Neuro-Behcet’s Disease, a Diagnostic Challenge

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Abstract

Neuro-Behcet’s disease (NBD) is a rare neurological manifestation of the systemic small vessel vasculitis called Behcet’s disease. It can present in various ways with predilection for the brain stem, thalamo-hypothalamic regions, cerebellum and basal ganglia. In this case, we describe a case of young stroke that was later attributed to NBD.

Introduction

Behcet’s disease (BD) is a multisystemic vasculitic disorder of unknown etiology. It is characterized by oral and genital ulcers, though the inflammatory perivasculitis can arise in almost any tissue. Neurological complications occur in about 5 to 25% of all patients with BD which accounts for long term morbidity and mortality. Here we present a case of BD with a solely neurological presentation.

Case Report

A nine-year-old girl presented for the first time with double vision and right ptosis. She had no preceding symptoms of vertigo, ataxia, dysphagia, fever or history of trauma. Neurological examination revealed a right third nerve palsy. MRI was inconclusive and her symptoms resolved with steroids. Four days later she had recurrence of symptoms with additional ataxia and drowsiness. MRI now showed a hyper-intense lesion in the right midbrain. Investigations for young stroke were performed which were normal. On enquiry, she mentioned that she suffered from infrequent oral ulcers. Ten years later she developed slurred speech and gait ataxia. A general examination documented an erythematous macular rash involving both her lower legs and a shallow ulcer anteriorly on her right leg (Figure 1). Neurological examination confirmed gait ataxia. MRI revealed a new lesion in the left half of the midbrain (Figure 2). Yet again her symptoms responded

Fig. 1: Shallow cutaneous ulcer and erythematous rash

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recurrence of the speech slurring along with left hemiparesis, headache and diplopia. Neurological examination confirmed left hemiparesis with grade 4 power and mild dysarthria. The rest of the Neuro-exam including the sensory system was normal.

MRI showed a new hyper-intense lesion on the right side of the midbrain. Inflammatory markers, Autoimmune panel and CSF study were normal. Skin biopsy was suggestive of vasculitis. P-ANCA and C-ANCA done following the biopsy results were within the reference ranges. At this stage, based on her clinical and radiological findings and by excluding other causes of stroke, the diagnosis of Neuro-Bechet’s disease. Also, since there is no specific laboratory diagnostic test, the diagnosis of NBD depends essentially on clinical findings, radiological findings and after careful exclusion of other possible diseases. The differentials to be considered are mainly autoimmune and demyelinating illnesses. MS being one such condition can be readily differentiated from NBD on MRI. The MRI image of our patient was compatible with the characteristics of NBD. Brainstem-thalamic-basal ganglia lesions in the proper clinical context can strongly support the diagnosis of acute/subacute parenchymal NBD, and on occasions can raise this possibility even when the systemic features of BD are scarce.

Of the systemic vasculitidies the common culprits affecting the brain include granulomatosis with polyangiitis (Wegners), Polyarteritis Nodosa and Behcets disease. Vasculitis secondary to SLE and RA must also be considered. Both pANCA and cANCA along with an autoimmune screen are helpful in ruling out these other causes when suspecting NBD. ESR has not been proven to be helpful in diagnosing or monitoring NBD disease activity. This was validated in our patient who had normal ESR and CRP.

The treatment of parenchymal NBD primarily consists of glucocorticoids (high-dose pulse intravenous and/or oral) and azathioprine. Because of its relatively predictable and low side effect profile, azathioprine is commonly used as a first-line disease modifying treatment in many centers for the serious manifestations of BD, particularly NBD.

References