Neuro-Behcet Disease – A Case Report and Review

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Abstract

Behcet’s disease (BD) is a multisystem autoimmune relapsing vasculitis with almost unknown etiology, which involves both large and small vessels. In Neuro-Behcet’s disease (NBD), the CNS can be involved generally in two ways. The parenchymal type is caused by development of an immunemediated meningoencephalitis, which predominantly involves the brainstem and diencephalon region. Non-parenchymal type is a consequence of thrombosis within the dural venous sinuses. Peripheral nervous system generally preserved or involved in very rare cases.

The diagnosis of NBD is based on the clinical presentation and the typical NBD lesions in brain magnetic resonance imaging (MRI). The development and disappearance of lesions at MRI correlate with the course of clinical neurologic deficits. Brain MRI in chronic NBD usually shows brain or brainstem atrophy and in some cases “black holes”.

We present a case report of a female with NBD and a thoral review of this disease.

Keywords: Behcet Disease • Neuro-Behcet’s disease • Clinical Presentation • Radiological findings • Neuroimaging

Introduction

Behcet’s disease (BD) is a disease with autoimmune etiopathogenesis characterized by recurrent genital and oral ulceration and uveitis. The pathogenesis is still uncertain, but the leading pathological finding is generalized vasculitis of all blood vessels - arteries and veins of any diameter [1]. This disease may present with other symptoms including involvement of central nervous system (CNS), inflammation and pain in the joints, gastrointestinal complaints, thrombophlebitis and thrombosis of the deep veins, arterial occlusion or aneurysms [2].

The neurological involvement called Neuro-Behcet’s Disease (NBD) is rare and remains a difficult diagnosis to establish as other diseases and conditions may have similar presentation [3]. It is crucial to early diagnose NBD and treat it properly because it is one of the main causes of long-term disability and mortality in BD [4]. We present a case report of a female diagnosed with NBD and review on clinical presentation, classification and neuroimaging pattern of NBD.

Case Report

We describe a case of a 25-year old female hospitalized in November 2019 in Second Clinic of Neurology of UMHAT “St. Marina”, Varna with sudden onset complaints of general weakness, which was more prevalent for the right limbs, slurred speech and swallowing disorders. Two days prior she complained of pain when trying to move her lower limbs and during swallowing.

Review of her medical history revealed Behcet’s Disease diagnosed in December 2018. She also suffered from secondary adrenal insufficiency and had two episodes of aseptic meningitis in June 2017 and March 2019.

On admission she was in poor general condition with severe dysarthria. She was in non-febrile state with regular pulse at 108 beats per minute, blood pressure 135/92 mm/Hg.

On neurological examination, the patient was conscious, with limited command response due to severe dysarthria. There were no signs of meningoradiculal irritation. The pupils were equal, with slow reaction to light, unaffected eye movements. There was central lesion of the right facial nerve, dysphonia and dysarthria. Motor activity examination revealed quadriparesis, the pattern being moderate to severe for the right limbs and mild on the left. Tendon reflexes were asymmetrical and diminished for the left side. Babinski’s sign was negative bilaterally. The neurological findings were noted to be consistent with clinical presentation of a vertebrobasilar cerebral infarction.

Initial laboratory tests showed elevated leucocyte count – 14,78 × 10^9, high C-reactive protein (CRP) - 62,14 mg/l, and increased cholesterol levels – 7,50 mmol/l as the LDL was 5,54 mmol/l. Renal and hepatic enzymes were normal.

Computed tomography (CT) scan of the head without contrast showed no pathological findings.

