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Cutaneous Lupus Erythematosus In Children

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SUMMARY

Lupus erythematosus (LE) in children is an important clinical problem in view of rarity, lack of data, syndromal association and difficulties in management. In this study, 13 patients, below 14 years of age were investigated clinically, histopathologically, and immunologically. Out of this number, 11 patients had discoid LE, two had subacute cutaneous LE, and one patient progressed from discoid LE to subacute cutaneous LE. Syndromal association was found in two cases of Russell Silver Syndrome. Chloroquine was not well tolerated.

Introduction

Lupus erythematosus in children is generally considered a rarity.[1] Damm et al.[2] reported 3-4% of all LE to occur in childhood. While childhood LE, as in adult LE, consists of different subsets,[3] chronic discoid LE (CDLE) is particularly rare.[4] The problem is important in view of occasional familial occurrence,[5,6] difficulties in drug therapy, association with other diseases such as chronic granulomatous disease[7,8,9] and hereditary complement deficiency.[10]

We studied 13 children with cutaneous LE with respect to clinical immunological, histopathological findings and therapeutic aspects. The purpose of this study is to document the difficulties in management and to find associations with other diseases, if any.

Material and Methods

Thirteen patients with LE, presented to the pediatric dermatology unit, Al Nahdha Hospital, Oman during the period 1986-1992 were included in this study. A detailed history was taken with particular reference to age at onset, cutaneous and systemic symptoms and family history. Detailed clinical cutaneous and systemic examination was done. All patients were subjected to the following investigations: peripheral blood examination, serological tests including LE cell, ANA, Anti dsDNA, Anti Ro and La antibodies, Anti ENA, serum complement levels, VDRL, and rheumatoid factor. Biopsies were taken from lesional and uninvolved skin (deltoid) for routine histopathology and direct immuno-fluorescence.

Results

All patients were aged less than 14 years, with male to female ratio of 1:1. The youngest patient was 3 years. Duration of lesions varied from 2 months to 6 years.

Table 1. The presenting complaints of the patients

COMPLAINTS	NO. OF PATIENTS	PERCENTAGE
CUTANEOUS:		
Erythematous Plaques (DLE lesion)	12	92.3
Photo Sensitivity	4	30.8
Malar Erythema	1	7.7
Oral Ulcers	2	15.4
Digital Scars	1	7.7
SYSTEMIC:		
Recurrent Fever	3	23.1
Polyarthralgia	4	30.8
Recurrent Fever	3	23.1
Recurrent Infections	1	7.7
Convulsions	1	7.7

Table 1 shows the presenting complaints. Rash on face (DLE-like) (12 cases) and photosensitivity (4 cases) were the common cutaneous presenting complaints. Malar erythema was the presenting complaint in one case. Only 4 cases had systemic complaints, such as recurrent fever (3 cases), polyarthralgia (4 cases), recurrent infections (3 cases) and convulsions (1 case). Family history of similar illness was found in 3 cases with first degree relatives also having CDLE.

Table 2 shows details of clinical examination. Typical DLE rash was noted in 12 patients. The lesions had typical features of location over exposed areas, erythema, and scarring. However scarring was mild. Follicular plugging was seen in only 2 cases. Tin-tac sign was found in only one case. Other cutaneous findings included photosensitive rash (5 cases), oral ulcers (2 cases), and alopecia (2 cases).

Table 2. Summary of clinical findings.

FINDINGS	NO. OF PATIENTS	PERCENTAGE
CUTANEOUS:		
DLE Rash	12	92.3
Butterfly Erythema	2	15.4
Oral Ulcers	2	15.4
Digital Scarring	1	7.7
Photosensitive rash	5	38.46
Psoriasiform lesion	1	7.7
Alopecia	2	15.4
SYSTEMIC:		
Generalized Lymphadenopathy	2	15.4
Growth retardation	3	23.1
Russell Silver Syndrome	2	15.4

In 12 cases, clinical diagnosis of discoid lupus erythematosus was made. During follow-up period, after 4 years of initial diagnosis, one patient developed psoriasiform lesions over forearms, oral ulcers, and polyarthralgia and was diagnosed as subacute cutaneous lupus erythematosus. One patient had digital scarring, oral ulcers, and malar erythema, with systemic findings of polyarthralgia and weight loss. He was diagnosed as possible subacute cutaneous lupus erythematosus. Two brothers with DLE had features of Russell Silver syndrome; such as poor somatic growth, characteristic faces with triangular jaw, normal mentality, and low birth weight.

