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An Adolescent Male Patient of SLE Presenting With Early Lupus Nephritis, Rheumatic Heart Disease and Iatrogenic Cushing Syndrome

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Abstract: *Systemic lupus erythematosus (SLE) is a multisystem autoimmune disorder. SLE is mimicked by other systemic disorders. Lupus nephritis is one of serious complications of SLE and is predictor of poor prognosis. Autoimmunity plays an important role in pathogenesis of SLE. We present herewith a rare case of young male who had presented with anasarca, palpitations, breathlessness, hypertension, proteinuria, with deranged renal function test, and was diagnosed as lupus nephritis on renal biopsy. The patient also had crossed renal ectopia, which makes this case very rare and one of its own kind in the whole world as yet. In addition, he also had a serious proliferative form of glomerular damage (ISN3 and ISN 4).*

Keywords: ***SLE, systemic lupus erythematosus, lupus nephritis, anasarca, autoimmune disease.***

Introduction

Systemic lupus erythematosus *(*SLE) is a rare, autoimmune disease which affects multiple systems. Usually SLE is a disease found more in adult females (female: male = 9:1). This difference is less marked in very young children and in the elderly (female: male =3:1). SLE may present acutely, but more commonly it is known to evolve over a period of time [1-2]. Majority of patients who are affected are between 20 years and 50 years of age. Female preponderance suggests hormonal influence, and sex hormone is known to modulate immune system and here the risk of SLE increases with use of estrogen containing contraceptives. SLE, especially the early disease, mimics several other systemic disorders. Therefore diagnosis is usually missed out in the initial stages, and more so if the patient happens to be a young teenager male. Lupus nephritis is one of serious complication of SLE, and is also a major predictor of poor prognosis [3-5].

Case report

A 15 year old male had presented with chief complains of decreased urine output since 10-15 days, joints pain (knee, wrist ,elbow and back), palpitations and breathlessness since 7-8 months which was initially on exertion, but from 15-20 days it is at rest as well. He had also complained of pinpoint and some large hemorrhages over trunk forearms, back, and legs since 20 days. In the history of presenting illness, patient gives a history of low grade fever with joint pain 1 year back, for which he took treatment from several doctors. His medical records confirmed that he was started on predinisolone at a dose of 60 mg daily per orally, 4 month back. He gained 16 kg weight (from 42kg to 68 kg) during this interval.

Figure 1- Moon face of patient

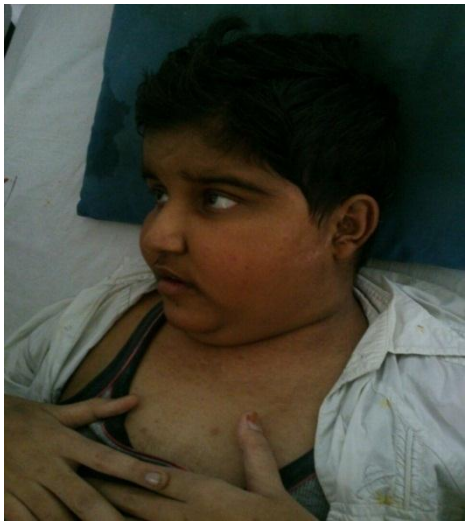
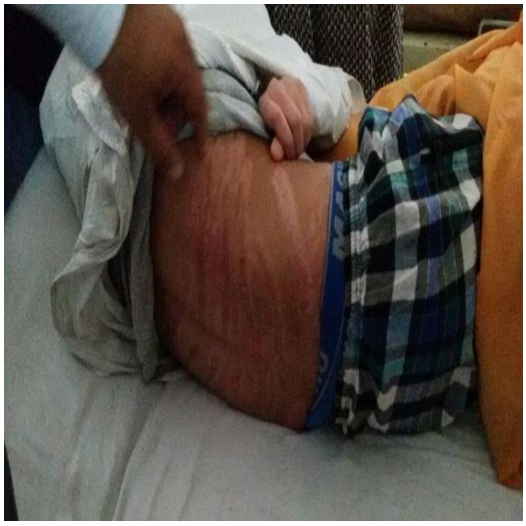


Figure 2- purple striae on back



On examination:

