

A Case of Reversible Blindness in Systemic Lupus Erythematosus

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Abstract

We report a case of reversible blindness in a lady with systemic lupus erythematosus (SLE) with nephritis and accelerated hypertension. CT scan brain revealed a symmetric hypodense lesion of the occipital lobes, which disappeared in two weeks time with treatment.

Key Words

SLE, Reversible blindness, CT scan

Introduction

Systemic lupus erythematosus is a multiorgan autoimmune disease with a wide spectrum of clinical manifestations affecting various organ systems, either singly or in any combination. Causes for blindness in SLE are protean, which may be due to a lesion in the visual pathway. It may be monocular or binocular, reversible or irreversible. The pathology may be due to vasculitis, thromboembolic events, haemorrhage, retinal detachment, antiphospholipid antibody syndrome, demyelination or related to drug therapy. Reversible blindness in SLE may be due to migraine, optic neuritis, retrobulbar neuritis and reversible posterior leucoencephalopathy (RPLE). Here we report an unusual case of reversible cortical blindness in SLE.

Case Report

We report a 34 yr old lady, a home manager and a mother of two healthy children, admitted to the Department of Medicine, with a history of headache of 6 months duration, fronto-occipital in nature and increasing in the early hours of morning. On examination she was found to be a hypertensive (180/110mmHg). Systemic examination revealed no abnormality, contributing to systemic hypertension in a young woman.

On evaluation she had a hemoglobin of 12gm%, total count of 9300/cumm with normal differential count

of P60,L38,E2, platelet count of 1.5 lakhs/cumm and a raised ESR of 120mm/hr. Chest skiagram, electrocardiogram and echocardiogram were within normal limits. Renal evaluation revealed nephrotic range of proteinuria (3.2gm/day), no active urinary sediments, azotemia (blood urea 60mg/dl and serum creatinine 2.3mg/dl) with normal sized kidneys on ultrasonogram. A renal biopsy was done, which revealed membranous nephropathy with focal atrophy of tubules and interstitial fibrosis with full house immunofluorescence suggestive of lupus nephritis, although she denied history of photosensitivity, skin rashes, joint symptoms, increased hair fall, mucosal ulcers and neuropsychiatric symptoms. Immunological tests revealed antibodies to nuclear antigens (ANA+++ homogeneous) and antibodies to double stranded DNA (dsDNA). Lupus anticoagulant and anti-cardiolipin antibodies were negative. Diagnosis of lupus nephritis class V was made and she was put on oral prednisolone 1mg/kg/day along with antihypertensive medications and discharged.

She came back to the emergency room in 6 weeks time with intolerable headache, accelerated hypertension (210/120mmHg) along with worsening of azotemia (blood urea 110mg/dl; serum creatinine 6.4mg/dl) and metabolic acidosis. Antihypertensive medications were stepped up

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and she was taken up for peritoneal dialysis, during which she developed vomiting, seizures, altered sensorium, worsening of headache and loss of vision in both eyes. She was referred to Rheumatology Department for evaluation of SLE with blindness. There was no focal neurologic deficit except for cortical blindness (blindness with retained pupillary reflexes). Fundus oculi revealed mild AV nipping and no papilloedema. Her CT scan brain revealed symmetric hypodense lesion in both occipital lobes (Fig.1). She was treated aggressively with anti-hypertensives to reach the target BP along with antiepileptics and antioedema measures. To our surprise her vision returned back to normalcy in 2 week's time and repeat CT scan brain revealed no abnormality (Fig.2).

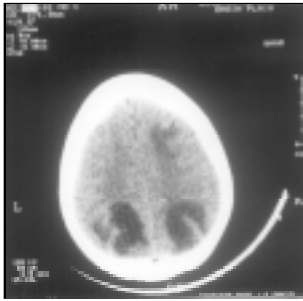


Fig. 1

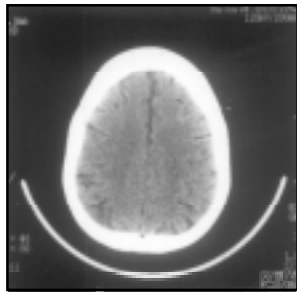


Fig. 2

Fig. 1 CT Scan brain showing symmetric hypodense shadows over the posterior regions of cerebral hemispheres.

Fig. 2 Repeat CT Scan brain done after two weeks showing complete clearance of the lesions.

Discussion

Acute visual loss in patients with SLE may be due to retinal or choroidal ischaemia from vasculitis or occlusion (vasculopathy), retinal detachment, infectious retinitis, glaucoma, optic retrobulbar neuritis or even cortical infarcts of the brain (1). RPLE syndrome is seen in some patients who are hospitalized for acute illness and develop a reversible syndrome with headache, altered mental functions, seizures and loss of vision accompanied with findings of extensive bilateral white matter abnormalities suggestive of edema in the posterior regions of the cerebral hemispheres (posterior leucoencephalopathy) on neuroimaging, described by Hinchey et al (2). RPLE may develop in patients with renal insufficiency or accelerated hypertension or who are on immunosuppressive drugs. The findings on neuroimaging are characteristic of subcortical edema without infarction.

The clinical signs and findings on neuroimaging in RPLE should be readily recognised and treated promptly by aggressive control of BP and or avoiding the offending immunosuppressive drug if any. RPLE has been reported in pregnancy induced hypertension, polyarteritis nodosa, Wegener's granulomatosis and malignant melanoma on cytotoxic drugs. The cerebral white matter is composed of myelinated fibres in a cellular matrix of glial cells, arterioles and capillaries that makes it susceptible to accumulation of fluid in the extracellular space due to capillary leakage and acute disruption of the blood brain barrier. RPLE should not be mistaken for cerebral infarction, where less aggressive control of BP is needed to prevent the infarct zone from extending (3). It is also mandatory to differentiate it from lupus cerebritis in view of avoiding immunosuppressant therapy which may indeed worsen RPLE (4). The calcarine and paramedian occipital lobe structures are spared in RPLE which differentiates it from bilateral cerebral infarction of posterior cerebral artery territory (2).

Radiologic diagnosis of RPLE can be made with CT scan brain when there is a symmetric white matter disease in the posterior cerebral region. Although magnetic resonance imaging (MRI) gives a higher resolution of image, it is not necessary for diagnosis of RPLE. The only advantage MRI offers is to show small focal abnormalities beyond the resolution of CT (5). In our patient the findings suggestive of RPLE on CT scan brain was evident and the visual loss was reversible with aggressive control of hypertension.

This phenomenon of RPLE may be considered as a silver lining to the dark gloomy cloud of blindness in atleast a few fortunate cases of SLE.

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