Systemic Lupus Erythematosus Presenting as Neuroretinitis

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Abstract
Neuroretinitis is the inflammation of retina and optic nerve. It is associated with optic disc edema accompanied by peripapillary or macular hard exudates. A 17 yr old female presented with headache and nausea of five days duration. She had periorbital edema and mild splenomegaly. Neurological assessment was non-contributory. She was found to have pancytopenia, albuminuria and a high ESR. Thereafter she developed blurring of vision of both eyes. Ophthalmological examination showed it to be due to bilateral neuroretinitis. ANA and anti-ds DNA were strongly positive. Renal biopsy with immunofluorescence study revealed diffuse global proliferative lupus nephritis with active lesions [class IV-G (A)]. She was diagnosed as a case of SLE presenting with neuroretinitis.

Introduction
Ocular manifestations in systemic lupus erythematosus (SLE) are usually due to vaso-occlusive disease involving retinal and choroidal vessels, however direct inflammatory damage may also occur. Neuroretinitis refers to an optic neuropathy in which optic disc edema is accompanied by peripapillary or macular hard exudates. Neuroretinitis due to direct inflammatory damage in SLE has been described rarely in SLE. We have presented a case of a young woman with bilateral neuroretinitis due to SLE.

Case Report
A 17 yr old female presented with headache and nausea of five days duration. Headache was diffuse and of moderate intensity. There was no history of fever, cough, respiratory distress, chest pain, jaundice, frothing or reddish colour urine, convulsion, altered sensorium or any skin rash and photosensitivity. There was no history of exposure to cats, birds or pets. However she had a history of pain in both knees and left shoulder joint for last three months. On examination she had mild bilateral periorbital edema and tachycardia. She was normotensive. She had splenomegaly which was 4 cm in length from the costal margin. The neurological examination was non-contributory. Oral ulcer, alopecia, swelling and deformity of joints, reddening and watering from eyes were absent.

Investigations revealed pancytopenia (hemoglobin 7.4 gm/dl, total count 3800/cumm, neutrophil 59%, lymphocyte 34%, monocyte 6%, eosinophil 1%, platelet 1.2 lakhs/cumm) and a high ESR of 110 mm/hour. CRP was positive (24 mg/L). Urine examination showed albuminuria which was quantified as 400 mg in a 24 hr sample. No active urinary sediment was present. Thyroid profile, liver function tests (bilirubin 0.6 mg/dl, SGOT 28 IU/dl, SGPT 12 IU/dl, serum protein 7 g/dl, albumin 3.7 g/dl and globulin 3.3 g/dl), lipid profile, urea, creatinine, electrolytes, blood sugar and ASO titre were all within normal limits. Imaging such as X-rays of the joints, chest X-ray, ECG, and echocardiography were normal. Ultrasonography of the abdomen showed splenomegaly of size 16 cm. Viral markers, i.e. anti-HAV IgM antibody, anti-HCV antibody, HBsAg, HIV, Mantoux test and VDRL were negative. Serological tests for toxoplasma, leptospira, herpes, mumps and tuberculosis were negative.

During the next one week her headache worsened and she started developing blurring of vision of both eyes. Ophthalmological check up showed a visual acuity of 6/12 in both eyes and fundoscopy showed bilateral optic disc swelling with macular star suggestive of neuroretinitis (Figure 1). There was no afferent pupillary defect.

Thereafter a CT scan of brain was done, which was within normal limit. It was followed by MRI of brain which was also normal (diffusion imaging showed no abnormal signals, bilateral optic nerves were normal, and flow in cerebral blood vessels was normal). A CSF study was done which showed albumino-cytological dissociation (2 cells, all lymphocytes; 354 mg protein), sugar 78 mg/dl, and adenosine deaminase 1 U/L (normal range <5 U/L). Following this, serological studies of antinuclear antibody (Hep2) (titre 1:100), and anti-double stranded DNA antibody (strength 1:67) were done which were strongly positive. Anti-ribosomal P was negative. Antiphospholipid antibodies including beta-2 glycoprotein were negative. D dimer assay and FDP were normal. Complement C3 level was decreased (0.51 g/L, normal range 0.9 to 1.8 g/L). Rheumatoid factor was negative (6.3 IU/mL, normal <35 IU/mL). Anti SS-A and anti SS-B antibodies were negative. Thus, the case was diagnosed to have SLE on the basis of pancytopenia, high titre of ANA, positive anti-ds-DNA, albuminuria and neuroretinitis. Renal biopsy with immunofluorescence study revealed diffuse global proliferative lupus nephritis with active lesions [class IV-G (A)] (Figure 2). She was then started on oral prednisolone 40 mg/day and mycophenolate mofetil 2 gm/day. Gradually her headache, blurring of vision and periorbital

Fig. 1: Showing left optic disc swelling with macular star

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Neuroretinitis is inflammation of retina and optic nerve. It is associated with swelling of optic disc, peripapillary and macular hard exudates (that may occur in a star-shaped pattern) and cells in vitreous. Neuroretinitis usually presents with acute unilateral painless loss of vision. Visual acuity at the time of initial examination ranges from 6/6 to light perception (PL). A relative afferent pupillary defect (RAPD) is usually present unless the condition is bilateral. Perimetry often reveals a central or centrocecal scotoma.

