IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 23, Issue 9 Ser. 5 (September. 2024), PP 39-41
www.iosrjournals.org

Intracranial Hypertension Revealing Neuro-Behçet Disease With Favourable Outcome After Ventriculoperitoneal Shunt

H.Majdaoui, S.Najim, J.Boucht, I.Elkhateb , R.Jennane, R.Belfkih
Neurology Départements Of Mohamed VI Teaching Hospital Of Tangier

Abstract:
A 39-year-old man had been diagnosed after rolling out all the other possible causes as having intracranial hypertension with bilateral papilledema secondary to Behçet's disease with normal MRI scan. Despite treatment with steroids, methylprednisolone and repetitive LPs, he had little to no improvement. The patient later underwent a ventriculoperitoneal shunt operation with corticosteroid therapy and adjuvant treatment with topiramate and acetazolamide, which resulted in symptoms improvement. Thus, it is important to consider NBD in the differential diagnosis of patients with intracranial hypertension and bilateral papilledema and also ventriculoperitoneal shunting as an effective therapeutic alternative when medical treatment fail to normalize the intracranial hypertension building up.

Keywords: *Behçet's disease, neurological involvement, intracranial pressure, CSF, ventriculoperitoneal shunt*

Date of Submission: 05-09-2024 Date of Acceptance: 15-09-2024

I. Introduction

Defined as a vasculitis with venous tropism, Behçet's disease (BD) is a chronic, relapsing and systemic disease due to its multiple visceral sites, including neurological involvement **[1]**.

According to the International Study Group's classification, a definitive diagnosis of BD requires recurrent oral ulcerations plus two of the following: recurrent genital ulcerations, skin lesions, eye lesions, and a positive pathergy test **[2].** The diagnostic criteria for neuro-Behcet's disease (NBD) are a current diagnosis of BD plus the presence of neurological symptoms not otherwise explained by known systemic or neurological diseases **[3]**. NBD has variable prevalence depending on the series but can represent around 10-25% of the affected patients in a large series **[4]**. Neurological manifestations may appear as a parenchymal central nervous system pattern (the commonest), an intracranial hypertension-like pattern, or a meningitis-like pattern **[5]**.

We discuss through this case report an unusual initial presentation of NBD and that is intracranial hypertension that's been shown in larger series have a rate of approximately 5% **[6],** with favourable outcome after ventriculoperitoneal shunt.

II. Case Report:

A 39-year-old man, with no medical history or medication use, admitted for subacute headaches with vomiting, especially in the mornings, for one month, associated with blurry vision in both eyes. He was afebrile and fully oriented, with no neck stiffness.

A cranial, orbital MR imaging (MRI), and intracranial venous MR angiography came back negatives (figure 1). Opening pressure measurement during lumbar puncture (LP) revealed an intracranial pressure of 42 cmH2O when he was lying on side, with no microscopic findings. The IgG index was normal and oligoclonal band was negative in cerebral spinal fluid (CSF).

Fundoscopic examination revealed stage III papilledema bilaterally, and the visual field was disturbed (figure 2). Complete blood count, electrolytes, liver function tests, thyroid function tests, urea and creatinine, cortisol, iron, vitamin A, B 12 and D were all within the normal limits. Antinuclear antibody titers, anti-doubled-stranded DNA antibodies were also normal.

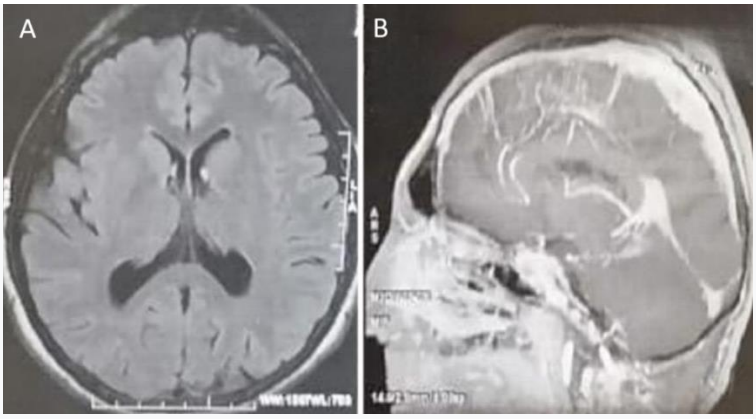

Figure 1: Normal neuroimaging
A: MRI showing non parenchymal involvement
B : Angiogram demonstrating good flow through the cerebral veins and venous sinuses.

While retaking the patient's history, it revealed several episodes of recurrent oral and genital ulcerations, and the pathergy test came back normal. BD with intracranial hypertension was suspected. We later administered Methylprednisolone 1 g IV for 5 days, followed by prednisolone 60 mg p.o with dosage's decreasing within three months period. Depletive LP was also performed without any improvement, with persistent stage III papilledema.

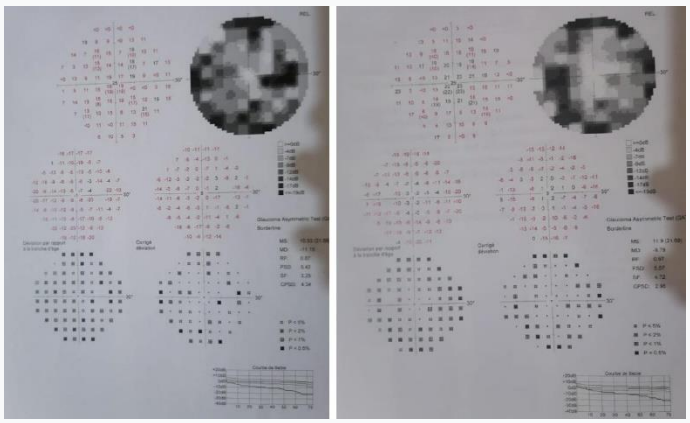

Figure 2: Humphery visual field demonstrates a remarkable visual field loss

The patient later underwent ventriculoperitoneal shunting and was placed on colchicine with corticosteroid therapy and adjuvant treatment with topiramate and acetazolamide, that were gradually tapered and stopped over weeks as his symptoms resolved.

