CASE OF THE MONTH

Neuro-Behcet’s Disease Presenting as a Young Stroke

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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 multi-systemic 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 diagnostically challenging presentation.

Case Report

A 20-year-old female presented to the clinic with acute onset slurred speech, heaviness of the left upper and lower limb, headache and diplopia. There was no history of vertigo, ataxia, dysphagia, fever or trauma. Neurological examination showed left hemiparesis with grade 4 power and mild dysarthria.

On enquiring about her past, she stated that 11 years ago she was admitted for complaints of double vision and eyelid droop. She was diagnosed to have a right third nerve palsy. Initial MRI was normal and with the suspicion of an autoimmune/demyelinating etiology, she was given an empiric dose of intravenous Methyl-Prednisone to which she responded. Four days later the diplopia recurred along with loss of balance and she became drowsy. A repeat MRI now showed a diffuse lesion in the right midbrain. She was worked up for potential causes of young stroke which were all within normal limits and she was started on Aspirin. Her symptoms resolved with another dose of steroids. A detailed history revealed that she had infrequent mouth ulcers. The patient had no subsequent complaints until ten years later when she developed slurred speech and ataxia. A clinical examination documented an erythematous macular rash involving both her lower legs and a shallow ulcer anteriorly on her right leg (Figure 1). Neurological examination showed gait ataxia. MRI revealed a new lesion in the left half of the midbrain (Figure 2). Her symptoms resolved after a dose of steroids. During this admission, a skin biopsy was also performed which came back normal.

Her current MRI showed a new hyper-intense lesion on the right side of the midbrain. Her ESR, CRP, haemogram, homocysteine, protein C and S, Sickle cell, RA factor, LE cell and Lupus Anticoagulant were all normal. A CSF study was within normal limits and her...
echocardiogram was also normal. A skin biopsy was suggestive of vasculitis (Figure 3). Both P-ANCA and C-ANCA done following the biopsy results were within the reference ranges and an ANA and dsDNA were normal as well. At this stage, based on her clinical and radiological findings and by excluding other causes of stroke and vasculitis; she was labelled as a case of Neuro-Bechet’s disease.

Subsequently she is in remission maintained on azathioprine.

Discussion

Behcet’s disease is a rare immune mediated small vessel systemic vasculitis that is characterized by the clinical triad of aphthous ulcers, genital lesions and ocular involvement.¹

The nervous system involvement is one of the most serious manifestations of BD, leading to headache, confusion, paresthesia, cranial nerve palsy, cerebellar ataxia, or meningeal irritation signs.² Of these headache is the most common neurological symptom accounting for about 70% of patients.³ In this case the patient did present for the first time when she was nine years old but diagnosis in children is difficult because all specific diagnostic clinical manifestations may not be present at the same time and long intervals may be needed before the appearance of characteristic clinical features to make a definite diagnosis.⁴ There are two categories of neurological involvement in BD that have been generally accepted: parenchymal involvement and non-parenchymal involvement, also called cerebral Angio-Bechet’s syndrome. 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 considered in our patient were SLE, MS and systemic vasculitis. The differentiation of MRI findings in NBD from those found in MS are based on the predilection for different regions of the brain. The most commonly affected regions of NBD are the mesodiencephalic area (46%), the pontobulbar region (40%), and the hypothalamic-thalamic region (23%).⁵ On the other hand, the lesions seen in MS affect predominantly the periventricular grey area and corpus callosum.⁶ 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.⁷

Of the systemic vasculitidies the common culprits affecting the brain include granulomatosis with polyangiitis (Wegners), Polyarteritis Nodosa and Behcet’s 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. Although raised ESR and other serum inflammatory markers have been found to be associated with disease activity in BD,⁸ no definite identifiable pattern has been recognized to be linked with NBD activity. This was further validated by 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 disease modifying treatment in many centers for the serious manifestations of BD, particularly NBD.⁹

References