Visual impairment due to venous sinus thrombosis in neuro-Behc¸et’s disease

Pérdida visual debido a trombosis de senos venosos en neuro-Behc¸et

Dear Editor,

Behc¸et disease (BD) is a multisystemic inflammatory disease of unknown aetiology characterised by aphthous ulcers in the mouth and genitals, ocular inflammation, skin lesions including erythema nodosum, and acneiform eruptions. Patients may also show neurologic, cardiovascular, and gastrointestinal involvement. Although BD has historically been considered more prevalent in men, recent studies point to a more balanced sex ratio. However, most patients with neurological involvement are men. BD affects young adults; age at onset is a predictive factor of clinical severity. Diagnosis is based on the presence of systemic and ocular clinical manifestations, since the disease has no pathognomonic signs. Ocular involvement in patients with BD usually includes bouts of inflammation occurring in the context of an underlying chronic retinal vascular inflammation. Bouts may affect the anterior pole, in the form of acute uveitis, or the posterior pole, with severe vitritis, retinal haemorrhages and exudates, cystoid macular oedema, or optic neuritis. BD may involve the nervous system in the form of recurrent meningoencephalitis typically affecting the brainstem, idiopathic intracranial hypertension (ICh) with or without sinus thrombosis, cranial mononeuropathies, cerebellar ataxia, myelitis, seizures, and even cognitive impairment. There are 2 clearly defined forms of BD: parenchymal and non-parenchymal. The most frequent manifestation of the latter is venous sinus thrombosis.

We present the exceptional case of a patient diagnosed with neuro-Behc¸et disease (NBD) and rare attacks of ocular inflammation. He displayed bilateral optic atrophy secondary to chronic papilloedema in the context of ICh due to dural venous sinus thrombosis.

Our patient was a 37-year-old white man who was referred to our hospital’s neuro-ophthalmology unit due to progressive and persistent vision loss in both eyes over the previous several months. He had been diagnosed with BD at the age of 18 based on the presence of recurrent mouth ulcers since adolescence, 2 bouts of bilateral anterior uveitis, and facial acneiform eruptions. At the age of 24, he was diagnosed with NBD due to thrombosis of the superior longitudinal, transverse, and sigmoid sinuses (Fig. 1) after consulting for symptoms of headache, intermittent vision loss, and papilloedema. He was treated with oral prednisone, azathioprine, colchicine, ciclosporin, and anti-coagulants. In the years previous to our evaluation, our patient reported fluctuations in vision quality that were attributed to papilloedema secondary to ICh caused by dural venous sinus thrombosis. Two lumbarperitoneal shunts to manage ICh achieved satisfactory results and decreased papilloedema and vision loss fluctuations. Mild papilloedema persisted one year later; doctors suggested a permanent lumbarperitoneal shunt placement but the patient refused surgery. He had no other ophthalmological manifestations or relevant family history. At the time of his visit, he was taking oral acetazolamide, azathioprine, antiplatelet agents, and anticoagulant agents. The ophthalmological examination revealed a visual acuity of 0.1 in both eyes. Biomicroscopy showed pigmented keratic precipitates in both eyes and no active signs of ocular inflammation. Intraocular pressure and intrinsic and extrinsic ocular motility were all within normal limits. Eye fundus examination revealed bilateral papillary pallor and no signs of inflammation. The Humphrey visual

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field test (SITA-Standard 24-2 programme) revealed diffuse absolute scotoma in both eyes. Optical coherence tomography showed decreased thickness of the nerve fibre layer of both optic nerves (Fig. 2). Further analyses including a biochemical study, a total protein test, and measurements of folic acid and vitamins B1, B6, and B12 yielded normal results. An additional lumbar puncture disclosed an intracranial pressure of 15 mm Hg. CSF analysis revealed no abnormalities. Visual evoked potentials revealed no response on either side. An MRI scan showed no additional findings. Our patient was diagnosed with bilateral optic atrophy secondary to chronic papilloedema despite successful management of intracranial pressure.

