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Case Report

Evaluation of Clinical Features and Imaging of Idiopathic Optic Perineuritis with just Conjunctivitis Symptoms: A Case Report

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Abstract

Optic perineuritis (OPN) is a rare orbital inflammatory disease that mainly involves the optic nerve sheath and surrounding tissues. Here, we report a 66 - year - old woman who was admitted to hospital with a history of pain, redness and swelling in her right eye for two months. On the ophthalmologic examination, best corrected visual acuity (BCVA) was 1/20 and 20/20 in the right and left eye respectively. With visual impairment, there was colour vision defect in the right eye. In addition to relative afferent pupillary defect (RAPD) positivity, there was also a manifest restriction of outward movement of the right eye. According to the clinical features and imaging results, idiopathic OPN was diagnosed in the right eye. The patient gave dramatic response to corticosteroid therapy. It is important that being sceptical not to misdiagnose the OPN patients applying with just conjonctivitis complaints like pain, redness and swelling.

Keywords: Optic Perineuritis; Orbital Inflammation; Corticosteroids; Optic Neuritis; Doughnut Sign; Tram-track Sign

Abbreviations

OPN: Optic Perineuritis; BCVA: Best Corrected Visual Acuity; RAPD: Relative Afferent Pupillary Defect; OD: Optic Disc; RNFL: Retinal Nerve Fiber Layer; OCT: Optic Coherence Tomography; MRI: Magnetic Resonance Imaging; ON: Optic Neuritis.

Introduction

Optic perineuritis (OPN) was first described by Edmunds and Lawford in 1883 [1,2]. It is a rare orbital inflammatory disease that mainly involves the optic nerve sheath and surrounding tissues. In fact, OPN is an uncommon variant of orbital pseudotumor and especially may be indistinguishable from retrobulbar optic neuritis [3]. It usually has idiopathic etiology [1-4]. However, OPN can be seen with variable inflammatory, vasculitic, infectious diseases like Sarcoidosis, Wegener granulomatosis, Giant cell arthritis, Leukemia, Neurosyphilis, after Influenza vaccination and toxic causes also have been reported [2,5-8]. While the visual prognosis of OPN is generally good, sometimes can be worse like lately treated patients and patients with optic atrophy after inflammation in the optic nerve sheath.

[1,4] Here, we report a patient who had just conjonctivitis symptoms like pain, redness and swelling in her right eye for two months in addition to didn't have any complaint of visual disturbance.

Case Report

A 66 - year - old woman was admitted to hospital with a history of pain, redness and swelling in her right eye for two months. She stated that she had been treated for her red eye, but her complaints were still continuing. The patient's history and systemic examination was unremarkable. On the ophthalmologic examination, best corrected visual acuity (BCVA) was 1/20 and 20/20 in the right and left eye respectively. With visual impairment, there was colour vision defect in the right eye. In addition to relative afferent pupillary defect (RAPD) positivity, there was also a manifest restriction of outward movement of the right eye. Chemosis, conjunctival hyperemia and mild eyelid edema were observed during the anterior segment examination of the right eye. On fundus examination of the right eye, the optic disc (OD) appeared to be pale and elevated with a obscure border (Figure 1a), while the left eye fundus examination was normal (Figure 1b).

Figure 1:

a. The optic disc appeared to be pale and elevated with a obscure border in the right eye.
b. Fundus appeared normal in left eye.
c. Decreased optic disc edema at 2 weeks of treatment.
d. Fundus appeared normal in left eye.

Intraocular pressure was 16 mmHg and 14 mmHg in the right and left eye respectively. The Hertel exophthalmometer measurement was 19 mm in the right eye and 17 mm in the left eye. As visual field evaluation with Humphrey 30-2 automated perimetry revealed blind spot enlargement in the right eye (Figure 2a), there was no defect in the left eye (Figure 2b).

Figure 2:

a. Blind spot enlargement in the right eye.
b. Normal visual field in left eye.
c. Decreased visual field defect at 2 weeks of treatment.
d. Normal visual field in left eye at 2 weeks of treatment.

