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CASE REPORT: A CASE OF YOUNG BOY WITH SLE, ANTIPHOSPHOLIPID SYNDROME & AIHA

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ABSTRACT
A 14 year old male child, diagnosed as a case of autoimmune hemolytic anemia since 3 years of age, on oral Prednisolone, was admitted with complaints of sudden onset of right eye vision loss and left hemiparesis and convulsions. He had cushingoid features and raised blood pressure. His workup showed anemia, thrombocytopenia, positive ANA, anti dsDNA, APLA and Coombs direct test. MRI Brain showed multiple infarcts and opthalm check up showed right eye CRVO. His final diagnosis was Systemic Lupus Erythematosus with antiphospholipid syndrome with AIHA, Right central retinal vein thrombosis, multiple infarcts in brain with left hemiparesis, hypertension, exogenous cushingoid.

CASE REPORT

A 14 year old male child, was admitted with complaints of sudden onset diminished vision of right eye and weakness of left upper and lower limb since 15 days. He developed 2 episodes of generalised tonic clonic convulsions on day of admission to Bharati Hospital. Patient was diagnosed as a case of autoimmune hemolytic anemia since 3 years of age and had received blood transfusions 4 times over the years. He was currently on oral Prednisolon since 1 year, and had developed cushingoid features (Figure 1). His birth and family history were insignificant; immunization status complete.

On examination he had grade 3 power in left upper and lower limbs and cushingoid features, raised blood pressure for age. He developed tingling sensation first in left upper limb. Nerve connduction study showed left upper limb sensory motor axonal neuropathy, EMG showed decreased activation. His hemiparesis improved over 2 weeks.

His workup showed anemia Hb 7.4g/dl, TLC 11100, thrombocytopenia 52000/cmm, raised ESR 89 mm at 1 hour, CRP 14.23; ANA, dsDNA, Coomb's test were positive. IgM Antiphospholipid antibodies were positive; phosphlipid profile showed raised titres of IgG anticardiolipin antibodies, equivocal titres of IgG phosphatidyl serine. MRI Brain showed multiple lacunar infarcts in right centrum semiovale, left high parietal lobe, both corona radiata with normal MR angiography of brain (Figure 2); MRI spine was also normal. Fundoscopy showed Right eye central vein thrombosis. Urine protein creatinine ratio and renal doppler were normal. He was treated with levetiracetam, warfarin, prednisolone and amlodipine. He is currently on azoran 50 mg once daily with prednisolone dose being tapered.

His final diagnosis was Systemic Lupus Erythematosus with antiphospholipid syndrome & AIHA, Right central retinal vein thrombosis, multiple infarcts in brain with left hemiparesis, hypertension, exogenous cushingoid.

DISCUSSION

Our patient is a rare case in many aspects. He is case of paediatric SLE, male patient which is less common. He fulfilled 4 out of 11 diagnostic criteria-- neurologic, hematologic, immunologic & antinuclear antibodies. Our patient initial presentation was 'Autoimmune Hemolytic Anemia (AIHA) at age of 3 years. In literature hemolytic

Fig 1. Patient with cushingoid features due to prednisolone

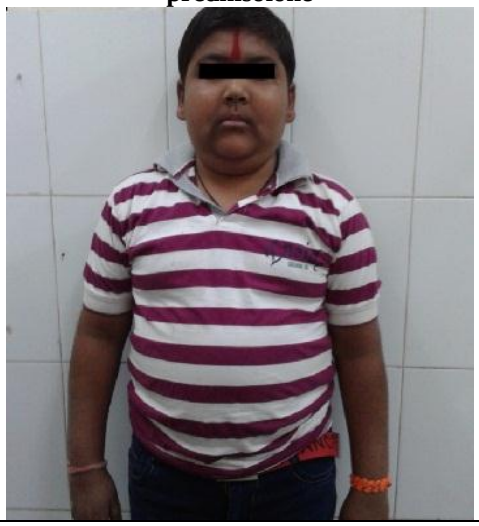
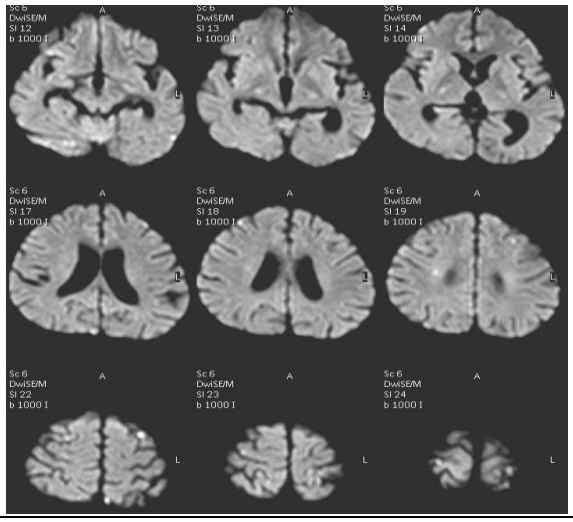


Fig 2. MRI Brain of patient showing multiple lacunar infarcts



anemia occurs in overall 10% patients with SLE. In a case series of AIHA with SLE by Kokori *et al*, 66% had hemolysis as initial disease presentation and was associated with renal involvement, thrombocytopenia and possibly thrombosis [1].

Antiphosphlipid antibodies can be present in upto 50% of SLE patients [2] and is associated with thrombosis, thrombocytopenia and fetal loss (in females). Our patient had arterial thrombosis in cerebral circulation, likely inflammatory type, and venous thrombosis in central retinal vein both occuring simultaneously, which is rare. Cerebral ischemia is most common arterial manifestation of APLA syndrome [3]. Central Retinal Vein Thrombosis (CRVO) is itself rare in paediatric cases; and it is significantly associated with antiphospholipid antibodies & anti cardiolipin antibodies; but not with lupus anticoagulant [4]. This was shortly followed by seizures, which can occur in upto 20% of patients with SLE.

ACKNOWLEDGEMENT

None

CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest.

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