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A Case of Bullous LE with Secondary Nephrotic Syndrome

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Abstract: Lupus erythematosus (LE) of the skin comprises an uncommon group of skin disorders, most often affecting young adult women (aged 20 to 50), but children, elderly and males may also be affected. Here we report a case of a 9 year old girl who presented with clinical features suggestive of non-minimal nephrotic syndrome with few vesicular skin lesions, hyperpigmentation and generalised lymphadenopathy which on further evaluation proved to be a case of bullous variety of cutaneous systemic lupus erythematosus.
Keywords: Cutaneous lupus, Non-minimal nephrotic syndrome, Vesicles, Bullous LE.

INTRODUCTION

Systemic Lupus Erythematosus (SLE), a condition often provoked by sun exposure [1] is classified as a autoimmune disease characterized by multisystem inflammation [2], and the presence of circulating autoantibodies directed against components of cell nuclei [1, 3].

SLE occurs in children and adults, disproportionately affecting women of reproductive age [4]. Lupus may affect any tissue or organ the most common being skin, joints, kidneys, blood forming cells, blood vessels and central nervous system [5].

In this article we describe the skin manifestations of lupus apart from the usual malar rash and discoid rash and emphasize on the fact that the clinical presentation of a simple case of nephrotic syndrome should not be dealt with ease and an underlying entity of SLE should always be suspected and managed especially in a female child.

CASE REPORT

A 9 year old female, the 1st order child of a non-consanguineous marriage was brought to the Out-patient department with complaints of generalised swelling all over the body since 2 weeks, decreased urine output and poor oral intake since 3 days, reddish discoloration of urine and stool for 2 days, vesicular skin lesions over the face since 2 days(Fig 1). The oedema started as facial puffiness which progressed over 2 weeks to involve the abdomen and legs. There was no history of recent pyoderma or sore throat, fever, seizures, abnormal behaviour, breathlessness, insect bite, yellowish discoloration of sclera or contact with tuberculosis. The mother gave insignificant past and family history. The antenatal, natal and postnatal periods were uneventful and the child was immunised for age.

On examination, the child was febrile(100F), pale, with generalised lymphadenopathy and bilateral pitting pedal oedema, vesicular skin lesions confined to the face, upper chest and upper back and healed skin lesions in the periumbilical area. There was hyperpigmentation and scaling all over the neck and upper trunk with multiple oral ulceration and mucositis. The pulse rate was 80 per minute, regular; blood pressure measured 140/90mm of Hg in the right upper limb in supine position. Abdomen was distended(Fig 2) and fluid thrill and shifting dullness was elicited. Chest was clear with normal vesicular breath sounds; cardiovascular and central nervous system examination was normal. Investigations revealed the Table-1.

Fundoscopic examination was normal and Chest X-ray revealed moderate pleural effusion on the right side. Ultrasonography of the abdomen revealed moderate ascites. A renal biopsy could not be done due to financial constraints. Skin biopsy revealed subepidermal blister with neutrophilic infiltrate(Fig 3) The child was admitted and managed with diuretics, 3 doses of methyl prednisolone, antibiotics to control infections, fluid intake and output chart was maintained and the severe hypertension which progressed during the hospital stay was controlled with 3 medications-calcium channel blocker (Nifedipine), angiotensin receptor blocker(Losartan)and a loop diuretic (Furosemide). Two packs of fresh frozen plasma was also transfused. A single dose of Cyclophosphamide

infusion was given in view of lupus nephritis and the child was discharged with oral prednisolone and hydroxychloroquine in divided doses and is on regular monthly follow up for monthly consecutive infusions of cyclophosphamide for 6 months followed by additional infusions every 3 months for 18 months. The skin lesions improved on Dapsone for 10 days(Fig 4).