The right eye mean retinal nerve fiber layer (RNFL) thickness was 291 µm as the left eye was 112 µm in the optic coherence tomography (OCT) examination (Figure 3a).

Figure 3:

a. RNFL thickness in first visit; 291 microns on the right, 112 microns on the left.
b. At the second week of treatment, the RNFL thickness is 149 microns on the right and 112 microns on the left.

Fundus fluorescein angiography showed fluorescein leakage on the optic disc in the right eye (Figure 4a-c) as the left eye appeared to be normal (Figure 4d).

In the laboratory tests; complete blood count, routine biochemical tests, urinalysis, erythrocyte sedimentation rate (ESR), C - reactive protein (CRP), angiotensin – converting enzyme (ACE), thyroid function tests (TFT), thyroperoxidase antibodies (Anti – TPO), anti - thyroglobulin, thyroid stimulating immunoglobulin (TSI) and thyroid beta receptor antibody (TRB) were within normal range. Anti - nuclear antibody (ANA), rheumatoid factor (RF), anti - neutrophil cytoplasmic antibody (ANCA) and other rheumatological autoantibody tests were negative. Syphilis (VDRL-RPR, TPHA), Epstein-Barr virus (EBV), Herpes Simplex Virus (HSV) Type 1 - 2, Varicella zoster virus (VZV), Cytomegalovirus (CMV), toxoplasmosis, measles, rubella, Hepatitis B Virus (HBV), Hepatitis C Virus (HCV) and Human Immunodeficiency Virus (HIV) tests were negative. Chest X - ray and Waters radiograph were both seen to be normal. No pathology was observed as a result of the patient's consultation with otorhinolaryngology, dental diseases and endocrinology departments. However, the brain and orbita Magnetic Resonance Imaging (MRI) showed the "tram - track" sign in T1 - weighted postcontrast fat - suppressed axial sections and the "doughnut" sign also with enhancement of extraocular muscles in coronal sections in the right eye (Figure 5a and 5b).

According to the clinical features and imaging results, idiopathic OPN was diagnosed in the right eye. High - dose intravenous methylprednisolone (1 gr/day) was initiated for 3 days followed by oral deflazacort treatment (90 mg/day). At the first week of treatment, visual acuity dramatically increased to 20/20 and also the patient's colour vision improved. The restriction of ocular movements, chemosis were decreased. At the second week the optic disc edema was observed to be highly regressed on the fundus examination (Figure 1c). The visual field loss decreased in the right eye (Figure 2c and 2d) and the mean RNFL thickness declined to 149 µm (Figure 3b). The treatment of the patient was gradually reduced and terminated in about 3 months.

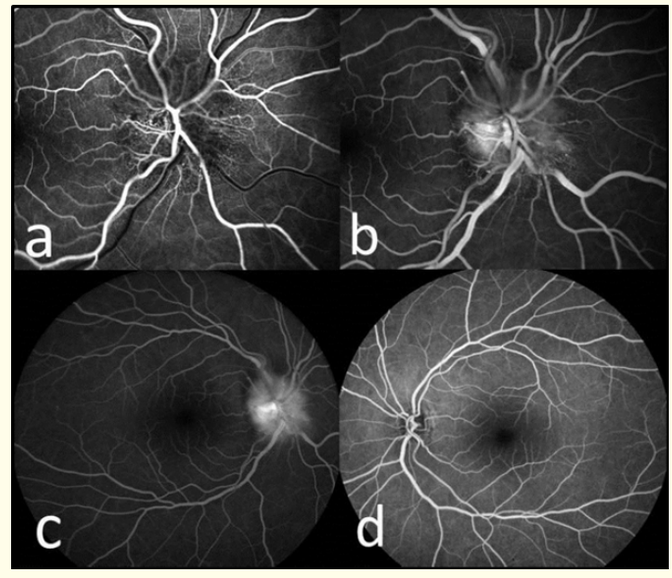

Figure 4:

a. Capillary plexus on optic disc was more visible in the arterio-venous phase in the right eye.
b. Venous phase fluorescein leakage in the right eye.
c. Late stage increased fluorescein leakage in the right eye.
d. Late stage normal angiography in left eye.