Because of the neurological symptomatic and the anamnesis for Behcet’s Disease a decision of performing MRI was made. The neuroimaging showed multifocal pathological signal intensity–hyper-intense on both T1 and T2 sequence. Due to the previous hospitalizations for meningitis, the patients had previous brain MRIs, and we were able to compare the dynamic of the neuroimaging findings. Compared to the previous MRI from March 2018 (8 months ago) development of new and disappearance of previously known lesions was noted (Figures 1 and 2).
A: Axial T1 – Hyperintense lesion at the left mesencephalon extending to crus cerebri and the posterior limb of capsula interna with mild mass-effect mimicking a mesencephalic tumor.
B: Axial T1 – Hyperintense lesions with different shape at right basal ganglia and thalamus; bilateral periventricular lesions and insular cortex and right inferior frontal gyrus.
C: Sagittal T1 – Hyperintense lesion at the left mesencephalon extending to crus cerebri and the posterior limb of capsula interna with mild mass-effect mimicking a mesencephalic tumor.
D: Axial T2 – Hyperintense lesions at right basal ganglia and thalamus; bilateral periventricular lesions.

Figure 1. MRI imaging from March 2019.

A: Axial T1 – Hyperintense lesion at the left mesencephalon extending to crus cerebri and the posterior limb of capsula interna, with reduced volume and reduced mass-effect in comparison with previous images.
B: Axial T1 – Newly formed hyperintense lesion in right basal ganglia with edema and mass-effect.
C: Coronal T1 – Newly formed hyperintense lesion in right basal ganglia with edema and mass-effect; new smaller lesions in left thalamus and in the right cortical area.
D: Axial T2 – Newly formed hyperintense lesion in right basal ganglia with edema and mass-effect.

Figure 2. MRI imaging from November 2019 – Definite diagnosis of Parenchymal Neuro-Behcet Disease with migration (development and disappearance) of lesions.
The largest new lesion was 18mm in diameter with perifocal edema and mass-effect in the periventricular basal ganglia on the right side (nucleus caudatus, putamen, globus pallidus and the posterior limb of capsula interna). Another new lesion 3.5mm was found in the left thalamus. Reduced in size previously known lesions were visible in the left mesencephalon with mild mass-effect and in the right pontine area near the cerebellar limb. Disappearance of some of the known lesions was noticed - near the right lateral ventricle and in the right inferior frontal gyrus and the left insular cortex.

In this case the patient had anamnesis for previously proven BD. The clinical presentation and the comparison of previous and the new images showed the typical changes with migrating lesions relevant to the clinical course. The fulfilled criteria for BD, the neurological syndrome supported by characteristic abnormalities on the MRI gave us the reason to accept diagnosis of Parenchymal Neuro-Behcet Disease.

After multidisciplinary discussion and considering the clinical findings, the patient underwent a treatment with Colchicine, Methylprednisolon and symptomatic therapy for the mucocutaneous lesions and the pain syndrome. The patient showed partial clinical improvement. At the discharge she was with mild quadriparesis and possible independent walking.

### Clinical Review

#### Clinical presentation and classification

Behcet's disease (BD) is a chronic, relapsing, systemic disorder of unknown etiology in which an inflammatory perivascularitis can affect the vessels in all tissues. The clinical presentation is characterized by recurrent oral and genital ulcers, uveitis, and other manifestations from multiple organ systems [6]. The diagnosis of BD is made on the basis of the combination of typical clinical symptoms and signs.

The diagnostic criteria for BD are presented in Table 1 [6].

In 2014, the International Team for the Revision of the International Criteria for BD proposed new classification based on the analysis of more than 2500 BD patients. This criterion is based on point system where ocular lesions, oral and genital aphthae are each assigned 2 points, whereas lesions of the skin, CNS involvement, and vascular symptoms are assigned 1 point each. The pathergy test, when used, was assigned 1 point. The score ≥4 points represents diagnosis of BD [7].

### Discussion

Neuro-Behcet's disease (NBD) is one of the most serious manifestations of (BD). Though NBD is relatively rare - occurs in 5% to 10% of patients, but being potentially treatable, neurologists need to consider it in the differential diagnosis of infective, inflammatory or demyelinating central nervous system (CNS) disorders [8]. It is fundamental to early recognize NBD and treat is properly because it is one of the main causes of long-term disability and mortality in BD [9].

