e-ISSN: 0976-822X, p-ISSN:2961-6042

Available online on http://www.ijcpr.com/

International Journal of Current Pharmaceutical Review and Research 2025; 17(9); 923-926

Case Report

Challenging Case of Isolated Neuro-Behcet Disease in an Adolescent, From Headache to Diagnosis: A Case Report

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Received: 01-06-2025 Revised: 15-07-2025 / Accepted: 21-08-2025

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Conflict of interest: Nil

Abstract

Background: Behcet disease is an immune mediated disease with multisystem involvement. The disease may have ophthalmological, neurological as well as cardiovascular system involvement along with mucocutaneous symptoms. Isolated neuro-behcet disease is a rare disease which is difficult to diagnose, as there are no specific diagnostic criteria of the disease, it poses a big challenge in confirming the diagnosis and management. The genetic factor HLA-B51/52 needed to support the diagnosis is positive only in 60 to 70% cases and negative in 30 to 40% cases. The pathergy test is also nonspecific but increases the chances of diagnosis.

Case Presentation: A 15-year-old male adolescent brought to our emergency unit with recurrent episode of headache, vomiting, diplopia and recent onset of similar complaint associated with horizontal and vertical gaze nystagmus, unilateral facial nerve palsy, dysarthria and ataxia. There is history of headache with aura for 7 to 8 months for which patient was treated as a case of migraine. Also history of unilateral left cervical granulomatous lymphadenitis 5 years back. The patient was diagnosed as a probable case of isolated Neuro-Behcet disease by MRI brain & spinal cord and MR angiography along with CSF and skin finding.

Conclusion: In the current study the role of MRI and MR angiography along with skin findings and CSF analysis were discussed in diagnosis of probable parenchymal Neuro-Behcet disease in the absence of other marker of disease. The strong differential diagnosis of Neurosarcoidosis and primary CNS vasculitis was kept.

Keywords: Neuro-Behcet disease, headache, Neurosarcoidosis.

Introduction

Behcet disease is a chronic multisystem immune mediated disease of unknown etiology. The disease is characterized by recurrent aphthous ulcers, genital ulcer and uveitis [1]. Skin manifestations in the form of acneiform nodules and erythema nodosum can occur in 75% of the patients [1]. The first description is believed to be from Hippocrates in fifth century BC. The first modern description was given by Turkish dermatologist Hulusi Behcet in 1937 [2].

The highest prevalence of the Behcet disease is seen in Turkey and Japan and the severity is increased in the Mediterranean and the middle east (i.e., silk route) [2]. The disease has equal male and female ratio and tend to have more severe course and morbidity in males. The etiology of the disease is unknown but certain factors such as inflammatory mediators, genetics, immunologic phenomena, infectious agents and clotting factors are likely factors to contribute [3]. The disease is sporadic, family clustering is also seen. Behcet disease is closely associated with HLA-B51 gene, it is found that the disease is more severe and 6 times more common in patients who are HLA-B51 positive. Pathergy test is commonly performed in Behcet disease, it refers to eruption of papulo-pustules at the site of the needle injury within 24 to 48 hours. Pathergy is not specific to Behcet disease and may be found in other disease such as pyoderma gangrenosum and sweet syndrome [4]. The neurological involvement was first discovered by Knapp in 1941[5].

Isolated neuro-Behcet disease is difficult to diagnose, there is no diagnostic criteria for isolated neuro-Behcet disease [5]. The diagnosis is usually made by the MRI/MRA, CSF analysis and clinical

findings of the patient [6]. Isolated neuro-Behcet disease is extremely rare and poses a great challenge to make the diagnosis. In this report, a 15-year-old male adolescent, has been diagnosed with probable isolated parenchymal type neuro-Behcet disease, is presented to emphasize the role MRI and MRA along with CSF analysis and skin manifestations in childhood.

