Case Report

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Neurobehcet mimicking multiple sclerosis

Abstract

Report on a case of female patient diagnosed with behcet disease over 10years duration, presented with neurological deficit in form of acute left sided weakness which after proper investigations (MRI findings) proved to be a case of neurobechet for which the patient received treatment with complete resolution of her condition.

The main challenge in our case is the importance of differentiation between MS and neurobechet which can be achieved by MRI brain which is one of most useful tools in assessment of NB based on distribution of the lesion and its characteristic enhancement.

The differentiation between these two conditions is crucial for proceeding to the management. So, the aim of our case presentation is to raise the awareness in importance of differentiation between Neurobehcet and MS.

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Introduction

Behcet disease is a multisystem vasculitic disease that can present with various systemic manifestations. Neuro-Behçet's disease is one of the more serious manifestations of Behçet's disease, it can be classified as parenchymal or non parenchymal (vascular) neurobehcet according to site of affection, which is an inflammatory multisystem disease,[1] closely resemble to MS demyelinating lesion.

So the differentiation between this two conditions is crucial for proceeding to the management. So, the aim of our case presentation is to raise the awareness in importance of differentiation between Neurobehcet and MS.

Case report

A 31year old female patient with no special habits of medical importance was diagnosed with behcet disease based on recurrent oro-genital ulcers and recurrent anterior uveitis for which steroids, hydrochloroquine and one dose of infleximab were prescribed. 1month before admission to our department, the patient developed acute onset left side weakness and dysarthria with no urine or stool incontinence, the condition was not associated with fever, headache or trauma.

On examination the patient was fully conscious, oriented with GCS 15, neurological examination showed left side weakness proximal more than distal with power -3 with hypertonia and hyperreflexia with no signs suggest cranial nerve affection and other systems review were apparently free.

Investigations showed normal CBC with elevated ESR (1st hour 110mm/hr) and CRP (60mg/l) with normal liver and kidney function. CT brain was normal. So MRI was mandatory and showed multiple scattered cerebral enhancing lesions suggesting evidence of MS versus neuro-behcet with cerebral activity.

CSF analysis was normal with no monoclonal antibodies, visual evoked potential was normal, those were done to exclude demyelinating disease like MS. In context of being bechet, neurobechet like MS lesion should be considered.

Accordingly, the patient received treatment in form of IV pulse steroids for 5days followed by 6 cycles of cyclophosphamide therapy with complete resolution of her condition (normal power).

Discussion

Behcet is a multisystem chronic relapsing disorder of unknown etiology that can present with variable clinical manifestations.[2] It was first described in 1937 by the Turkish dermatologist Hulusi Behçet as a triple-symptom complex consisting of recurrent uveitis, aphtous ulceration of the mouth and genitalia.[3] Most of the characteristic lesions of Behçet's disease are due to vasculitis which is obligatory for the diagnosis in populations at risk.[4]

In 1990, the International Study Group (ISG) criteria for diagnosis of BD were developed and still remain the most widely used and well-accepted criteria among experts[5] (Figure 1).

Sign/symptom	Score
Oral aphthosis	2
Genital aphthosis	2
Ocular manifestations	2
Skin manifestations	1
Vascular manifestations	1
Central nervous system involvement	1
Positive pathergy test	1

Point score system: scoring ≥4 indicates BD diagnosis. Pathergy testing is optional and the primary scoring system does not include pathergy testing. However, where pathergy testing is conducted, one extra point may be assigned for a positive result.

Figure 1 Revised ICBD.

In 2006, The International Criteria for Behçet's Disease (ICBD) which is a new diagnostic criterion were developed. These criteria

expanded the 5 items of the ISG criteria to 6, including vascular manifestations, and assigned a weighted point value system. Later in 2010, these criteria were themselves revised, neurologic signs were added and the pathergy test was considered an extra criterion to be used if conducted and positive, given its high specificity[6,7] (Figure 2).

Recurrent oral ulceration (obligatory)	minor aphthous, major aphthous, or herpetiform ulceration (observed by physician or patient); recurring at least 3 times in one 12-month period
Plus 2 of	
Recurrent genital ulceration	aphthous ulceration or scarring (observed by physician or patient)
Eye lesions	anterior uveitis, posterior uveitis, or cells in vitreous on slit lamp examination; or retinal vasculitis observed by ophthalmologist
Skin lesions	erythema nodosum (observed by physician or patient), pseudofolliculitis, or papulopustular lesions; or acneiform nodules (observed by physician) in post-adolescent patients not on corticosteroid treatment
Positive pathergy test	read by physician at 24–48 h

Findings applicable only in absence of other clinical explanations.