NBD is defined by the Consensus Status Agreement as the neurological predominant symptoms of a patient who has clear diagnosis of BD and suffers from all of the other systemic symptoms [8].

The diagnostic criteria for NBD are presented in Table 2.

The neurological symptoms can be manifested by damage of both central and peripheral nervous system (PNS) [8]. The CNS is the usual target of NBD and there are two main categories of CNS involvement – parenchymal (P-NBD) and non-parenchymal (NP-NBD) involvement [10].

P-NBD accounts for the majority of NBD – about 80% of the cases [11]. This form is featured by diffuse brain stem, cerebral, optic, and spinal cord symptoms. Different neurological symptomatic can appear depending on the site of involvement [12]. P-NBD commonly presents with an attack of hemiparesis, cognitive disturbances, pelvic-reservoir troubles and fever. Recent study in South Korea reports the frequency of these manifestations - pyramidal signs (52.0%), headache (45.9%), dysarthria (42.9%), and fever (31.6%) [13]. In summary the most common clinical findings are pyramidal tract and brain stem signs. The symptoms may have sudden onset – acute attack. The last one may be followed by a relapsing or progressive course of the disease [14].

Because of the heterogeneous clinical presentation and progression, there is a further division of P-NBD into an acute type and a chronic-progressive type [15]. The first type is presented by acute and transient symptoms such as fever and hemiparesis accompanied by inflammatory features from the blood samples and in the cerebrospinal fluid (CSF). The chronic type is characterized by slowly progressive over time ataxia, dementia, incontinence, and brainstem atrophy [11].

NP-NBD accounts for 20% of the cases and usually affects major intracranial vessels with frequent involvement of the venous sinuses, cerebral veins and less commonly the arteries [12]. In very rare cases thrombosis and aneurysms of the large cerebral arteries are described [18]. The most frequent vascular manifestation is venous sinus thrombosis, followed by thrombosis of the deep and the cortical cerebral veins [17]. An association between venous parenchymal infarcts and venous thrombosis can be observed, but it depends on the collateral circulation. The extensive intra- and extracranial collateral circulation usually leads to good compensation in the early stages [18]. NP-NBD usually manifests with sudden onset of neurological symptomatic. The arterial involvement commonly is presented with stroke with progression of the neurological deficit over time [19]. Increased intracranial pressure is the leading clinical manifestation of venous sinus thrombosis. The clinical picture headache, papilledema, focal neurological symptomatic, seizures and coma [20].

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**Table 1. International Study Group (ISG) criteria for the diagnosis of Behcet's disease (BD)**

| International Study Group (ISG) criteria for the diagnosis of Behcet's disease (BD) |
|---|
| **1. For diagnosis, patient must have had the following symptoms:** |
| - Recurrent oral ulceration—minor or major aphthous, or herpetiform ulceration observed by physician or patient that recurred at least three times in a 12-month period |
| **1. Plus two of the following:** |
| - Recurrent genital ulceration – aphthae or scarring, observed by physician or patient |
| - Eye lesions—anteriour 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, papulo-pustular lesions; or acniform nodules observed by physician in post-adolescent patients not on corticosteroids |
| - Positive pathergy test—read by physician at 24–48 h |

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The major classification and forms of NBD are presented in Table 3.

Neuroimaging in Neuro-Behcet Disease

The diagnosis of NBD is challenging and relies on the exclusion of other neurological conditions with similar clinical picture such as CNS infections, brain tumors, demyelinating disorders and neurological involvement due to immunosuppressive therapies. There are some additional criteria, which are based on neuroimaging (MRI in particular, but also MR angiography and venography) serum profiles, HLA determination, neurophysiological examination, and eventually, nervous tissue biopsy [21].

Magnetic Resonance Imaging (MRI) is crucial for the diagnosis and is the gold standard for neuroimaging examination to prove NBD. There are different typical findings in parenchymal and non-parenchymal NBD [8].