III. Discussion:

NBD can be diagnosed if there is central nervous system involvement and the BD diagnostic criteria are satisfied. It occurs more commonly in men, with a male to female ratio of up to 4:1 **[7]**. The mean age of onset was found to be 32.0±8.7 years **[8]**. Papilledema and headache are common manifestations of intracranial hypertension, which was also in our case, the patient was a 39-year-old man with lumbar CSF pressure of 42 cmH20 and a history of recurrent oral and genital ulcerations.

For the diagnosis of NBD with isolated intracranial hypertension, certain conditions related to cerebral pseudotumor syndrome must be met in accordance with the diagnostic criteria of BD. These conditions include: 1. bilateral papilledema; 2. lumbar CSF pressure >250 mmH2O; 3. no mass lesions or hydrocephalus on brain imaging **[9]**. Our patient's imaging was normal, showing no signs of parenchymal or non-parenchymal abnormalities which helped making the proper diagnosis.

Although intracranial hypertension in patients with known BD has been described, this may be confounded by steroid treatment, which predisposes to this condition. Our patient was not known to have BD or history of taking any steroids.

In 40% of patients the CSF is normal **[10]**, which is similar to our case that had elevated CSF opening pressure. If not treated rapidly, intracranial hypertension with the eventual development of optical atrophy might cause irreversible loss of sight, which necessitates an urgent and quick intervention to prevent it.

Glucocorticoids, immunosuppressants (azathioprine), and dehydrating agents are commonly used in the treatment of NBD with isolated intracranial hypertension **[11]**. In our case, our patient was given steroids, acetazolamide and topiramate with no improvement in the symptoms.

The management of those patients in whom the intracranial hypertension does not respond to treatment with corticosteroids is unclear. Similar to the case presented here, additional cases of BD treated with lumboperitoneal shunt have been described in the literature.

Bank and Weart documented a BD patient with intracranial hypertension secondary to dural sinus thrombosis, despite medical treatments for more than a month and repeated lumbar punctures, the patient's symptoms associated with raised intracranial pressure persisted. A lumboperitoneal shunt resulted in complete resolution of the neurological complaints of the patient without any relapse in symptoms **[12]**.

IV. Conclusion:

BD should be routinely looked for in adult patients, especially males in their third and fourth decades, who present with intracranial hypertension syndrome. Lumbar puncture, after a normal contrast-enhanced CT or MRI scan, is absolutely mandatory to establish that the constituents of CSF are normal in every patient in whom benign intracranial hypertension is a consideration. In BD patients in whom the intracranial hypertension developed but failed to normalize after medical treatment, ventriculoperitoneal shunting should be considered as an effective therapeutic tool which we tried to emphasize through this case as well.

References:

[1] Thierry Chabin. La Maladie De Behcet : A Propos D'un Cas De Neuro-Behcet D'aspect Pseudo-Tumoral. Médecine Humaine Et Pathologie. 1989.

[2] Sevda A.K, Ahmet A, Necati C, Aysu K.A. Neuro-Behcet's Disease In A Case With Papilledema And Intracranial Hypertension As The Initial Presentation. North Clin Istanb, 2018 Apr 17; 5(2): 153–156.

[3] Seema K, Alan S, Gulsen A-D, Saeed B, Afshin B-H, Cris S.C Et Al. Diagnosis And Management Of Neuro-Behçet's Disease: International Consensus Recommendations. J Neurol. 2014; 261(9): 1662–1676.

[4] Akman-Demir G, Serdaroglu P, Tasçi B. Clinical Patterns Of Neurological Involvement In Behçet's Disease: Evaluation Of 200 Patients. The Neuro-Behçet Study Group. Brain. 1999;122:2171–82.

[5] Ascaso F.J, Rodriguez A, Cristóbal J.A. Documenta Ophthalmologica. 2002;105(3), 291–299.

[6] Walker RW. Idiopathic Intracranial Hypertension: Any Light On The Mechanism Of The Raised Pressure? J Neurol Neurosurg Psychiatry. 2001;71: 1-5.

[7] Siva A, Altintas A, Saip S. Behçet's Syndrome And The Nervous System. Curr Opin Neurol. 2004;17:347–57.

[8] Saip S, Akman-Demir G, Siva A. Neuro-Behçet Syndrome. Handb Clin Neurol. 2014;121:1703–23.

[9] Akdal G, Yaman A, Men S, Et Al. Pseudotumor Cerebri Syndrome Without Cerebral Venous Sinus Thrombosis In Behçet's Disease. J Neurol Sci. 2017;383:99–100.

[10] Hirohata S, Kikuchi H, Sawada T, Nagafuchi H, Kuwana M, Takeno M, Et Al. Clinical Characteristics Of Neuro-Behcet's Disease In Japan: A Multicenter Retrospective Analysis. Mod Rheumatol. 2012;22:405–13.

[11] Yali W, Wei Y, Shufang L, Shasha W Et Yan D. Diagnosis And Management Of Neuro-Behçet Disease With Isolated Intracranial Hypertension: A Case Report And Literature Review. BMC Neurology. 2023; 23:335.

[12] Bank I, Weart C. Dural Sinus Thrombosis In Behçet's Disease. Arthritis Rheum. 1984;27:816-8.