A patient with Behçet's disease presenting with acute urinary retention

Sare Gülfem Akyüz[1], Aysun Çaltık[2], Mehmet Bülbül[1], Özlem Aydoğ[1], Gülay Demircin[3], Ayşe Aksoy[4], Berna Uçan[5]

Departments of [1]Pediatric Nephrology, [4]Pediatric Neurology, and [5]Radiology, Dr. Sami Ulus Children's and Maternity Hospital, and [2]Keçiören Training and Research Hospital, and [3]TOBB University Faculty of Medicine, Ankara, Turkey. E-mail: saredr@gmail.com

SUMMARY: Akyüz SG, Çaltık A, Bülbül M, Aydoğ Ö, Demircin G, Aksoy A, Uçan B. A patient with Behçet's disease presenting with acute urinary retention. Turk J Pediatr 2014; 56: 196-198.

Behçet's disease (BD) is a multisystemic inflammatory disorder of unknown etiology. Neurologic involvement is known to be the most devastating feature of BD. The frequency and types of neurologic involvement in the pediatric age group are not clear, and the available information is limited to case reports. Here, we report a BD patient who presented with urinary incontinence as the initial feature of spinal cord involvement.

Key words: Behçet's disease, urinary retention, spinal cord.

Behçet's disease (BD) is a multisystemic inflammatory disorder of unknown etiology[1].The reported incidence of neurologic involvement varies from 5-10% in non-selected series[2,3], and it may be parenchymal or non-parenchymal. Encephalomyelitis, spinal cord abnormalities, aseptic meningitis, and benign intracranial hypertension may be recognized in patients with BD[3]. Here, we report a 16-year-old boy who presented with urinary retention as a feature of spinal cord involvement of BD.

Case Report

A 16-year-old boy was admitted with urinary retention for three days. He had a history of BD for five years. He had been diagnosed as BD on the basis of recurrent oral and genital ulcerations and positive pathergy reaction. He had been using only colchicine, but had not presented for follow-up visits for two years.

In his physical examination on presentation, there were no mucocutaneous, musculoskeletal, cardiovascular, or ocular findings. On the neurological examination, his cognitive, cerebellar, cerebral, and cranial nerve functions were intact; deep tendon reflexes were normal in both upper and lower limbs. A Foley catheter was inserted.

On laboratory investigations, complete blood count, acute phase reactants, and renal and liver function tests were in normal limits. Urine analysis showed eumorphic erythrocytes. Abdominal ultrasonography and echocardiography were normal.

During his follow-up, he developed paresthesia of the legs, difficulty walking and gait disturbance. Motor function in the legs was diminished bilaterally (3/5). Deep tendon reflexes were exacerbated, and cremasteric and abdominal reflexes were absent. Cerebrospinal fluid (CSF) examination could not be done because his family did not give permission. Cranial magnetic resonance imaging (MRI) was normal. MRI of the lumbar spine was normal on T1- and T2-weighted images, but revealed contrast enhancement after gadolinium injection, and these findings were consistent with myelitis (Fig. 1).

According to the patient's history of BD and lumbar spine MRI findings, our patient was diagnosed as neuro-Behçet's disease (NBD) with spinal cord involvement.

He was treated first with intravenous (i.v.) pulse methylprednisolone (30 mg/kg/day, for 3 days). Motor functions in his legs normalized, but urinary retention did not resolve. Thus, i.v. pulse cyclophosphamide therapy (750 mg/m^2/monthly, twice) was added, and clean intermittent catheterization

for urinary retention was continued. We also continued tapering the dose of prednisolone orally (starting dose of 2 mg/kg/day, tapered to 0.75 mg/kg/day in 45 days). His muscle strength had almost fully recovered in the second month of the treatment, but he still required clean intermittent catheterization.

Discussion

Behçet's disease (BD) is a multisystemic inflammatory disorder of unknown etiology[1]. A higher incidence in families with more than one member with BD suggests a genetic anticipation[4]. Although the usual onset of BD is in the third or fourth decade, pediatric cases have also been reported[4,5]. In addition to mucocutaneous and ocular involvement, BD may also involve the joints, blood vessels, and gastrointestinal and nervous systems[4,5].

Neurologic involvement was reported in 5-10% of the cases in a large series of BD[2]. NBD is mainly in two major forms: parenchymal central nervous system involvement and cerebral venous sinus thrombosis (CVST). CVST mainly occurs in the pediatric age group as compared to adult-onset NBD, in which parenchymal neurologic involvement is more frequent. The frequency and types of the neurologic involvement in the pediatric age group are not clear, and available information is limited to case reports[5,6].

In NBD, while the brainstem and diencephalon are affected frequently, spinal cord involvement is detected rarely[2]. Spinal cord involvement in NBD has been known since the 1950's[2]. Whereas initial reports on this issue were based on histopathological studies, cases reported more recently were diagnosed by spinal MRI scans[4]. The rate of spinal cord involvement has varied between 2.5-30% in various studies. In a study of Uludüz[5], only one child had spinal cord involvement among 26 patients with pediatric BD. Her neurological picture was consistent with a relapsing myelitis, resolving without sequelae over many years. In a study of Shakir[7], neurologic involvement of BD was reviewed, and paraparesis, sensory deficits indicating a spinal level, sphincter and/or sexual dysfunction, and CSF lymphocytic pleocytosis were reported to be cardinal features of spinal cord involvement. Persistent disability was only noted in patients with spinal cord involvement. In our patient, although the motor function in his legs normalized, urinary retention has not yet resolved, and he is still on treatment with clean intermittent catheterization in the first month of the treatment.

