

CASE REPORT

NEURORETINITIS: A RARE CASE REPORT OF SUDDEN LOSS OF VISION IN YOUNG ADULT

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ABSTRACT: Neuro retinitis (NR) though a rare disease occurring in young adults, mostly unilateral is an inflammatory disorder characterized by swelling of optic nerve head and adjoining retinal nerve fibre layer resulting in a typical Macular star configuration due to lipid exudation within macula and fovea. Neuroretinitis can be due to an infectious process involving the disc, a post viral event, of an autoimmune inflammatory mechanism or sometimes idiopathic in nature resulting in a sudden loss of vision in affected eye along with multiple pupillary defects in form of Anisokoria (dilated on affected side), Marcus Gunn pupil or ill sustained pupillary reactions. Patients may also present with painful ocular movements mostly adduction and elevation of the eye. Features common to above all 3 groups included age, absence of pain and fundus findings. Preceding systemic symptoms and visual field loss are more common in patient with Cat scratch disease and Multiple Sclerosis and uncommon in those with recurrences. With today's technological advances, decision making regarding evaluation and treatment of NR is made accurately to reduce the incidence of optic atrophy which leads to total loss of vision.

KEYWORDS: Neuroretinitis, Acute unilateral sudden loss of vision, Pain full ocular movements.

INTRODUCTION: Theodor Leber, back in 1916 described the condition of Stellate Maculopathy. Gass in 1977 coined the term Neuroretinitis. It is peculiar form of optic neuropathy characterized by an acute unilateral visual loss causing optic disc swelling accompanied by hard exudates characteristically arranged in the star shaped pattern in fovea. It can affect all age groups although more common in 3rd & 4th decade of life, with no gender predilection. It is mostly unilateral entity and is precipitated by various etiological factors ⁽¹⁾ whose management leads to good visual prognosis. Though the term neuroretinitis emphasises clinical involvement of both disc and retina the pathologic locus is within optic nerve head and macula is not the primary disease locus. Our objective here is to present a rare case report of 26 year old female with neuroretinitis and highlight the need for its early diagnosis and treatment to have a good visual prognosis back.

CASE REPORT: A 26 year old married female presented with painless progressive decrease in vision, insidious in onset in right eye for 15 days. There was no history of trauma or contact with cats or pets. There was no recent history of joint pains or swellings, fever, headache, malaise, myalgia, flu-like symptoms, or lymphadenopathy. Patient had no history of blood transfusion or admission to hospital. History of cough or any pulmonary symptoms, sexual transmitted skin lesions, sinus infections, metabolic disorders (DM) and any adjuvant ocular problems were negative with no travel history to endemic Lyme areas.

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On examination, it was a unilateral entity. The best corrected visual acuity at presentation was HM (+) in right eye (affected eye) with no pin hole improvement and 6/6 in left eye. Colour Vision was impaired in right eye. At initial presentation a mild relative afferent pupillary defect was noticed in right eye and rest of anterior segment was normal. Fundus Examination of the affected eye (RE) (Fig. 1) revealed few overlying vitreous inflammatory cells making the media hazy. Optic disc was hyperaemic, edematous with blurred margins and had peripapillary oedema. Slit Lamp Examination with the 90 D lens confirmed presence of mild macular oedema upto +3D which seemed to be confined to the outer plexiform layer of retina. Star shaped hard exudates were distributed in the macula namely "Macular Star"⁽²⁾ and the foveal reflex was absent. There was deep subretinal exudates along inferotemporal quadrant suggestive of vasculitis, as vessels were torturous and congested with arterio-venous ratio 1:2. Venous pulsations were appreciated. Rest of the fundus was normal. Central field of the patient revealed a Centrocaecal scotoma (RE) and normal left eye fields in perimetry. ERG was normal. Ultrasonography (B Scan) of right eye showed widening of the optic nerve sheath with mild elevation of 1 mm of optic nerve head. Fundus Fluorescein Angiography (Fig. 2) was done which revealed slight leakage in the disc vessels and minimal peripapillary leak along with mild leak in inferotemporal vessels. There was no aneurismal dilatations of arterioles or macular leak. OCT (Fig. 3) revealed retinal thickening, subretinal fluid, fluid and exudates in outer plexiform layer (Henle's layer). The other eye (LE) was unremarkable in all aspects. The remaining cranial nerves and results of neurological and physical examination was normal. ELISA for parasitic & bacterial infections was negative. Serum HIV, other viral serology and cold agglutinins were negative too. ANA antibodies and HbA1c revealed negative result. Patient blood was sent to be tested for Cat Scratch Disease. CT Scan and MRI showed no findings suggestive of Multiple Sclerosis. X-ray for nose and paranasal sinuses were normal. A clinical diagnosis of Acute Idiopathic Stellate Neuroretinitis of Right eye was made for our case, following which treatment according to ONTT was started, 250mg methyl prednisolone QID along with painkillers. Methylprednisolone powder was reconstituted in distilled water and diluted in 500ml NS infusion before starting it i.v. The vision improved to 6/18, following this stability in vision for 3 days i.v was stopped and oral prednisolone 1mg/kg/day was started and then tapered for next 12 days. The deep exudates remained but otherwise fundus became normal (Fig. 4) with return of visual acuity to 6/9. Colour vision and Field became normal. No neurological symptoms supervened at any time. The patient remained asymptomatic during follow up period for future weeks for 6 months.