Table 1: Laboratory investigations

Laboratory Parameter	Observed Values
Haemoglobin(gm/dl)	7.8
Total Leukocyte Count(/mm3)	3800
Differential count	N68L32M0E0
Total platelet count(/mm3)	98000
ESR(mm/hr)	56
CRP(mg/dl)	3.8
ANA	+VE
Anti-ds DNA	+VE
Serum C3,C4	Reduced
Urine	Albumin-3+,RBC-10,RBC cast+, Pus cells-3-5/hpf
Serum Albumin(g/dl)	2.2
Serum Cholesterol(mg/dl)	278
Serum Creatinine (mg/dl)	3.9

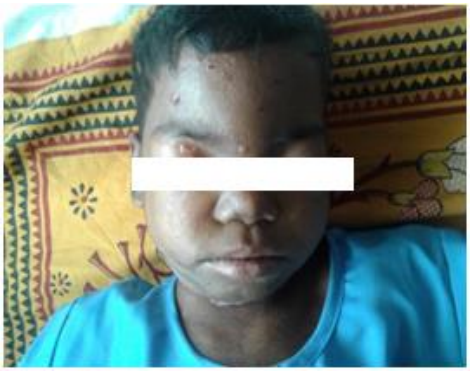

Fig. 1: Vesicular skin lesions on face

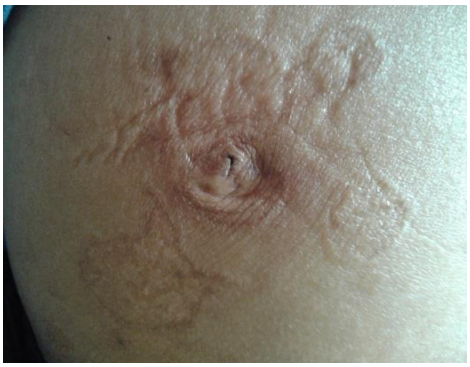

Fig. 2: Distended abdomen

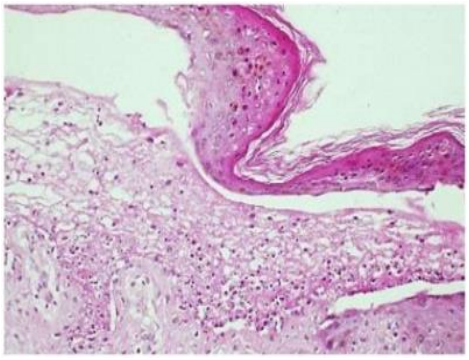

Fig. 3: Skin biopsy showing subepidermal blister with neutrophilic infiltrate

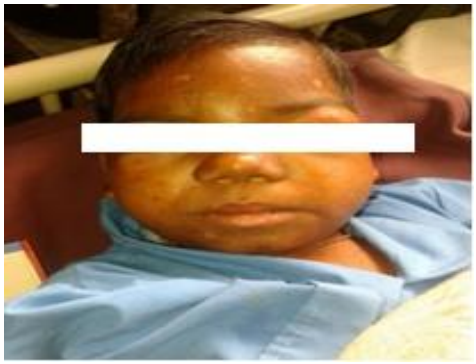

Fig. 4: Healed skin lesions

DISCUSSION

Although statistics reveals that 76% of patients develop skin lesions during the course of SLE, hardly 1% of these can be attributed to vesiculobullous lesions [2]. Among the vesiculo bullous diseases, the most distinctive presentation is that of an entity known as bullous SLE (BSLE). Young adolescent females are more affected with equal affection of both sun exposed and unexposed areas. The lesions are characterised by the appearance of blisters subepidermally on an erythematous base. Face, proximal extremities and upper part of trunk are most commonly affected. The differential diagnosis includes close mimickers such as linear IgA disease and bullous pemphigoid (BP). The inflammatory variety of EBA (epidermolysis bullosa acquisita) is also often confused with the entity of BSLE [7, 8].

Improved diagnostic criteria have paved way to better understanding of the underlying pathology of this cutaneous condition [7, 8]. 6 criteria were put forward by two renowned scientists, Gammon and Briggaman, based on which BSLE was classified into 2 types [9]:

Type 1: All 6 diagnostic criteria are met

Type 2: Only 1-4 criteria met. In type 2 the target is an undetermined antigen or any dermal antigen excluding type 7 collagen.

The 6 diagnostic criteria are as follows:

- Fulfilment of American Rheumatism Association (ARA) criteria for SLE;
- Acquired vesiculo bullous eruption on (but not limited to) sun-exposed skin;
- Histological evidence of sub epidermal blister with neutrophilic infiltrate;
- Presence of IgG, IgM, IgA, and C3 at the BMZ;
- Evidence of antibodies to type VII collagen;
- Co distribution of immunoglobulin deposits with anchoring fibrils/type VII collagen by immune electron microscopy [9].