Optic disc atrophy is an ophthalmological sequela that results in permanent interruption of axoplasmic transport in the optic nerve head. In the context of BD, it is usually unilateral or asymmetrical and occurs after repeated bouts of posterior uveitis (viritis, vasculitis, neuroretinitis, optic neuritis). Early loss of visual acuity and neuroretinal rim pallor are essential clinical signs for distinguishing optic disc atrophy from glaucomatous optic neuropathy. As in our case, NBD may contribute to damaging the optic nerve in the form of either optic neuritis or papilloedema secondary to ICH. In these cases, involvement is usually bilateral and symmetrical. The most frequent cause of ICH in NBD is dural venous sinus thrombosis. Our patient had experienced only 2 episodes of uveitis previously, but ICH had been present for several years. Despite good management of intracranial pressure, ICH had caused papilloedema; although medical and surgical treatment had achieved partial resolution of papilloedema, it persisted over time, leading to atrophy of the optic nerve head.

Prevalence of NBD varies greatly among studies (5% to 49% of the cases). Parenchymal involvement includes a wide
Figure 2  A) Eye fundus. Optical atrophy: optic discs with neuroretinal rim pallor. B) Humphrey visual field test (SITA-Standard 24-2 programme): diffuse absolute scotoma in both eyes. C) Optical coherence tomography (Cirrus): decrease in the nerve fibre layer of both optic nerves.
range of clinical findings compatible with sensory-motor
ictal symptoms. Corticosteroids, immunosuppressants,
colchicine, and biological products seem to provide
good treatment options, although scientific evidence is
insufficient to support them. Non-parenchymal
involvement is frequently associated with signs and symp-
toms of ICH and mainly caused by dural venous sinus
thrombosis. These patients have a better prognosis than
those with parenchymal involvement, which suggests
that these 2 forms possess different etiopathogenic
mechanisms. Recent studies have shown that anticoagulant
treatment is decisive for improving prognosis. Refrac-
tory patients and those with precisely located thrombi may
benefit from mechanical thrombectomy with or without
thrombolytic treatment.

Conclusion

Papilloedema secondary to ICH is a rare finding in patients
with NBD. Prognosis is usually favourable, although the
condition may cause optic atrophy and even vision loss
regardless of the level of previous ocular inflammation.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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References

AT, editors. Diagnosis and treatment of uveitis. Philadelphia:
2. Bertol V, Ara JR, Oliveros A, Gutiérrez AI, Samperiz P, Gros B,
4. Rodríguez-Carballeira M, Alba MA, Solans-Laqué R, Castillo AJ,
Registry of the Spanish network of Behçet’s disease: a descrip-
Suppl. 84:S33—9.
5. Benamour S, Najj T, Alaoui FZ, el-Kabli H, el-Aidouni S. Neu-orological involvement in Behçet’s disease 154 cases from
a cohort of 925 patients and review of the literature. Rev Neurol.
2006;162:1084—90.
6. Tugal-Tutkun I, Onal S. Uveitis in Bechet disease: an analysis
7. Diaz Valle D, Méndez Fernández R, Benitez del Castillo Sánchez
José M. Actualización en el tratamiento de las uveítis. Comuni-
cación solicitada, 83. In: Congreso de la Sociedad Española de
8. Borhani-Haghighi A, Samangooei S, Ashjazadeh N, Nikseresht A,
Shariat A, Yousefpour G, et al. Neurological manifestations of
Study Group. Clinical patterns of neurological involvement in
Behçet’s disease: evaluation of 200 patients. Brain. 1999;122
Pt 1:2171—82.
10. Morrissey SP, Miller DH, Hermaszewski R, Rudge P, MacManus DG,
Kendall B, et al. Magnetic resonance imaging of the central nerv-
E. Neurologic involvement in Behçet’s syndrome: A prospective
C, et al. Biologics, colchicine, corticosteroids, immunosup-
pressants and interferon-alpha for neuro-Beheçet’s syndrome.
13. Saadoun D, Wechsler B, Resche-Rigon M, Trad S, Le Thi Huong D,
14. Siddiqui FM, Dandapat S, Banerjee C, Zuurbier SM, John-
son M, Stam J, et al. Mechanical thrombectomy in cerebral
venous thrombosis: systematic review of 185 cases. Stroke.

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