception and deep tendon reflexes were normal. The extensor plantar responses were bilaterally absent. Sense of rectal filling was not disturbed but he had urinary retention. There were no motor or sensory deficits detected in both upper extremities. His cognitive and cerebellar functions were normal. Ophthalmologic examination revealed bilateral acute posterior uveitis. In laboratory investigation, erythrocyte sedimentation rate and C-reactive protein were 80 mm/h and 142 mg/dL, respectively. Immunological tests including antibodies against the SM, RNP, SS-A, SS-B, SCL-70, Jo-1 antigens, rheumatoid factor, and anti-cardiolipin antibodies were all negative. Activated partial thromboplastin time was within normal limits. All bacterial cultures and viral markers were found to be negative. His cranial and cervical magnetic resonance imaging (MRI) exams were normal. Thoracic spinal MRI revealed multiple discrete hyperintense cord lesions without cord swelling on T2-weighted images (Fig. 1). Furthermore, the patient had undergone conventional spinal angiography to check vascular pathology, which revealed patent spinal arteries (Fig. 2).

Considering the above mentioned clinical features, his presenting symptoms were sufficient to make a diagnosis of BD, and by demonstrating the ischemic infarction in spinal cord by means of MRI, we have established the diagnosis of anterior spinal cord syndrome associated with BD. Treatment consisted of 1,000 mg per day prednisolone (IV) for 5 days and 1,000 mg cyclophosphamide; (IV) infusion was started. Except for the neurological signs, other findings, including uveitis, were completely resolved within the first week of treatment. Consequently, maintenance therapy consisting of 60 mg per day of oral methylprednisolone and 1,000 mg per month of cyclophosphamide was begun. After completing 12 doses of cyclophosphamide, his treatment was switched to azathioprine (150 mg per day). Furthermore, he has been on a rehabilitation program since the first dose of cyclophosphamide. At last follow-up, he could walk with the aid of a walker and muscle strength in his legs was graded as 4 on a scale of 0-5 (where in grade 0 no movement

**Figure 1** Initial midsagittal (A) thoracic fat-suppressed T2-weighted magnetic resonance image shows longitudinally extensive T2 hyperintense lesion within the cord (arrow). B and C show corresponding transvers images at the level of T3 and T7, respectively.
is observed and in grade 5 the muscle contracts normally against full resistance). Magnetic resonance images taken at three months of the disease onset have revealed marked resolution of the previous cord lesions (Fig. 3).

**Discussion**

Anterior spinal cord syndrome comprises ischemia or infarction of the spinal cord in the distribution of the anterior spinal artery, which supplies the ventral two-thirds of the spinal cord. The typical presentation of ASCS is an acute and painful myelopathy. Clinical features include weakness and loss of pain and temperature sensation below the level of the involvement, with relative sparing of position and vibratory sensation. Bladder and bowel control is impaired in most of the cases. However, as seen in our case, sometimes a partial syndrome is seen with the gray matter of the cord preferentially affected, and in this situation, sphincter control is well preserved. Diagnosis of ASCS is based on the clinical examination and MRI findings. In this regard, the parenchymal lesions in the spinal cord (MRI) of our patient were consistent with infarction, which supports the diagnosis of ASCS.

It is a rare condition with a variety of precipitating factors. The syndrome most commonly results from diseases within the aorta, including aortic aneurysms, surgery of the aorta, aortic dissections, traumatic rupture, and emboli from atheromatous plaques. Other causes include sickle cell disease, polycythemia, protein S deficiency paradoxical embolism through a patent foramen ovale, vasculitis, and some infections (tuberculosis, schistosomiasis, and Neisseria meningitidis).

This young man had an aggressive and abrupt onset BD that involved mucocutaneous, ocular, and nervous system simultaneously. This case is of interest because of the simultaneous development of all BD related symptoms within days and the protection of proprioception as well as vibratory sensation despite the involvement of the spinal cord. Although parenchymal neurological involvement in patients with BD mostly manifests itself with a brainstem syndrome, occasionally spinal cord can also be involved. In those rare cases having spinal cord involvement, the most common presentation is transverse myelitis, which affects several segments of the cord longitudinally. To our knowledge, ASCS has not been previously reported in patients with BD. Despite extensive investigation, no risk factors that have been shown to be associated with ASCS were identified in our patient. In this regard, we think that the vasculitis of the
anterior spinal artery or its branches due to BD is the most likely abnormality that caused the thoracic cord lesion in our patient. Pathologic evidence of spinal cord involvement in the literature has not clearly shown a vasculitic process, while the information available is so scarce due to the limited number of histopathologic examinations. Therefore, any further comment on this issue would be highly speculative at this level of knowledge.

Given the rarity of the diagnosis, there is no consensus for any particular treatment approaches. Therefore, treatment of ASCS is directed at the underlying cause, if known. In the present case, we treated our patient as NBD with parenchymal involvement, while there have been no controlled or comparative trials of treatment of any aspect of neurological involvement in BD. High-dose corticosteroid therapy has been reported to be used in acute and sub-acute phases with variable effectiveness. Generally, response to corticosteroids seems to be efficient; however, there are some patients who showed a poor response. The timing of the treatment is the most important determinant of the prognosis; namely, early invention leads to better outcomes. Other immunosuppressive agents such as cyclophosphamide or interferon-α can also be used, as in other life-threatening involvements of BD. In our case, both high-dose steroid and cyclophosphamide were administered early in the course with resultant improvement in the neurological symptoms, albeit rather slowly.

In conclusion, this case indicates that spinal involvement of NBD can manifest itself as ASCS. Hence, BD with neurologic involvement should be considered as an etiological factor for ASCS, especially when the patient has a classical triad of oral and genital ulcerations with uveitis.

Disclosure Statement
None of the authors have a financial or proprietary interest in the subject matter or materials discussed, including, but not limited to, employment, consultancies, stock ownership, honoraria, and paid expert testimony.

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