Case Report

A 15-year-old male adolescent presented to our emergency unit with complaint of headache, projectile vomiting, diplopia, vertical gaze nystagmus and ataxia. The patient had history of headache associated with photophobia and phonophobia since past 7 to 8 months for which he was treated at multiple hospitals as migraine, sometime responding to analgesic, rarely awakening from sleep at night, during that period MRI brain was also done which was normal. Now the patient presented with sudden onset of severe headache associated with diplopia and blurring of vision during headache, horizontal and vertical gaze nystagmus, projectile vomiting and ataxia. On examination he was found to have left facial nerve UMN type palsy, an emergency NCCT head followed by MRI BRAIN and MR ANGIOGRAPHY was done and CSF analysis was performed. CSF was suggestive of lymphocytic pleocytosis and mildly elevated proteins. In laboratory investigations CBC, Electrolytes, vit-B12, vit-D, ESR, CRP, ANA, C3, C4, anti-dsDNA and TFT were normal. Nonurinary abnormalities. Ophthalmology evaluation was unremarkable. Secondary infectious etiology was ruled out. Brain MRI showed T2/FLAIR hyperintensities in midbrain, pons, medulla, bilateral middle cerebral peduncle and white matter of cerebellum and superiorly seen extending along corticospinal tracts involving posterior limb of bilateral internal capsule, bilateral corona radiata, right capsulo-ganglionic region, right external capsule, optic chiasma and radiation with no diffusion restriction and no post contrast enhancement. Bilateral P1 segment of PCA, bilateral A1 segment of ACA shows multiple constrictions on MR angiography giving beaded appearance with multiple foci of microhemorrhages seen in midbrain, pons, bilateral capsulo-ganglionic region, right frontoparietal lobe likely vasculitis induced. MRI spine was normal.

On the basis of radiological and clinical grounds we kept the possibility of Isolated Neuro-Behcet disease [6] [3]. However, there is no history of oral and genital ulcers, no history of blurring of vision (except during headache), joint compromise and lung compromise and no history of hypertension. Although the patient has acneiform nodules on abdomen and past history of unilateral cervical lymphadenopathy 5 years back.

In his family history there was no parental consanguinity, birth difficulty, hypertension and headache. Protein excretion was normal in 24 hour urine, Uveitis was not observed in eye examination and Pathergy test was negative. HLA B51/52 allele was sent and found negative. To rule out other vessel involvement a CT angiography of thorax and abdomen was done which was grossly normal. Due to the migrainous character of headache and MRI findings, after ruling out secondary causes, we strongly kept the possibility of isolated neuro-behcet disease (parenchymal type) [6].

