An 8-year-old Behcet Disease girl initially presenting with bilateral retrobulbar optic neuritis

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Case Report

Keywords: Behçet Disease, Retrobulbar optic neuritis, Ocular involvement

Posted Date: March 11th, 2020

DOI: https://doi.org/10.21203/rs.3.rs-16794/v1

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Abstract

Background: Behçet disease (BD) is a polygenic and chronic autoinflammatory multisystem vasculitis. Acute optic neuritis has been rarely reported in patients with BD, especially in children.

Case presentation: We reported an 8-year-old girl with suddenly visual loss and color vision impairment. The patient has a history of recurrent oral aphthous ulcers, genital ulcers, and chronic abdominal pain. On ophthalmic examination, anterior and posterior chambers and funduscopy of both eyes were normal. The results of laboratory tests for infectious and rheumatic diseases were normal. Brain magnetic resonance imaging and the result of cerebrospinal fluid analysis for oligoclonal bands autoantibodies were normal too. Pathergy skin test and human leukocyte antigen (HLA) B5 and HLA-B51 were positive. The patient was recognized as a case of BD-related bilateral retrobulbar optic neuritis and was treated by corticosteroid, azathioprine, colchicine, and infliximab.

Conclusion: Retrobulbar optic neuritis may be first manifestation of BD.

Background

Behçet Disease (BD) is a chronic disease associated with venous and arterial vasculitis, which represented by recurrent oral aphthous ulcers, genital ulcers, skin lesions, neurologic and ocular involvement, arthritis and gastrointestinal (GI) involvement (1). Ocular involvement, which is seen in 30%-60% of BD cases, is more commonly associated with panuveitis, which is usually bilateral. Iridocyclitis and posterior uveitis are ocular manifestations of BD. Retinal vasculitis, retinal detachment, and optic neuritis also occur rarely (2,3). Retrobulbar optic neuritis is one of the very rare manifestations of retinal involvement in BD cases, especially in children (4). Here we report BD-related bilateral Retrobulbar optic neuritis, which is a very rare case of ocular involvement in children.

Case Report

An eight-year-old girl was admitted in the pediatric neurology ward of Imam Hossein Children's Hospital, Isfahan University of Medical Sciences due to a sudden visual loss in both eyes and headache in the occipital area. The patient had no fever and she did not have any symptoms of meningitis, including neck stiffness, Brudzinski, and Kerning in physical examinations. Cranial nerves examination were normal, visual acuity of the right eye was hand motion of 50 cm, and the left eye was 100 cm. The patient had visual impairment in recognizing green and red colors. On ophthalmic examination, anterior and posterior chambers and funduscopy of both eyes were completely normal. The patient has a history of recurrent oral aphthous ulcers (5–6 times a year), genital ulcers, chronic and vague abdominal pain, diarrhea, and intermittent constipation. She had been treated with intravenous immunoglobulin due to Kawasaki when she was three years old.
The results of laboratory tests including complete cell count, erythrocyte sedimentation rate, C-reactive protein, serum electrolytes, infectious diseases serology for toxoplasmosis, cytomegalovirus, tuberculosis and rheumatologic tests including anti nuclear antibody, angiotensin converting enzyme, antiphospholipid antibodies and serum complement levels were normal. The result of mantoux test was negative. Chest X-ray and Brain magnetic resonance imaging (MRI) results were normal either. The result of cerebrospinal fluid analysis including cell, sugar, protein, smear, culture, oligoclonal bands, Neuromyelitis optica (NMO) antibody, Myelin oligodendrocyte glycoprotein (MOG) were normal. Fecal calprotectin level was 252 µg/mg. (NL < 50). GI endoscopy and colonoscopy results were normal. The patient was transferred to pediatric rheumatology ward. The result of a pathergy skin test, on the forearm area performed with intradermal needle 21-gauge, was positive. Both human leukocyte antigen (HLA) B5 and HLA-B51 tests were positive.