Optic disc swelling is the earliest sign of neuroretinitis. It may be severe and associated with peripapillary edema and splinter hemorrhages. Resorption of serous fluid around the macula leaves lipid precipitates in the form of a macular star. Macular star may take up to a year. Edema improved over the next few weeks. Ophthalmoscopy revealed disappearance of neuroretinitis.

**Discussion**

Neuroretinitis is inflammation of retina and optic nerve. It is associated with swelling of optic disc, peripapillary and macular hard exudates (that may occur in a star-shaped pattern) and cells in vitreous. Neuroretinitis usually presents with acute unilateral painless loss of vision. Visual acuity at the time of initial examination ranges from 6/6 to light perception (PL). A relative afferent pupillary defect (RAPD) is usually present unless the condition is bilateral. Perimetry often reveals a central or centrocecal scotoma.

Optic disc swelling is the earliest sign of neuroretinitis. It may be severe and associated with peripapillary edema and splinter hemorrhages. Resorption of serous fluid around the macula leaves lipid precipitates in the form of a macular star. Macular star tends to become more prominent one to two weeks later or when optic disc swelling resolves. Hence there is need to re-examine patients with acute papillitis with a normal macula within two weeks for the development of a macular star. Resolution of macular star may take up to a year. Dyschromatopsia is a common feature due to affection of ganglion cells and macula. Posterior inflammatory signs consist of vitreous cells and venous sheathing. Mild anterior uveitis may also occur. Neuroretinitis often resolves spontaneously and most patients have full restoration of visual function.

Neuroretinitis may be idiopathic (idiopathic optic disc edema with a macular star). Bacteria and viruses can cause this condition. Infectious organisms causing neuroretinitis include *Bartonella henselae* (cat-scratch disease), *Toxoplasma gondii*, *Borrelia burgdorferi* (Lyme disease), leptospira, *Rickettsia typhi*, *Treponema pallidum*, *Mycobacterium tuberculosis*, *Histoplasma capsulatum*, and various viruses, such as HIV, mumps, herpes zoster, hepatitis B and herpes simplex virus. Immunological disorders are rarely reported to cause neuroretinitis. Anecdotal reports of neuroretinitis are seen in ulcerative colitis and multiple sclerosis. Neuroretinitis due to SLE with or without antiphospholipid syndrome has been reported in literature very rarely. Other ocular manifestations of SLE include sicca syndrome and conjunctivitis.

**Neuroretinitis should be distinguished from several fundoscopically confusing conditions such as papilledema, ischemic optic neuropathy, optic neuritis, compressive lesions, toxic/nutritional deficiencies, and hereditary forms. Time course, presence or absence of pain, pattern of visual loss, visual field defects and fundoscopic appearance help to differentiate it from the others (Table 1).**

**Neuroretinitis as an acute presentation in SLE is uncommon. However autoimmune profiles including ANF should be investigated if a patient has any suggestive feature of immunological disorder. High index of suspicion is needed to diagnose SLE in a patient with neuroretinitis, otherwise diagnosis may be delayed.**

**Table 1: Differences between neuroretinitis and other funduscopically resembling entities**

<table>
<thead>
<tr>
<th></th>
<th>Neuroretinitis</th>
<th>Papillitis</th>
<th>Papilledema</th>
<th>Central retinal vein occlusion (CRVO)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visual acuity</td>
<td>6/6-6/12</td>
<td>Light perception</td>
<td>No visual loss</td>
<td>Moderate to severe visual loss</td>
</tr>
<tr>
<td>Pupillary reaction</td>
<td>RAPD+</td>
<td>RAPD+</td>
<td>Normal</td>
<td>Normal / RAPD+</td>
</tr>
<tr>
<td>Laterality</td>
<td>Unilateral, rarely bilateral</td>
<td>Unilateral / bilateral</td>
<td>Always bilateral</td>
<td>Unilateral</td>
</tr>
<tr>
<td>Eye pain</td>
<td>Nil</td>
<td>In upward gaze</td>
<td>Nil</td>
<td>Nil</td>
</tr>
<tr>
<td>Visual field</td>
<td>Centrocecal scotoma</td>
<td>Central / centrocecal scotoma</td>
<td>Enlarged blind spot</td>
<td>Normal</td>
</tr>
<tr>
<td>Colour vision</td>
<td>Severely impaired, disproportionate to visual loss</td>
<td>Severely impaired, disproportionate to visual loss</td>
<td>Disc swelling higher up to 8-9</td>
<td>Disc edema, macular star along with hemorrhages and soft exudates</td>
</tr>
<tr>
<td>Fundus</td>
<td>Disc hyperemic, with swelling of 2D, macular star</td>
<td>Disc swelling rarely above 2D, venous engorgement and hemorrhage less marked</td>
<td>Disc swelling higher up to 8-9</td>
<td>Disc edema, macular star along with hemorrhages and soft exudates</td>
</tr>
<tr>
<td>Fluorescence angiography</td>
<td>Leakage from disc and peripheral retina</td>
<td>Leakage from disc and peripheral retina</td>
<td>Leakage from disc and peripheral retina</td>
<td>Areas of capillary non-perfusion</td>
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<tr>
<td>VEP</td>
<td>Decreased in amplitude, increase in latency</td>
<td>Decreased in amplitude, increase in latency</td>
<td>Normal</td>
<td>Normal / decreased amplitude</td>
</tr>
</tbody>
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**References**