**Case Reports**

**Discoid lupus erythematosus: First clue to autoimmune haemolytic anaemia in a child**

*K M D Mallesh, Arun Basavanahalli Chandregowda, Prakash Javarappa*


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**Introduction**

Discoid lupus erythematosus (DLE) is uncommon in the first decade of life. It has been described as part of systemic lupus erythematosus (SLE) or in isolation in adults. We report a case of severe haemolytic anaemia with discoid lesions that gave a clue to diagnosis and management.

**Case report**

A 10 year old girl presented with headache, fever, weakness and skin lesions over face for 1 month. Child had been transfused with blood earlier for severe anaemia. Examination revealed severe pallor, jaundice, hepatosplenomegaly and congestive heart failure. There were two discoid, scaly macules with irregular margins and peripheral hyperpigmentation in the left side of the face below lower eyelid (Figure A).

Investigations revealed a haemoglobin level of 4.7 g/dl, dimorphic anaemia, positive direct Coombs test, positive antinuclear antibodies, negative venereal disease laboratory test, negative double stranded deoxyribonucleic acid, negative anti-phospholipid antibodies and negative ribonucleoprotein. Renal function tests were within normal limits. There was cardiomegaly in chest x-ray and mild left ventricular dysfunction with no evidence of pericardial effusion or valvular regurgitation on 2-dimensional echocardiography. Skin lesion biopsy showed basal cell degeneration and hyperkeratosis with follicular plugging suggestive of DLE. Only three criteria instead of four were present in the child out of 11 as per revised American College of Rheumatology Criteria for diagnosing SLE. Child was treated with pulsed methyl prednisone (30 mg/kg) for five days followed by prednisone 1 mg/kg/day for 4 weeks tapering over next 4 weeks. Congestive heart failure was managed with furosemide and enalapril. Discoid lesions were treated with topical triamcinolone acetonide 0.1% cream and sun screening. Lesions responded to treatment over 4 weeks (Figure B). Haemoglobin level improved and child was asymptomatic at latest follow up.

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1Department of Paediatrics, Bangalore Medical College, 2Bangalore Medical College and Research Institute, Fort, Bangalore, Karnataka, India, *Correspondence: drkmallesh@rediffmail.com*

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Discussion

Discoid lesions without other manifestations may be mistaken for pityriasis alba and pityriasis versicolor. It should be confirmed by skin biopsy and immunological tests. Revised College of Rheumatology criteria for the classification of SLE should be applied to differentiate this from SLE².

The median age at diagnosis of DLE is 30 years¹. SLE appears in 17-30% presenting as only discoid lesions and discoid lesions appear in 8-28% of SLE³,⁴,⁵. When confined to head and neck DLE is less likely to progress to SLE (5%) than when it presents below the neck (20%)⁶,⁷. Reported median age at diagnosis of SLE with DLE is 41.5 years¹. SLE patients with discoid lupus are less likely to present end-stage renal disease⁸.

Treatment of DLE includes topical steroids, tacrolimus 0.1% ointment, pimecrolimus 1% cream and sunscreening⁹. Tacrolimus 0.1% ointment and pimecrolimus 1% cream are safe and effective in resistant cases of DLE or facial DLE where topical steroid use heightens risk of thinning and telangiectasia⁸. Persistent, severe lesions could be treated with full thickness skin graft. Recurrence with skin grafting is treated with topical steroid¹⁰.

The presence of DLE should be searched for in all autoimmune haemolytic anaemia regardless of age.

References