In patients with P-NBD there are focal or multifocal CNS abnormalities in the clinically affected areas. The lesions are located in the brainstem, thalamus and basal ganglia. Rarely, they can affect the cerebral hemispheres, cerebellum, and the spinal cord [14].

Typical MRI findings are small foci of high signal intensity on T2-weighted images, which are iso- or hypointense compared to the normal brain parenchyma on T1-weighted images. Besides in some cases T1 lesions can be hyper-intense [22]. Lesions may be with different shape - circular, linear or irregular. The migration - development and disappearance of lesions on MRI correlates with the course of clinical neurologic deficits and is typical feature of NBD [23]. NBD lesions may diminish in size in response to steroid and immunosuppressive treatment [24,25].

Parenchymal NBD lesions have predilection to the brainstem-thalamic-basal ganglia region. The meso-diencephalic junction is typically affected as this is commonly seen during acute attacks [12]. The second most common location is the ponto-bulbar region as the pontine base is most affected and occasionally the lesion extends to brachium pontis. The white matter is more often involved, but lesions also might be seen within the grey matter structures, including brainstem nuclei [25]. In one-third of the cases the basal ganglia and internal capsule region is affected [14]. Diffusion-weighted imaging may help in the cases, presenting with stroke-like episodes, revealing an increase in the diffusion coefficient in NBD lesions. The clinical similarity between successive attacks may also be helpful in diagnosing NBD [8].

Atypical presentation of P-NBD is seen as a space-occupying lesion mimicking a unilateral brain tumor. In such situations the diagnosis is difficult, especially if there is no exacerbation on the systemic Behcet's disease signs. Steroid administration may be a diagnostic option in uncertain clinical cases [26].

In patients with NP-NBD the most common presentation is venous thrombosis. In more rare cases there might be an arterial aneurism, arterial occlusions or dissection and aseptic meningitis [2].

The most common MRI findings in NP-NBD are occlusion of the cerebral venous sinuses without or with venous infarcts [8], MRI in conjunction with magnetic resonance venography (MRV) is highly sensitive in detecting such lesions [19]. Neuroimaging features are similar to those in patients with central venous thrombosis due to other causes. The most common occlusion locations are sinus sagittalis superior, sinus transversus, deep cerebral veins, and the cavernous sinuses [27]. Magnetic resonance angiography (MRA) shows direct and indirect signs for thrombosis. Direct signs include lack of typical high-flow signal, and the appearance of flow signal after recanalization. Indirect signs are formation of collateral vessels, an occasional prominent flow signal from deeper medullary veins, cerebral hemorrhage, visualization of emissary veins, and signs of increased intracranial pressure [22].

Differential diagnosis of Neuro-Behcet's Disease

The differential diagnosis (DD) of NBD remains a major challenge. Neuro-Behcet Disease have wide specter of differential diagnoses including demyelinating process, neuroinfection, systemic vasculitis, brain tumors and cerebrovascular disease [28].

### Table 2. International consensus recommendation (ICR) criteria for NBD diagnosis [8].

| International Consensus Recommendation Criteria for the diagnosis of Neuro Behcet’s disease (NBD) |
|-----------------------------------------------|
| **I.** Definite diagnosis of NBD - must be meeting all of the following three criteria |
| 1. Satisfy the ISG criteria for BD (presented in Table 1) |
| 2. Neurological syndrome (with objective neurological signs) recognized to be caused by BD and supported by characteristic abnormalities seen on either or both: Neuroimaging and CSF |
| 3. No better explanation for the neurological findings |
| **II.** Probable diagnosis of NBD – must be meeting one of the following two criteria in the absence of a better explanation for the neurological findings: |
| 1. Characteristic neurological syndrome as in definite NBD, with systemic BD features but not satisfying the ISG criteria |
| 2. A non-characteristic neurological syndrome occurring in the context of systemic BD satisfying the ISG criteria |