Table 3 shows the results of investigations. ESR was raised in 2 cases, antinuclear antibody of speckled pattern was positive in titters of 1:160 in two cases, and anti Ro antibody was positive in one case.

Histopathological examination with routine H&E stain was confirmatory in 9 cases and suggestive in 4 patients. Direct

Table 3. Results of investigations.

INVESTIGATIONS	NO. OF PATIENTS	PERCENTAGE
ESR	3	23
Anemia	4	30.8
Leucocytosis	1	7.7
Thrombocytopenia	-	-
ANA	2	15.4
Anti Ro, Anti La	1	7.7
Anti DNA	-	-
Serum complement	N	-
Nitroblue Toluidin T	N	-
BIOPSY H&E		
-Confirmatory	9	69.2
-Suggestive	4	30.8
DIF (Involved skin)		
-Confirmatory	7	53.8
-Suggestive	4	30.8
-Non specific	2	15.4
DIF (Uninvolved skin)	2	15.4

immunofluorescence examination of involved skin was confirmatory in 7 cases, and suggestive in 4 cases, but revealed only nonspecific findings in 2 cases in which the lesions were of less than 3 months duration. DIF of uninvolved skin showed deposition of IgM and C3 in a thready pattern in 2 cases that have been clinically diagnosed as SCLE.

Treatment

Table 4 shows the drugs used and the response to treatment. Only 3 patients responded well to conservative management of sun protection and local steroids. Chloroquine was poorly tolerated in 3 patients who had gastrointestinal side effects (2 cases), and changes in visual acuity (1 case). Etretinate (Tigasone) gave good results in 4 patients. Cheilitis necessitated withdrawal of the drug in 1 patient. In 3 patients not responding to any of the above, oral steroids gave good results.

During follow-up period (varying from 6 months to 5 years), one case developed signs of subacute cutaneous LE, such as psoriasiform lesions, arthralgia, ANA positivity, and anti Ro antibody positivity. One other case had digital scars, oral ulcers, ANA in low titter, and IgM deposition in uninvolved skin. However the latter did not satisfy the ARA criteria for diagnosis of SLE and was diagnosed as possible subacute cutaneous LE who needed further observation.

Discussion

Our series, though small, shows that cutaneous LE in children is not rare as previously reported.[1,2,5] As in adults, different subsets do occur.[4] Female predominance seen in adults is not striking in children.

Familial occurrence, (2:1 in our series) has been reported. Prystowsky et al [11] reported significant family history in 4%. Steagal et al[6] reported 25 families with Discoid LE or systemic LE. This may be important in view of reported HLA association in LE.[15,16]

Table 4. Results of Treatment.

Drug	No of patients	Responders	Non-Responders	Stopped treatment*
Local treatment	13	3	10	-
Chloroquin	10	6	4	3
Etritinate (Tigason)	7	4	2	1
Oral steroids	3	3	-	-

*Stopped treatment due to side effects.

While clinical picture in children is generally reported similar to that seen in adults, scarring was less and follicular pluging was not a prominent feature.

Syndromal association with Russell Silver Syndrome has not been previously reported. Routine investigations were not of much help in diagnosis. Anti Ro antibody was positive in one case of SCLE.

Routine H&E and DIF were helpful in confirming diagnosis. IgM deposition in uninvolved skin was found in 2 cases, suggesting systemic involvement.

Treatment in children presents many problems. Avoidance of sun is not always possible and many of the drugs are poorly tolerated. Chloroquin, a safe drug in adults, is poorly tolerated, and deaths have been reported after chloroquin ingestion.[14] However, Rasmusson [10] in a recent report, concluded that safety profile in children is equivalent to that in adults. Recommended dose of chloroquin is 3.5 mg/kg. [11]

Gastrointestinal side effects are common as in our series. Tigasnon was well tolerated in our patients, except in one case with lesion on lip that was exacerbated by Tigasone-induced cheilitis. Steroids were useful in resistant cases.

Data on progression of CDLE to SLE in children is limited.[13] Follow-up duration in our series has been variable. However, 2 patients developed evidence of systemic involvement as previously described, thus emphasizing the need for proper follow up.

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