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Case Report

Drowsy on the Silk Road-A Case of Neuro-Behcet's Disease

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Introduction

Behcet's disease is a chronic, multisystem inflammatory disorder characterized by recurrent oro-genital ulceration, ophthalmologic, neurologic, or dermatologic manifestation. There is no pathognomonic laboratory test available therefore the diagnosis can be challenging, especially in patients with limited clinical features. Nervous system involvement termed as Neuro-Behcet's is a relatively rare occurrence. It is sub-classified into two major forms i.e. parenchymal and non-parenchymal. Here we describe a case of Neuro-Behcet's in a young woman with a rather bizarre presentation.

Case

A 38-year-old female presented to the Emergency Department with the complaint of altered sensorium for 3 days. This was preceded by bilateral eye redness for 7 days, and low grade fever for 5 days. Physical examination was notable for an irritable young woman with a GCS of E3V3M5 with no focal deficit on neurological exam. Ophthalmologic examination was consistent with anterior uveitis. MRI brain was performed which demonstrated multiple T2 high signal intensity areas in cortical and subcortical regions of bilateral frontal, parietal, occipital, temporal lobes, left caudate and lentiform nuclei, and both cerebellar hemispheres (Fig 1, 2). Majority high signal on T1. MRA and MRV showed normal vessels. Lumbar puncture was performed and CSF DR revealed increased protein content; CS, Viral PCR, MTB PCR, and Oligoclonal bands all negative.

She was diagnosed as a case of Behcet's disease 3 years ago when she had presented to Rheumatology clinic with a cold left foot associated with ankle swelling. CT angiogram left leg at that time was consistent with arterial thrombosis, demonstrating partial mural thrombosis in the distal abdominal aorta, non-opacification of left superficial femoral artery and partial thrombosis of left popliteal artery. 10 years prior to this presentation she had an episode of pan-uveitis which was treated with oral corticosteroids. Laboratory tests were significant for a positive fluorescent ANA but negative dsDNA, ENA profile, Anti-phospholipid antibodies, and ANCAs.

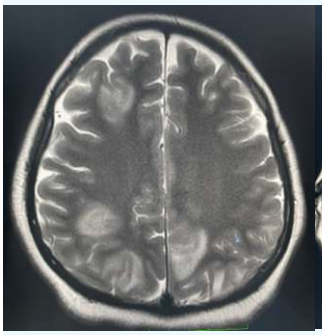

Figure 1

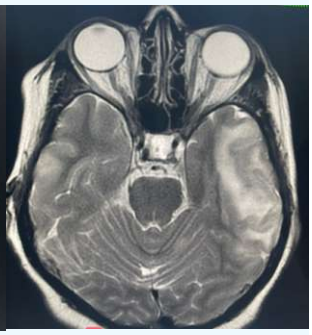

Figure 2

She was advised Methotrexate and Corticosteroids, but discontinued follow-up 2 years prior to the current presentation to the emergency department.

We started her on IV Methylprednisolone 500mg/day for 5 days. 2 days later, patient showed remarkable improvement in irritability and GCS improved to E4V4M6.

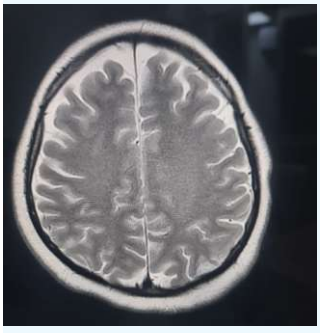

Figure 3

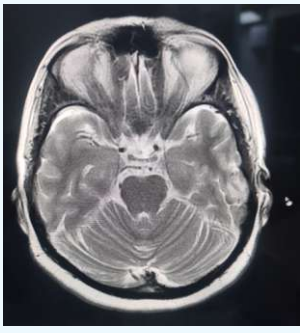

Figure 4

She was discharged on oral corticosteroids and methotrexate. One month later she was seen as an outpatient with follow-up MRI scan which showed resolution of previously identified lesions (Fig. 3, 4).

Discussion

Behcet's disease (BD) is an immune-mediated disorder that can involve multiple systems.[1] A small percentage of patients with Behcet's syndrome, approximately 10%, have neurological involvement. This is referred to as Neuro-Behcets disease (NBD).[2] Because of its rarity, there is limited data to provide high quality evidence for NBD. Published data majorly comprises of personal experiences and single-center studies. NBD should be considered in patients presenting with neurological symptoms who have history of recurrent oral or genital ulcers, uveitis, or other systemic features of BD. The gold standard for diagnosis of NBD is Magnetic Resonance Imaging.[3] Acute form of NBD manifests as meningoencephalitis and responds well to corticosteroids.[4] Neuro-Behçet's disease can either show parenchymal or non-parenchymal involvement. Parenchymal disease involves the brainstem, cerebral hemispheres, cerebellum or the diencephalon and makes up approximately 80% of Neuro-Behçet's disease cases.[5] Non-parenchymal disease includes venous sinus thrombosis, intracranial hypertension, acute meningeal syndrome, ischemic stroke, dissection or aneurysm.[5] Our patient presented with the parenchymal disease pattern. Parenchymal NBD lesions are typically demonstrated in the following areas on MRI in a descending order of occurrence; brainstem being the most common, and typically in the pons more than the basal ganglia (bilateral in one-third of cases), then thalamus, subcortical white matter and spinal cord.[6] Lesions are characterized by multiple small areas of high signal intensity on T2-weighted images. This case fulfills Definite NBD criteria of the international consensus[3] with her history of panuveitis, arterial thrombosis, recurrent oral aphthosis, and now presenting with multifocal involvement of the brain. Differential diagnoses of NBD commonly include Neuroarcoidosis, Multiple Sclerosis (MS), CNS infections, and Lupus Cerebritis. MS classically involves the periventricular regions which are most often spared in NBD[7] a as in our patient while neurosarcoidosis has a wide range of CNS manifestations, including leptomeningeal involvement and intraparenchymal mass like lesions.[8] Neuro-psychiatric SLE has a predilection for cerebral white matter mainly parietal and frontal lobes.[9] Our patient had these lobes involved but did not have other features to be classified as SLE according to the criteria. The CSF constituents are altered up to 70-80% in patients with NBD.[10] Typically there is pleocytosis and an increase in proteins,[11] and absences of oligoclonal zone, AQP4, MOG and MBP[12]. Glucose is usually normal, and low levels point toward CNS infections.[3] Ophthalmologic involvement can cause severe life - threatening consequences and may manifest as uveitis, retinitis, conjunctivitis, corneal ulcer, choroiditis, and optic neuritis.[13] Acute parenchymal NBD requires treatment with IV pulse methylprednisolone 1 gram per day followed by oral prednisolone 1mg/kg/day for a month, thereby 5-10 mg taper every 10-15 days along with steroid sparing agents drugs.14 Severe parenchymal disease, persistent or recurrent clinical course of NBD, despite adequate immunosuppressive therapy justifies use of biological agents.

Conclusion

This case report underscores the complexity and variability of Neuro-Behcet's disease presentation. It signifies early recognition and prompt intervention to prevent potentially devastating consequences of this rare disease. Most of the literature on Neuro-Behcet's disease across the globe comprises of personal experiences and reports; our case is a contribution to this growing body of literature to enhance better understanding of the disease.

Conflict of interest: *None*

Funding source: *None*

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