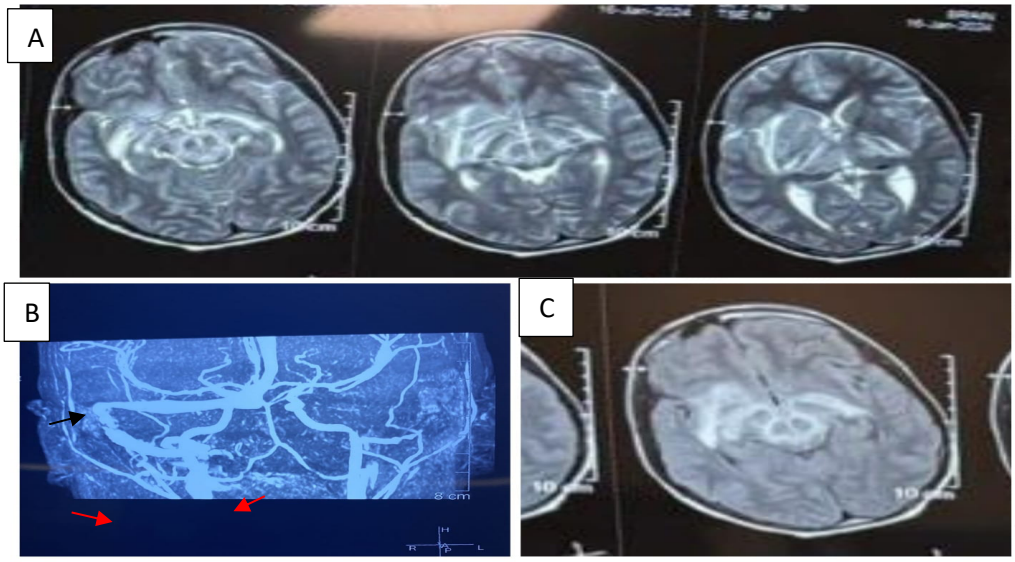

Figure 1: a and c denotes T2 WI/FLAIR showing hyperintensity in upper part of midbrain, posterior limb of bilateral internal capsule and thalami; b denotes Time-Of-Flight MRA showing Beaded appearance of P1 part of right PCA (marked with black arrow) and Beaded appearance of bilateral A1 part of ACA (marked with red arrow).

Discussion

Isolated neuro-Behcet disease is a rare disease. The disease has no definitive diagnostic criteria which poses a great challenge to the physicians to reach the diagnosis. The characteristic feature of the disease is recurrent oro-genital ulcers, uveitis and skin manifestations. The disease rarely present in an isolated neurologic form in which the first presentation is neurologic system involvement.

The most common symptom is migrainous headache which may be throbbing and tension type. The headache occasionally responds to analgesics and rarely leads to night time awakening from sleep.

In our case the patient presented with complaint of headache associated with photophobia and phonophobia. The patient received treatment for migraine for 7 to 8 months and detailed work up for headache including MRI brain was normal at initial presentation. The patient also had history of right sided unilateral cervical lymphadenopathy 5 years before the onset of neurological symptoms.

There were no evidence of hypertension and or any evidence of infectious etiology for the disease. Later he presented with aggravated manifestations of headache and MRI with MRA revealed characteristic lesion of the Neuro-Behcet disease [4]. The characteristic MRI finding of neuro-Behcet disease are lesions extending from upper brainstem to the thalamus and basal ganglia, iso-hypointense in T1A, hyper-intense in T2A and may have a mild mass effect and contrast enhancement [7]. In our case, the area of hyperintense was seen in T2A and FLAIR series, starting from midbrain, pons, medulla, bilateral middle cerebral peduncle and white matter of cerebellum and superiorly seen extending along corticospinal tracts involving posterior limb of bilateral internal capsule, bilateral corona radiata, right capsule-ganglionic region, right external capsule, optic chiasma and radiation. No diffusion restriction and no post contrast enhancement was seen.

In children mostly vascular involvement is seen in most of countries including Israel, France and Saudi Arabia. Our case had parenchymal involvement. There is no specific CSF finding in isolated neuro-Behcet disease nor a diagnostic criteria. There may be pleocytosis and elevated proteins according to some studies. In our patient CSF was suggestive of Lymphocytic pleocytosis and mildly elevated proteins due to parenchymal involvement which is compatible with literature [5]. HLA-B51/52 allele positivity is strongly associated with Behcet disease, however it may be negative in 30% to 40% of cases and may not found positive in all cases.

Those who are HLA-B51/52 were found to have more severe disease and is a marker of bad prognosis and poor outcome, compared to those who are HLA-B51/52 allele negative [5]. In our patient, HLA-B51/52 allele were negative, after confirming the disease on clinic-radiological grounds treatment was started with pulse therapy of methylprednisolone 1000mg iv for 5 days followed by oral prednisolone @1mg/kg/day. As the patient is considered to have CNS vasculitis with first possibility of neuro-behcet disease (parenchymal type), patient was started on mycophenolate mofetil.

After initiation of management the patient showed significant improvement in neurological deficit. Started walk without support within one week of initiation of therapy and motor verbal response become normal gradually. The patient kept on close monitoring and follow up and it was planned to repeat MRI after 45 days for resolution of lesions. As the definitive confirmatory test is brain biopsy, we strongly kept the other possibilities of primary CNS vasculitis and neuro-sarcoidosis.

Conclusion

In the current study the role of MRI and MR angiography along with skin findings and CSF analysis were discussed in diagnosis of probable parenchymal neuro-behcet disease in the absence of other marker of disease. The strong differential diagnosis of Neurosarcoidosis and primary CNS vasculitis was kept.

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