Case Reports

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

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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[1]. 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 4.7g/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 (30mg/kg) for five days followed by prednisone 1mg/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.

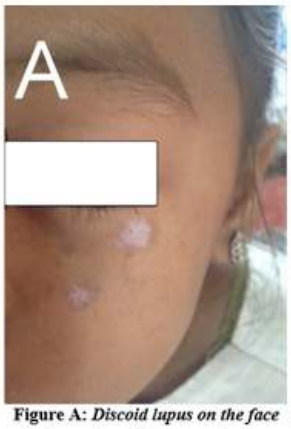

Figure A: *Discoid lupus on the face*
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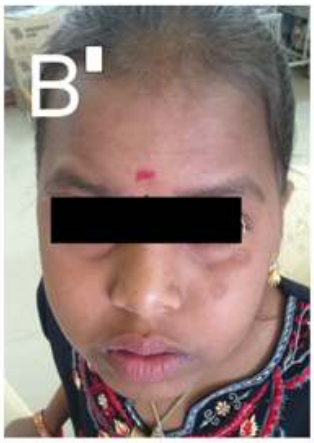

Figure B: *Discoid lupus healing after topical steroid and sun screening*
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Discussion

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 SLE[2].

The median age at diagnosis of DLE is 30 years[1]. SLE appears in 17-30% presenting as only discoid lesions and discoid lesions appear in 8-28% of SLE[3,4,5]. When confined to head and neck DLE is less likely to progress to SLE (5%) than when it presents below the neck (20%)[5,7]. Reported median age at diagnosis of SLE with DLE is 41.5 years[1]. Haemolysis is commoner with discoid lesions[1] and in paediatric SLE. SLE patients with discoid lupus are less likely to present end-stage renal disease[8]. Our case did not fulfil diagnostic criteria for SLE.

Treatment of DLE includes topical steroids, tacrolimus 0.1% ointment, pimecrolimus 1% cream and sunscreening[9]. 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 telangiectasia[8]. Persistent, severe lesions could be treated with full thickness skin graft. Recurrence with skin grafting is treated with topical steroid[10]. The presence of DLE should be searched for in all autoimmune haemolytic anaemia regardless of age.

References

1. Skare TL, Weingraber E, Stadler B, De Paula DF. Prognosis of patients with systemic lupus erythematosus and discoid lesions. *An Bras Dermatol* 2013; **88**(5):755-8. http://dx.doi.org/10.1590/abd18064841.20132042 PMid: 24173181 PMCid: PMC3798352

2. Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. *Arthritis and Rheumatism* 1997; **40**: 1725. http://dx.doi.org/10.1002/art.1780400928 PMid: 9324032

3. Zecević RD, Vojvodić D, Ristić B, Pavlović MD, Stefanović D, Karadaglić D. Skin lesions: an indicator of disease activity in systemic lupus erythematosus? *Lupus* 2001; **10**: 364-7. http://dx.doi.org/10.1002/art.1780400928 PMid: 9324032

4. Magro CM, Crowson AN, Harrist TJ. The use of antibody to C5b-9 in the sub-classification of lupus erythematosus. *British Journal of Dermatology* 1996; **134**:855-62. http://dx.doi.org/10.1046/j.13652133.1996.117852.x PMid: 8736325

5. Gilliam JN, Sontheiner RD, Distinctive cutaneous subsets in the spectrum of lupus erythematosus. *Journal of the American Academy of Dermatology* 1981; **4**: 471-5. http://dx.doi.org/10.1046/j.13652133.1996.117852.x PMid: 8736325

6. Berbert ALCV, Mantese SAO. Cutaneous lupus erythematosus - Clinical and laboratory aspects. *An Bras Dermatol* 2005; **80**: 119-31.

7. Callen JP. Management of skin disease in patients with lupus erythematosus. *Best Practice and Research. Clinical Rheumatology* 2002; **16**: 245-64. http://dx.doi.org/10.1053/berh.2001.0224 PMid: 12041952

8. Santiago-Casas Y, Vilá LM, McGwin G Jr., Cantor RS, Petri M, Ramsey-Goldman R, et al. Association of discoid lupus with clinical manifestations and damage accrual in profile: A multiethnic lupus cohort. *Arthritis Care and Research (Hoboken)* 2012; **64**(5): 704–12. http://dx.doi.org/10.1053/berh.2001.0224 PMid: 12041952

9. Han YW, Kim HO, Park SH, Park YM, Four cases of facial discoid lupus erythematosus successfully Treated with Topical Pimecrolimus or Tacrolimus**.** *Annals of Dermatology* 2010; **22**(3):307-11. http://dx.doi.org/10.5021/ad.2010.22.3.307 PMid: 20711267 PMCid: PMC2917684

10. Neuman Z, Shulman J, Ben-Hur N. Successful skin grafting in discoid lupus erythematosus: Recurrence checked by triamcinolone ointment (Ledercort). *Annals of Surgery* 1961; **154**(1):142-4. http://dx.doi.org/10.1097/00000658-196107000-00023 PMid: 13728537 PMCid: PMC1465846