

CASE REPORT

ATYPICAL AND MISDIAGNOSED CASE OF OPTIC NEURITIS IN A YOUNG FEMALE

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HOW TO CITE THIS ARTICLE:

Inderjit Kaur, Prempal Kaur, Mona Chatrath, Pratibha, Shaurya Sharma. "Atypical and Misdiagnosed Case of Optic Neuritis in a Young Female". Journal of Evolution of Medical and Dental Sciences 2014; Vol. 3, Issue 09, March 3; Page: 2115-2120, DOI: 10.14260/jemds/2014/2112

ABSTRACT: AIM: To report an atypical and misdiagnosed case of optic neuritis in a young female.

METHOD: 26 year healthy female was referred to the Regional Institute of Ophthalmology, Amritsar with the diagnosis of angle closure glaucoma both eyes. Patient was on 2% pilocarpine eye drops q.i.d and tablet acetazolamide 250 mg BD. On examination, visual acuity was 2/60 OD and 1/60 OS. Near vision was 6/36 OU. There was no circumcorneal congestion OU, no corneal edema OU, though anterior chamber was shallow OS >OD with pin pointed non-reacting pupil OU. IOP 14.6mm Hg OD and 17.3mm Hg OS. Gonioscopically open angle grade 2. IOP and gonioscopy were repeated after pupil dilatation with tropicamide, findings were consistent with the previous ones. Fundoscopy showed bilateral disc edema with hyperemia and normal foveal reflex. Color vision was defective. Low vision prevented a conclusive perimetry. ONTT protocol was given, on the basis of decreased visual acuity, abnormal color vision, headache, disc changes, open angle and a normal MRI. Patient's vision improved to 6/6 OD 6/9 OS in thirteen days. **CONCLUSIONS:** Bilateral optic neuritis is rare and an atypical presentation can be misdiagnosed as angle closure glaucoma in presence of severe headache, OU visual loss and shallow anterior chamber.

KEYWORDS: Misdiagnosed, bilateral, optic neuritis, young female.

INTRODUCTION: Optic neuritis, or primary inflammation of the optic nerve, is referred to as papillitis when the optic disc is swollen, retrobulbar neuritis when the disc appears normal and neuroretinitis when inflammation of optic nerve is associated with retinal inflammation. The most common form of optic neuritis is acute demyelinating optic neuritis. The annual incidence of optic neuritis, as estimated in population-based studies, is approximately 3–5 per 100,000 per year.

The majority of patients who develop optic neuritis are between the ages of 20 and 50 years. Women are affected more commonly than men. In most cases, the pathogenesis of optic neuritis is inflammatory demyelination, whether or not MS is diagnosed clinically. Sub-acute or acute visual loss, periocular pain or pain on eye movements and unilateral involvement are typical findings.

CASE REPORT: A primary care center referred a 26 year old female with the diagnosis of angle closure glaucoma to the Regional Institute of Ophthalmology Amritsar with the following notes: reduced visual acuity of <6/60 OU, shallow anterior chamber both eyes. Treatment prescribed by the referring doctor was 2% Pilocarpine eye drops q.i.d and tablet Acetazolamide 250mg BD.

Patient complaint of sudden visual loss 5 days back. This episode occurred in the evening and was accompanied by severe headache. H/o further deterioration of vision over a period of five days
On general examination

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- Patient was conscious and co-operative but apprehensive.
- Vitals –Blood pressure 120/80 mm of Hg, Pulse 74/min, afebrile.
- Non-diabetic and Non-hypertensive
- VA 2/60 OD and 1/60 OS
- No improvement with pinhole
- Color vision testing by Ishihara chart: Impaired color vision

O/E

- Interpalpebral fissure was normal.
- Lids normal.
- No superficial or deep conjunctival congestion
- Cornea clear
- A.C shallow both eyes, OS > OD
- Pupil miotic, non-reactive OU. RAPD could not be elicited.
- IOP 14.6 mm Hg OD and 17.3 mm Hg OS with Schiottz Tonometry
- Gonioscopy: Grade 2 angle width (Shaffer's Grading)
- Post dilation IOP and Gonioscopy: 17.3 mm Hg and 18.9 OU and grade 2 angle width.