Most of the reported cases have dramatically responded to dapsone, with cessation of new lesions in 1-2 days and healing within several days. Similar observation was noted in our case with dapsone (2 mg/kg/day).Relatively low doses (25-50 mg) have also been shown to be efficacious [8, 10].

The remaining types of cutaneous LE are enumerated below:

Discoid Lupus Erythematosus

It may be localised or widespread erythematous patches with scaling which leave behind a pigmented area and scar following healing. Discoid LE predominantly affects the face but can also involves the upper back, V of neck, and rarely the palms and/or soles (palmo plantar LE). Scarring alopecia is common. It can also affect the oral cavity causing ulcerations which are precancerous and can later give rise to squamous cell carcinoma [1, 11].

Subacute Lupus Erythematosus

This is a non scarring variety and mostly occurs on the light exposed areas causing a non itchy rash. It may be of 4 types such as annular, papulosquamous, vasculitic or nodular [1].

Lupus Erythematosus Tumidus

This dermal form of lupus attacks the sun exposed areas forming erythematous raised lesions sometimes ring shaped (annular). It clears in the cold months without scarring [1].

Lupus Profundus

Otherwise called as ‘lupus panniculitis’. It classically affects the subcutaneous fat of mainly the face causing firm nodular lesions which may last for months. Eventually the adipocytes are totally subjected to lipodystrophy causing unsightly scars [1].

Cutaneous Lupus Mucinosis

It is a rare presentation with dermal deposition of mucin. Sun exposed areas are mainly affected in the form of nodules, papules and plaques [1].

Chilblain Lupus Erythematosus

Cutaneous LE patients especially those living in cold climates, smokers may be affected with chilblains or Raynaud phenomenon [1].

Systemic Lupus Erythematosus

It affects several organs such as skin, joints and kidneys. Blood tests reveal circulating auto-antibodies. The clinical features of SLE are variable; may overlap with other diseases and conditions. SLE is more severe in smokers. Clinical features include Malar rash, mucosal erosions and ulceration, annular lesions, polycyclic lesions, hair loss, photosensitivity. Chronic variety has features that include Classic discoid lupus, Tumid lupus, Lupus panniculitis/ profundus, Mucosal lupus (lips, nose, mouth, genitals), Chilblain lupus etc. [1, 12].

Moving our focus to the second entity in our case; nephrotic syndrome is the kidney disease characterized by heavy proteinuria (3 g/day or more), hypoalbuminemia, hypercholesterolemia and edema. It can be divided two types: primary and secondary. Primary nephrotic syndrome is that in which only the kidney is affected; glomeruli are usually the target. While in secondary nephrotic syndrome is caused by systemic diseases. It may be due to several causes such as diabetes mellitus, juvenile idiopathic arthritis, systemic lupus erythematosus, familial Mediterranean fever and secondary renal amyloidosis. It is important to detect the secondary reason for the therapeutic point of view, as many times treatment of the secondary causes usually resolves the renal pathology [13].

In our case, the type of lupus nephritis could not be confirmed as renal biopsy, a mandatory test was not done. However in view of the persistent hematuria, proteinuria and features suggestive of nephrotic syndrome, a presumptive diagnosis of lupus nephritis of severe grade(3, 4 or 5) was made and intravenous infusions of cyclophosphamide was started to reduce the risk of progressive renal dysfunction.

CONCLUSION

Hence to conclude, in the diagnostic approach to adolescents and young adults, secondary causes of nephrotic syndrome such as Henoch-Schönlein nephritis, lupus nephritis, secondary renal amyloidosis, and infection-associated nephropathies, especially hepatitis B must first be excluded since finding an underlying aetiology alters the therapeutic options and disease course. The case has been presented for its typical and distinctive features which would prompt an astute physician to suspect and investigate the underlying systemic manifestations of SLE. Also, the dramatic response to dapsone has been highlighted which remains the drug of choice for the management of this cutaneous variety of lupus.

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