### Table 3. Consensus classification of neuro-Behcet’s disease [8].

| Consensus classification of Neuro-Behcet’s disease | Peripheral nervous system |
|-------------------------------------------------|--------------------------|
| Parenchymal (P-NBD)                              | Peripheral neuropathy    |
| • Multifocal or diffuse                          |                          |
| • Brainstem                                      | Mono-neuritis multiplex   |
| • Spinal cord                                    | Myopathy                 |
| • Cerebral                                       | Myositis                 |
| • Asymptomatic (silent)                          |                          |
| • Optic neuropathy                               | Acute meningeal syndrome |
| Non-Parenchymal (NP-NBD)                         |                          |
| • Cerebral venous thrombosis                     |                          |
| • Intracranial hypertension                      |                          |
| • Intracranial aneurysm                          |                          |
| • Cervical extracranial Aneurysm/dissection      |                          |

Mixed form of Parenchymal and Non-parenchymal NBD
First, NBD should be primarily differentiated from MS. The age of onset of the two diseases is about the same (20–40 years), but MS is generally more common in women, whereas NBD is seen frequently in men [29]. Some symptoms are present in both the conditions, but the frequency of their presentation varies. Optic neuritis, sensory symptoms, cerebellar symptoms such as dysarthria or ataxia, and spinal cord involvement are common in MS and are quite rare in NBD. On the other hand, headache, pseudobulbar speech, and cognitive–behavioral changes are more common in NBD [30]. Lesions of the brainstem that commonly extend to the basal ganglia and diencephalic structures support the diagnosis of NBD, whereas MS lesions preferentially involve periventricular areas and the corpus callosum [21]. The migration of the lesions - development and disappearance of lesions are typical feature for NBD [23].

The second DD is bacterial meningitis. It is essential as it is condition of medical emergency. Clinical presentation of headache, an altered mental status, seizures and focal cerebral signs can be common manifestations of both the infectious condition and NBD [8]. Anamnesis for toxic-infectious syndrome and continuous monitoring of the patient’s temperature might help, as the fever is usually high in bacterial meningitis, while temperature may be normal or of moderate degree in NBD. The CSF analysis is very useful in distinguishing the two conditions. In infective encephalitis there is decrease of the CSF glucose levels, proteinorrachia (protein levels above 0,45g/l usually between 1-5g/l) pleocitosis (up to 1000–10000 white blood cells/mm³). Specific blood tests may be performed to establish the identity of specific pathogens [21].

The third DD of NBD is the autoimmune disease causing primary or secondary CNS vasculitis. Patients with primary vasculitis do not have systemic signs, which are always present in NBD [31].

Atypical NBD space-occupying lesions are mimicking primary or secondary brain tumors or even abscesses. A stereotactic biopsy would show perivascular infiltration of leukocytes and microglia, oligodendroglial degeneration and areas of necrosis in NBD. Steroid administration may be a diagnostic option in uncertain clinical cases as they can shrink in size the lesion [26]. In the context of Behcet's disease the typical clinical picture is more important that a brain biopsy [32].

Finally, acute P-NBD can simulate acute stroke-like manifestation. Patients with BD may also exhibit a higher risk of vascular stroke, the etiology of which is uncertain [31]. Diffusion-weighted MRI is useful in differential diagnosis of both the infectious condition and NBD [8].

**Conclusion**

In differential diagnosis of NBD should be considered cerebrovascular diseases, brain tumors, and demyelinating processes. The MRI findings of NBD are distinct and they can prove the diagnosis even in cases without typical systemic manifestations. Stereotyped localization and migration over time of the lesions within the brain parenchyma are characteristic of NBD. Vascular NBD should be considered in cases with venous sinus thrombosis.

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How to cite this article: Tsalta-Mladenov, Mihael, Darina Georgieva-Hristova and Silva Andonova. “Neuro-Behcet Disease – A Case Report and Review.” J Neurol Disord 8 (2020): 425 doi: 10.37421/J Neurol Disord.2020.8.425