Purtscher like retinopathy as the initial presentation of paediatric systemic lupus erythematosus in a 4 year old child

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Introduction

The prevalence of systemic lupus erythematosus (SLE) in children is 1-6 per 100,000 and it is rare before 5 years of age[1]. We present a 4 year old girl with SLE whose diagnosis was first suspected based on the finding of Purtscher like retinopathy.

Case report

A 4 year old girl, the product of a non-consanguineous marriage, presented with a history of poor weight gain in the past two years, intermittent fever, skin rash and alopecia for 2 months. On examination she was febrile with pallor, tachycardia, stage 3 hypertension, generalised lymphadenopathy, oral ulcers, malar rash involving nasal bridge and patchy alopecia. Examination also revealed diminution of vision. On funduscopic examination, there were multiple cotton wool spots and flame shaped haemorrhages in both eyes suggestive of Purtscher like retinopathy (PLR) (Figure 1).

Figure 1: A. Purtscher like retinopathy B. Resolution of Purtscher like retinopathy after treatment C. Malar rash D. Improvement of malar rash after treatment

**Permission obtained from parents to publish photograph*

There was anaemia (haemoglobin 8.9g/dL), with normal total leucocyte count and platelet count, blood urea and serum creatinine. Erythrocyte sedimentation rate was 105mm at the end of one hour. C3 complement level was 73mg/dL (normal 80-160mg/dl), C4 complement level 12mg/dL (normal 16-48mg/dl) and C1q level 7.8mg/dL (normal 5-8.6mg/dL). She had raised transaminases with reversal of albumin to globulin ratio (albumin-2.9 g/dL globulin 6.2g/dL). Spot urine protein–to-creatinine ratio was 3.61 (Normal <0.2), suggestive of nephrotic range proteinuria. Antinuclear antibody (ANA) was homogeneously positive on indirect immunofluorescence. Anti-double-stranded DNA (dsDNA) titre was 1286 IU/mL (normal <100 IU). Antiphospholipid antibody was negative. The child was diagnosed as a case of SLE with a Systematic Lupus International Collaborating

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Clinics (SLICC) criteria score of 7 with PLR with Systemic Lupus Erythematosus Disease Activity Index (SLEDAI**)** 32. As there was no differential diagnosis, renal biopsy was not done.

In view of impending blindness and renal disease, she was immediately started on intravenous (IV) pulsed methylprednisolone (30mg/kg body weight) followed by pulsed IV cyclophosphamide with oral prednisolone as per National Institute of Health (NIH) protocol. Child responded to the treatment with improved vision and clearing of the eye lesions after 3 months of follow up. IV cyclophosphamide was given for 6 months followed by azathioprine maintenance. Child is currently on follow up for the past one year and is on hydroxychloroquine, low dose prednisolone (0.125mg/kg body weight) and azathioprine.

Discussion

Paediatric SLE has a higher morbidity and mortality than adult SLE[2]. Ocular manifestations can be present, of which SLE retinopathy is the most common. It is mostly seen in active disease and is associated with antiphospholipid antibodies[3], but it was negative in our case.

Purtscher retinopathy was first described by Otmar Purtscher, an Austrian ophthalmologist, in 1910, in a middle aged man with head injury and comprised multiple areas of whitening and haemorrhage in the posterior poles of both eyes[4]. Purtscher-like retinopathy (PLR) describes similar features on fundoscopy in conditions other than trauma. It is classically characterised by cotton wool spots, retinal haemorrhages, optic disc oedema and purtscher flecken which may lead to temporary or permanent blindness. PLR, as an initial manifestation of adult SLE, is well described in the literature but it is very rare in children. Pathogenesis of PLR is unknown. One proposal is that complement activation and leuco-embolisation by leucocyte aggregation with secondary lymph extravasation are responsible for PLR[5,6].

PLR is a rare manifestation in SLE and to the best of our knowledge has not been reported in paediatric lupus with onset below 5 years of age. It is imperative for clinicians to know about this manifestation to enable early recognition and vision-preserving strategies.

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