DISCUSSION: Neuroretinitis^(3,4) as the name suggests there is an inflammation of the neural retina and the optic nerve. Inflammation is mainly targeted on the optic disc vasculature, leading to leakage of fluid and proteins from the disc capillaries into the subretinal space and outer plexiform layer of peripapillary retina. Resolution of fluid component leaves behind lipid exudates as a "Macular Star". Only the aqueous phase is able to pass through the external limiting membrane to accumulate beneath the neurosensory retina.⁽⁵⁾ The degree of optic disc swelling ranges from mild to severe depending on the timing of the first examination. The posterior inflammatory signs consisting of vitreous cells and venous sheathing along with mild anterior uveitis also occurs.

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Neuroretinitis classified as:

1. According to fibres of the optic nerve affected: Axial Neuritis, Peri Neuritis and Trans Neuritis
2. According to the fundal picture: Papillitis, Retrobulbar Neuritis, Optic Neuritis.

The etiopathology of neuroretinitis is obscure, mediated by any infectious or immune process precipitated by number of different agents. In absence of any proven etiology, the condition is diagnosed as Leber's Idiopathic Stellate Retinopathy. Factors are classified accordingly:

Sl. No.	Etiological factors	Causes
1	Inflammation spreading from adjunct ocular structures:	- Retinitis - Choroiditis - Posterior Scleritis - Sympathetic Ophthalmitis
2	Inflammation spreading from adjunct anatomical structures:	- Sinusitis - Meningitis/ Encephalitis
3	Degenerative diseases of brain	- Demyelinating diseases - Multiple Sclerosis - Schneider's disease
4	Metabolic Disorders	- Diabetes Mellitus - Pregnancy - Lactation
5	Nutritional Disorders	- Anaemia - Hypoproteinemia - Beriberi - Wernicke's Encephalopathy
6	Vascular Group	- Leukemia - Eale's Disease - Ophthalmoplegic Migrane & Vasculitis
7	Systemic Diseases	- Cat Scratch Disease (commonly include 2/3 rd cases) - Tuberculosis - Syphilis - Leprosy - Leptospirosis - HIV, Herpes Simplex, Varicella Zoster - Toxocariasis

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8	Miscellaneous	- Inflammatory Bowel Disease - IRVAN - Poly arteritis nodosa - DUSN - Trauma
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Table 1

Investigations into etiology of neuroretinitis should begin with careful history taking & complete physical and ocular examination, CT Scan and MRI. Diagnostically, fundus picture may be confused with a few entities: Papillitis, Papilledema, Central retinal vein occlusion, anterior ischemic optic neuropathy, infiltration of optic disc by tumor and Systemic hypertension. Treatment of neuroretinitis depends on underlying infectious or inflammatory condition, only symptomatic treatment required for Idiopathic causes. Systemic corticosteroids and antibiotics along with oral corticosteroids have been reported to be effective here as per ONTT. Hence, early diagnosis and treatment of cause and symptoms ensure a good visual prognosis.

CONCLUSION: In most of the cases, neuroretinitis represents a self-limiting, benign, systemic inflammatory process with rarely a specific etiology. ONTT gives the best corrected vision to the patient containing steroids in them.

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Fig. 1: Fundus picture at time of presentation (RE) showing Macular Star with disc and peripapillary oedema



Fig. 2: FFA revealed a slight leakage in disc vessels and peripapillary leak along with mild leak in inferotemporal vessels

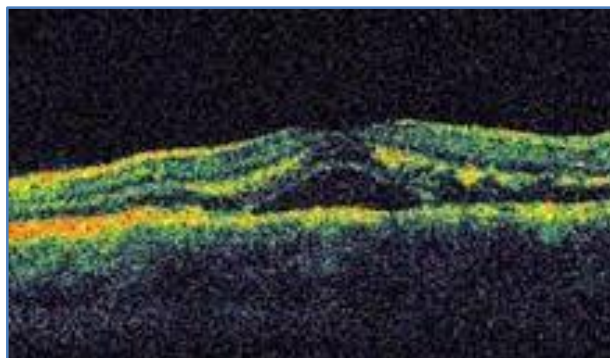


Fig. 3: OCT showing subretinal fluid

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Fig. 4: Followup of the patient after prompt treatment with steroids (ONTT)

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