Fundus examination OU

- Blurring of disc margins
- Hyperemic disc
- C:D 0.3:1
- Normal vasculature
- Normal foveolar reflex

IV Methylprednisolone therapy 1 gram IV for 3 consecutive days was given. Antiglaucoma medication was stopped. Patient was advised investigations and sent for medical and otorhinolaryngologist opinion.

On 3rd day

- VA improved to 6/36 OD and 6/18 OS
- IOP 14.6 mm of Hg OD and 17.3 mm of Hg OS

Investigations

ESR - 7mm/hr.
 TLC -7000/mm³
 FBS- 126 mg/dl,
 X-RAY Chest- normal
 X-RAY Paranasal sinuses- normal
 MRI- Showed normal axial study of brain.
 Mantoux test - Negative
 HIV - Non-reactive
 CRP- 4mg/dl

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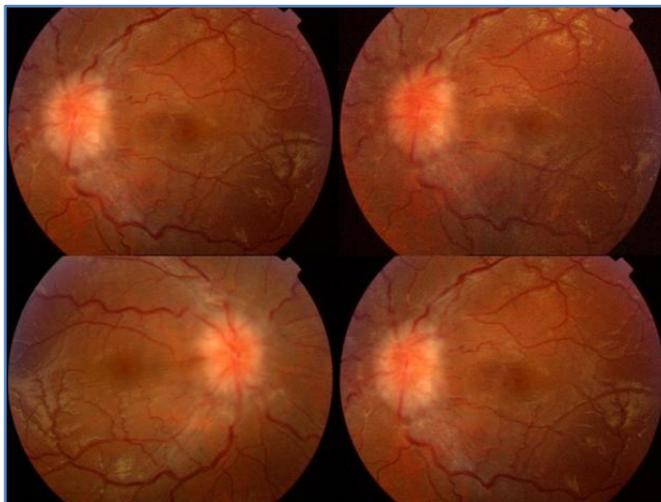
Medical specialist did not find any sensory or motor deficit. ENT specialist also found no relevant problem.

Patient was discharged on Tablet Prednisolone 1mg/Kg body wt. X11 days

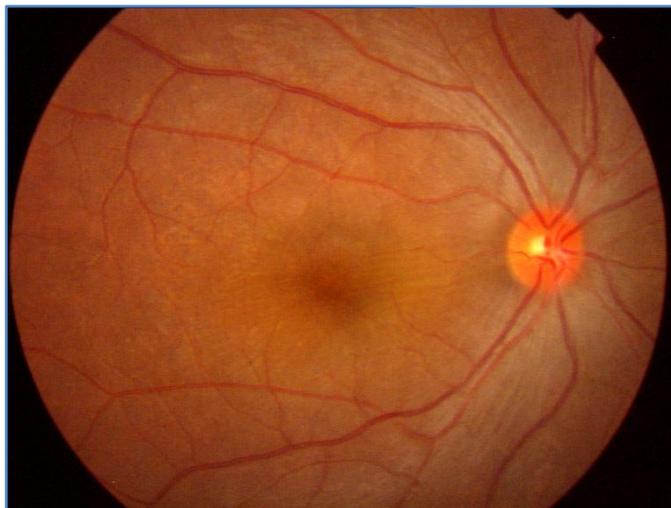
On 13th day

- VA 6/6 OD and 6/9 OS

FUNDUS PICTURE:



Fundus picture on presentation OU



Fundus picture on 13th day of treatment OD

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DISCUSSION: Optic neuritis (ON) is the most common cause of acute unilateral visual loss in young adults¹. Patients typically present with the triad of sub-acute unilateral visual loss, periocular pain and impaired color vision. The symptoms usually worsen over the course of a few days to 2 weeks but spontaneously recover in >90% of cases after 2–3 weeks regardless of steroid treatment. In most patients a relative afferent pupillary defect (RAPD) is obvious, although this is not specific to ON but is present in most optic neuropathies². An atypical clinical presentation, e.g. missing pain, a marked visual loss, bilateral visual loss or absence of spontaneous recovery after 2–3 weeks should always give reason to careful diagnostic search for other differential diagnoses.