Figure 2 ISG criteria.

Neurological involvement in behcet disease, what´s called neurobehcet (NBS) is well recognized nowadays with a variable degree of prelevance in different socities.[8]

The prevalence of neurological involvement in BD has been reported in a wide range (2–44%) from different geographical regions,[9,10] two major studies from Istanbul region showed a prevalence of around 5%,[11,12] The majority of these cases presented with parenchymal CNS involvement.[13] Which were a meningoencephalitis involving mainly the brainstem (BS), diencephalic structures and basal ganglia.

Pallis et al.,[14] and Wadia et al.,[15] was first described a neurological involvement in Behçet's syndrome in the early decades of this century.[14,15] NBS have multiple facies of presentations, which may be an acute focal or multifocal CNS dysfunction; it commonly resembles multiple sclerosis (MS).[16]

It may be misdiagnosed as multiple sclerosis such as our lady case so the differentiation between the two conditions is a very important entity. There are different methods to differentiate between two conditions as Visual evoked potentials, however still it is difficult to differentiate.[17]

In our case the MRI was one of most benefit tool to differentiate between the two conditions as there periventricular distribution together with picture of enhancement lesions and venous enhancement was characteristic of vasculitis as shown in Figure 3 so in the context of bechet disease, the diagnosis of neurobehcet with cerebral activity was confirmed.

The reason for the difference between NB and MS is that pathogenesis in NB points to non-specific inflammations such as neuro-infections in contrary to multiple sclerosis which considered as autoimmune disorders, suggesting that a currently unknown infection might be the trigger of a vasculitic process in the central nervous system (CNS).[18]

Neurological involvement in BD is more commonly due to involvement of central nervous system (CNS), spinal affection can occur in NB but is less prevalence than CNS affection.[19,20]

There were several prior studies that have noticed the MS like neurobechet such as A. Ozturk who described a 21years old female patient with Behcet's disease which started as Neuro-behcet's disease: Mimicking multiple sclerosis attack, and concluded that NB disease must be one of differential diagnoses in the presence of MS like lesions even though other manifestations of BD are not obvious.[21] And, Hadfield MG , Aydin F , Lippman HR , Sanders KM who described a case of NB without vasculitis who presented with intermittent neurological symptoms which were not responding to treatment and there neuroradiological findings were closely resemble MS.[22]

Also, A prospective study of neurologic manifestations of Behcet's disease in 96 Iranian patients: Neuro-Behcet's disease: A masquerader of multiple sclerosis.[23] Concluded that neurological manifestations in NB is relatively rare however more serious and should be included in differential diagnosis of MS.

The main challenge in our case is the importance of differentiation between MS and neurobechet which can be achieved by MRI brain which is one of most useful tools in assessment of NB[24] based on distribution of the lesion and its characteristic enhancement. Infliximab is tried in some patients with relapsing NB with good results.[25]

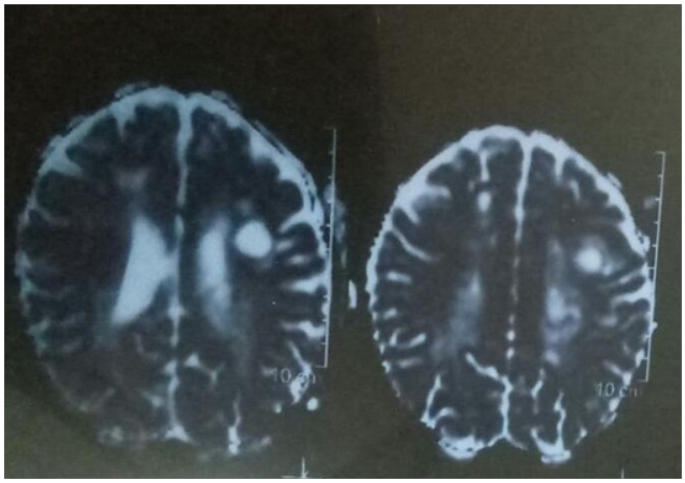

Figure 3 MRI brain.

Conclusion

Behcet disease is a multisystem vasculitic disease that can be presented with variable neurological involvement called neurobehcet which should be differentiated from MS mainly and other neurological diseases.

We presented a case of behcet disease presented with neurological deficit resemble MS, and diagnosed as neurobehcet disease based on characteristic MRI finding, received treatment with pulse steroids and cyclophosphamide injections with complete improvement of her condition.

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Competing interests

The authors declare no potential competing interests, including financial interests, activities, relationships, and affiliations.

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Conflict of interest

The author declares that there is no conflict of interest.

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