According to the criteria of the International Study Group for Behcet Disease, the patient was recognized as a case of BD-related bilateral retrobulbar optic neuritis and the treatment started by prescribing Pulse Methylprednisolone 30 mg/kg/d (max 1gr) for three days. Then in the fourth day of treatment, intravenous infusion with Infl iximab 8 mg/kg was given. The patient was discharged in the fifth day given oral Prednisolone 2 mg/kg/d, Azathioprine 2 mg/kg/d and Colchicine 1 mg/d. After a one-week follow-up, the patient had no color vision impairment and both eyes could see two meters away. Ocular exam through slit lamp and fundoscopy was normal. She did not have headache and nausea. After a one-month follow-up, the patient had no color vision impairment and ocular exam through slit lamp and fundoscopy was normal too. Both eyes visual acuity was around 20/40. Infusion of Infl iximab 8 mg/kg was given and oral Prednisolone 1 mg/kg/d with Azathioprine and colchicine with similar doses were continued. Three months after discharge, the patient did not have any complain about aphthous ulcers and GI system problems. Visual acuity of both eyes was 20/20 and ocular exam was normal. Azathioprine, colchicine and Infl iximab infusion continued monthly. Oral Prednisolone was tapered to 0.2 mg/kg/d.

**Discussion**

Behçet disease (BD) is a polygenic and chronic autoinflammatory multisystem vasculitis characterized by mucocutaneous, musculoskeletal, neurological, gastrointestinal and ophthalmological lesions. Childhood-onset BD is uncommon, accounting for 3–7% of all cases. Prevalence in children is probably not more than 10% of the adult counterparts in eastern Mediterranean countries.

BD rarely presents with acute optic neuritis. Few publications have reported an association between BD and optic neuropathy. Retrobulbar neuritis (optic neuritis) is a rare manifestation of neuro-Behçet disease. The diagnosis in children is difficult, as the disease is uncommon and clinically resembles other diseases, such as multiple sclerosis. Optic neuritis is a consideration whenever monocular or biocular blindness develops, suddenly in a child. The initial feature in some children is pain in the eye, but for most it is blurred vision, progressing within hours or days to partial or complete blindness. Visual acuity reduces to less than 20/200 in almost all affected children within 1 week. Visual evoked response testing
further confirms the diagnosis. MRI of the orbital may reveal swelling and demyelination of the optic nerve. Examination of the cerebrospinal fluid may be helpful to check for markers of demyelinating disease, such as oligoclonal bands and anti-NMO antibodies. Results of ophthalmoscopic examination may be normal at the onset of symptoms if neuritis is primarily retrobulbar(7,8).

Etiologic factors in cases of retrobulbar optic neuritis may be local, such as inflammation associated with sinusitis, or general, as multiple sclerosis and undulant fever. Acute type are due to virus infection of the central nervous system. Differential diagnosis of retrobulbar optic neuritis is Multiple Sclerosis, neuromyelitis optica, idiopathic optic neuritis, ischemic optic neuritis (is rare in children). Usually occurs as a sudden segmental loss of vision in one eye, but slow or stepwise progression over several days. Color vision loss is roughly equivalent in severity to visual acuity loss, whereas in optic neuritis the disturbance of color vision is greater than of visual acuity. Ophthalmoscopic examination reveals diffuse or partial swelling of the optic disk. It gives the appearance of papilledema and flame-shaped hemorrhage appear adjacent to the disc margin.), traumatic optic neuropathies, toxic optic neuropathies and psychogenic blindness(9,10).

The severity of visual loss and degree of response to treatment can vary but patients usually benefit from steroid treatment. Another recommended treat for inflammatory eye manifestation of BD is Azathioprine and for resistant eye manifestation Anti-TNF and Alpha Interferon considered. TNF-α, are known to be elevated in active BD, suggesting that anti-TNF-α therapy might be effective. Clinically, significant improvement of various(8,11).

**Conclusion**

Although acute optic neuritis has been rarely reported in patients with Behcet disease (BD), our experience suggests that retrobulbar optic neuritis may be first manifestation of BD, that patient's are coming with suddenly visual loss. Retrobulbar optic neuritis can be bilateral, can affected both eyes simultaneously, can be recurrent, and can lead to severe visual loss.

**Declarations**

**Ethics approval and consent to participate**

We confirm that the written informed consent form has been provided by the parents to have the case details published. Also, we restate that institutional approval is not required to publish the case details.

**Consent for publication**

We agreed

**Availability of data and material**

Not applicable
Competing interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Funding

Not applicable

Authors' contributions

Jari M: Design and final writing
Mohammadi T AND Taheri En: Writing primary manuscript

Acknowledgements

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for profit sectors.

Abbreviations

BD: Behçet Disease, GI: gastrointestinal, HLA: human leukocyte antigen,
MOG: myelin oligodendrocyte glycoprotein, MRI: magnetic resonance imaging, NMO: neuromyelitis optica, TNF: tumor necrotizing factor

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