In most individuals ON is caused by idiopathic inflammatory demyelination. This may occur as an isolated syndrome or in association with multiple sclerosis (MS). Approximately 50% of patients with isolated ON develop definite MS within 15 years³. The most predictive factor for the development of MS after ON is the presence of asymptomatic demyelinating lesions in the central nervous system (CNS). In the Optic Neuritis Treatment Trial (ONTT) the 5-year risk for definite MS was 52% in those patients with one or more asymptomatic white matter lesions on brain MRI compared with a 5-year risk of only 16% in patients with normal brain MRI.

Nonetheless, inflammatory ON may also be caused by other autoimmune diseases such as sarcoidosis, systemic lupus erythematosus (SLE), Sjögren's syndrome (SS) or Behçet's disease⁴. Although infectious causes of ON such as herpes Zoster, Lyme disease, syphilis, tuberculosis or toxoplasmosis are rare, they should be kept in mind because a specific treatment regime is required rapidly. Many other conditions such as tumors or ischemic diseases can also cause an optic neuropathy and clinically mimic idiopathic inflammatory ON.

In those cases in particular, in which the patient history reveals atypical clinical aspects for ON, a careful clinical work up is required in order to establish the correct treatment regimen. Furthermore, in those patients with ON with a high risk of developing MS, an early and accurate diagnostic assessment to rule out other causes is necessary to initiate a disease-modifying treatment⁵. We should go for MRI, CSF, additional blood tests if possible biopsy to reach specific diagnosis and management. Recommended blood tests are CRP, CBC, Blood Chemistry, Vitamin B12, Glucose, ANA, Serology for Lyme, Brucellosis these are considered to be mandatory. Additional blood tests are RF, ACE, Anti dsDNA, HIV, Serology for syphilis, tuberculosis.

Typical ON	Atypical ON
Young Adult Patient <50 years	Age >50 or < 12 years
Acute or Sub-acute visual loss	Sudden visual loss
Progressive over a few days upto 2 weeks	Progressive visual loss >2 weeks
Unilateral visual loss with reduced color and contrast vision, any type of visual field defect	Severe visual loss to PL-ve Bilateral visual loss
Periocular pain and painful eye movement	No pain/severe or persistent pain >2 weeks
Previous history of ON or MS	Previous history of systemic vasculitis, systemic infection, neoplasia etc.
Neurological signs and symptoms suggestive of MS	Clinical symptoms suggestive of other diseases than MS (NMO, connective tissue disorders, sarcoidosis, vasculitis etc.)

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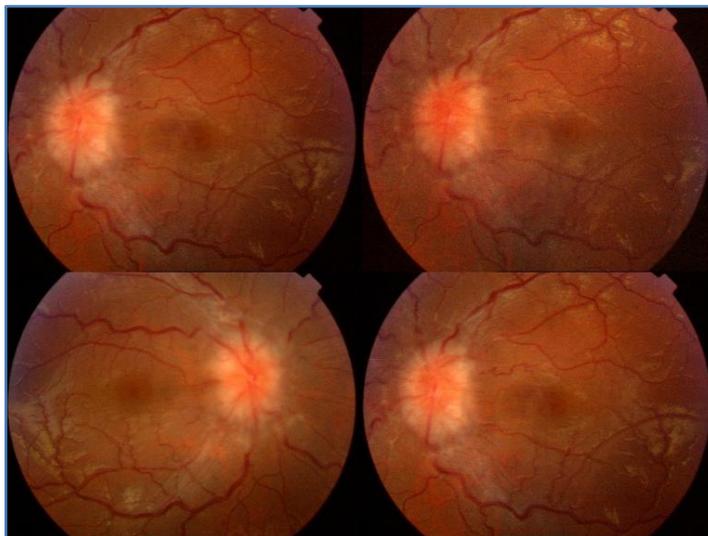
Normal or swollen optic disc	Severe optic disc edema
Normal macula and peripheral retina	Optic disc hemorrhage
Uveitis or retinal periphlebitis possible	Marked
	Optic Atrophy without h/o ON or MS
Spontaneous improvement after 2-3 weeks	Absence of recovery >3 weeks after onset
No deterioration after withdrawal of steroids	Deterioration

Typical and atypical presentations of optic neuritis

ON: optic neuritis; MS: multiple sclerosis; NMO: neuromyelitis optica

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PAPILLITIS

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Date of Submission: 24/01/2014.
Date of Peer Review: 25/01/2014.
Date of Acceptance: 13/02/2014.
Date of Publishing: 25/02/2014.