Figure 5:

a. Sign of "tram - track" in T1 - weighted postcontrast fat - suppressed axial sections in the right eye.
b. Sign of "doughnut" (arrow) and enhancement of extraocular muscles (stars) in T1 - weighted postcontrast fat - suppressed axial sections in the right eye.

Discussion

Optic perineuritis is an uncommon variant of the idiopathic orbital inflammatory diseases or orbital pseudotumor that also include periscleritis, orbital myositis, and dacryoadenitis [1]. It is mainly the inflammation of the optic nerve sheath and sometimes periorbital fat tissue. The lymphoid infiltrate on the effected tissues consists of lymphocytes, plasma cells, macrophages and polymorphonuclear cells [3]. The pathological studies commonly show that thickening of the optic nerve sheath due to the nonspecific fibrosis. Also some other studies show that necrobiotic

collagen or granulomatous inflammation in the sheath and optic nerve demyelination, infarction or optic nerve vasculitis in patients who have visual loss [1]. A wide spectrum of clinical features have been reported related to etiology and spread of the inflammation along the orbit or optic nerve. In addition to the external inflammation findings like eye lid swelling, conjonctival chemosis, scleritis; proptosis, restriction of eye movements, relatif afferent pupillary defect may be seen with a variety of symptoms like eye pain, visual loss and visual field defects [1-3,5,7,8]. Clinically, the OPN is commonly misdiagnosed as optic neuritis (ON) [1,3,7]. In fact, the external inflammation signs are the only clues in the differantial diagnosis of OPN with ON. In our case also conjunctival hyperemia, chemosis and mild eyelid edema were seen as external inflammation signs that was not commonly encountered in the literature. According to spread of inflammation, compartment effect due to the compression of thickened optic nerve sheath [4] and optic nerve involvement; optic disc edema, visual loss, visual field defects and optic atrophy in lately treated patients may be seen. In this case, the patient didn't emphasize a visual disturbance that may be related to her age and cognition level and this situation complicated the diagnosis at first. But complete ophthalmologic examination that includes visual level evaluation revealed the main entity. Optic perineuritis is usually diagnosed by characteristic clinical features and MRI findings [1,3]. Thickening around the optic nerve in patients with OPN is typically being monitored as "tram-track" sign on axial sections and 'doughnut" sign in coronal sections in MRI [1,3].

Optic perineuritis patients have been reported to give dramatic responses to corticosteroid therapy. However, the recurrence is observed when the treatment regimen is reduced or interrupted [1]. It has been reported that the visual prognosis is poor in some patients when the treatment is delayed or the optic nerve atrophy that initiated by various mechanisms, is occured as a result of recurrent attacks of the disease [1,4]. Differential diagnosis of idiopathic OPN and the idiopathic ON is usually made by MRI findings and clinical features. Characteristic differences of idiopathic OPN from idiopathic ON are, occurance particularly in elderly patients, visual field defects that are often in the form of paracentral or arcuate scotomas, slow onset of symptoms, external inflammation, extraocular muscle involvement and dramatic response to oral steroid therapy that contraindicated in ON patients [1,3]. Optic atrophy that may be related to retrograde axonal degeneration, delayed treatment and recurrent attacks worsen the prognosis [1,4].

In conclusion**,** OPN patients may apply to hospital with various complaints and findings like diplopia due to extraocular muscle involvement, painful eye movements, ptosis, chemosis and conjunctival hyperemia. As a result, the OPN may be complicated with many other clinical pathologies and the diagnosis and treatment may be delayed. The visual prognosis is poor in lately diagnosed and treated patients. It is important that being sceptical and having all possible patients through complete basic eye examinations.

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Declaration of Interest

The authors report no conflicts of interest or funding/support. The authors alone are responsible for the content and writing of the paper. The authors have informed consent and all permissions granted by patient.

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