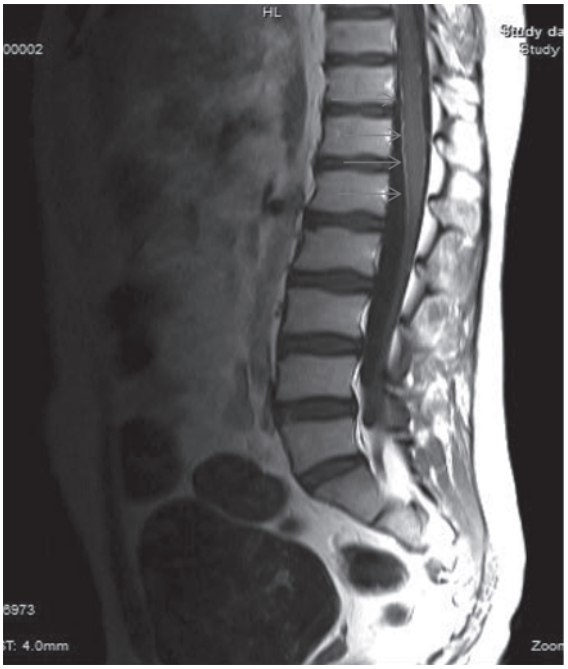

Fig.1. Continuous diffuse pial postcontrast enhancement is observed at the level of T11-L1 A, at T1 weighted sagittal magnetic resonance imaging (thin arrows).

There are no controlled studies concerning the treatment of pediatric NBD. Treatment focuses on the reduction of the inflammation that causes vascular or parenchymal lesions. Treatment choices in neurological diseases are based mainly on anecdotal reports and experience[8]. High- dose corticosteroids are given during attacks, followed by maintenance oral corticosteroids tapered over 2-3 months[5]. If the patient has poor prognostic factors (spastic paraparesis, sensory deficits, sphincter and/or sexual dysfunction, CSF lymphocytic pleocytosis, or persistent disability) or a second neurologic episode has occurred, another immunosuppressant agent such as azathioprine, i.v. cyclophosphamide or methotrexate should be added[8]. Interferon (IFN)-α and tumor necrosis factor (TNF)-α antagonists have been used with some success in resistant cases[9-12]. Because the urinary incontinence of our patient did not resolve with pulse i.v. methylprednisolone, we added high-dose i.v. cyclophosphamide.

In conclusion, neurologic involvement is known to be the most devastating involvement in BD. Spinal cord involvement is very rare, and might be overlooked. Considering the dismal outcome of spinal cord presentation in young males, early and vigorous treatment is recommended in this group of NBD.

REFERENCES

1. Yazıcı H, Yurdakul S, Hamuryudan V. Behçet's syndrome. Curr Opin Rheumatol 1999; 11: 53-57.
2. Al-Araji A, Kidd DP. Neuro-Behçet's disease: epidemiology, clinical characteristics and management. Lancet Neurol 2009; 8: 192-204.
3. Akman-Demir G, Serdaroğlu B, Taşçi B; Neuro-Behçet Study Group. Clinical patterns of neurological involvement in Behçet's disease: evaluation of 200 patients. Brain 1999; 122: 2171-2181.
4. Yeşilot N, Mutlu M, Güngör O, Baykal B, Serdaroğlu P, Akman-Demir G. Clinical characteristics and course of spinal cord involvement in Behçet's disease. Eur J Neurol 2007; 14: 729-737.
5. Uludüz D, Kürtüncü M, Yapıcı Z, et al. Clinical characteristics of pediatric-onset neuro-Behçet disease. Neurology 2011; 77: 1900-1905.
6. Siva A, Saip S. The spectrum of nervous system involvement in Behçet's syndrome and its differential diagnosis. J Neurol 2009; 256: 513-529.
7. Shakir RA, Sulaiman K, Kahn RA, Rudwan M. Neurological presentation of neuro-Behcet's syndrome: clinical categories. Eur Neurol 1990; 30: 249-253.
8. Hatemi G, Silman A, Bang D, et al.; EULAR Expert Committee. EULAR recommendations for the management of Behçet disease. Ann Rheum Dis 2008; 67: 1656-1662.
9. Nichols JC, İnce A, Akduman L, Mann ES. Interferon-α2a treatment of neuro-Behçet disease. J Neuroophthalmol 2001; 21: 109-111.
10. Fujikawa K, Aratake K, Kawakami A, et al. Successful treatment of refractory neuro-Behçet disease with infliximab: a case report to show its efficacy by magnetic resonance imaging, transcranial magnetic stimulation and cytokine profile. Ann Rheum Dis 2007; 66: 136-137.
11. Alty JE, Monaghan TM, Bamford JM. A patient with neuro-Behçet's disease is successfully treated with etanercept: further evidence for the value of TNF-α blockade. Clin Neurol Neurosurg 2007; 109: 279-281.
12. Ribi C, Sztajzel R, Delavelle J, Chizzolini C. Efficacy of TNF-α blockade in cyclophosphamide resistant neuro-Behçet disease. J Neurol Neurosurg Psychiatry 2005; 76: 1733-1735.