Here was a teenager male whose face was round like a moon (moon face). He had a conspicuous hump between the shoulders (buffalo hump). His abdomen was distended with associated peripheral thinning of limbs. Skin seemed to be thinned out and purple coloured striae were noticed over back, trunk, abdomen, and limbs, with pin point hemorrhages and ecchymotic patches over all limbs and trunk. On examination his blood pressure was 170/120 mm HG, pulse 128/min, regular, synchronous, and good volume. He had pallor, and deep pitting edema was detected over lower limbs up to knees. There was cyanosis of nails. However there was no icterus, clubbing, or lymphadenopathy. Suprasternal pulsations were noticed, with hyperdynamic apex beat and hyperdynamic precordium. On auscultation, a muffled first heart sound with loud second heart sound, and a loud rumbling pan-systolic murmur was detected over the whole precordium, which was best heard in aortic and neo-aortic area (Erb's area), with radiation to carotid, suprasternal notch, both shoulders, axilla, interscapular and infrascapular regions. On further auscultation, he also had crepts in axillary and infraaxillary and infrascapulary regions.

Investigations:

Hb-9.2, TLC-14.8*103, SGOT-18, SGPT-18, Alkaline phosphatase 59 ,bilirubin total-0.55, bilirubin direct 0.39, bilirubin indirect-0.16; Blood urea-240; Serum Creatinine-1.50; BUN-94.3; ASO titre - <200IU/ML; CRP <0.6; Serum Protein 5.89, S.Albumin-2.14, S.globulin-3.75, S.A/G ratio-0.6, Serum C3-99.08, SC4-6.25, LDH-507, Serum total cholesterol-201,S.HDL-39,SLDL-148,S.TG-216, CHOL :HDL 5.1:1, LDL:HDL3.7:1, SANA-6.284(POSITIVE), anti ds-DNA; HIV1&2 and HBsAg-ve. Routine examination of urine showed 20-25 pus cell per high power field. Albumin 4+, RBC-15-20 per HPF. Proteinuria >500mg/24hr. Ultrasonography of whole abdomen showed crossed renal ectopia (unilateral fused kidney on right side), and very small right renal calculi (concretions).

Needle biopsy of kidney had revealed:

a) Immune complex mediated, diffuse proliferative necrotizing crescentric glomerulonephritis (crescents in 9 of 14 glomeruli sampled-5 cellular and 4 cellular) displaying secondary segmental tuft sclerosis involving 7(50%) glomeruli.
b) DIF studies reveals full house immunostaining pattern with co-domjkhyuminant ,granular capillary wall and mesangial staining igG,less igM,igA and C3 and no evidence of light chain restriction.Extra glomerular staining for igG ,light chains and C1q is noted.
c) Features of patchy acute tubular injury with interstitial oedema. NO evidence of tubule interstitial chronicity is seen in the sampled cortical parenchyma.DIF findings in this case suggest renal involvement in SLE/systemic collagen vascular disease.

2D-Echo confirmed LV thickness to be normal. Rheumatic heart disease was ruled out. Mitral valve cusps showed AML and PML were mildly thickened with moderate MR, and moderate AR. Associated finding was a mild TR with severe PAH, with PASP 61 mmHg. The LVEF was 45%. There was no aortic stenosis or regurgitation.
This patient was hospitalized in intensive care, and ICU protocol was strictly followed. Management strategies revolved around managing hypertension with anti-hypertensives; management of Lupus nephritis with predinisolone 40 mg / day, hydroxychloroquine, along with mycophenolate mofetil 500mg 12hrly. Fluid intake and output was recorded carefully, and 4-hourly record of vital parameters was strictly maintained. After a short stay in ICU, this patient was shifted to the general ward, and was discharged once the blood pressure was controlled and steroids tapered off to necessary minimum. At the time of discharge from hospital, he was also advised to have a regular follow up with a nephrologist.

Discussion:

This is a very rare case of SLE in a young male teenager, who had presented with multiple organ involvement and renal ectopia. Patient had gained 16 kg weight, had anasarca, and was hypertensive that was suggestive of active nephritis. He had several lab abnormalities, and his urine showed microscopic hematuria and albuminuria 4+. Renal biopsy had revealed the patient having a dangerous proliferative form of glomerular damage (ISN3 and ISN 4). In lupus nephritis there is usually microscopic hematuria, hypertension, proteinuria (>500mg/24hr), and extensive peripheral edema that is suggestive of diffuse proliferative nephritis. Our patient also had fever, oral ulcers, joint swelling (arthritis), as well as petechiae and purpura which are more common with focal and diffuse proliferative lupus nephritis. Nephritis is most serious manifestation of SLE. Since nephritis is asymptomatic in most lupus patients, urine analysis should be ordered in any person suspected of having SLE. Early renal function tests in case of SLE should not be omitted in order to detect any renal involvement, because early detection and treatment can improve clinical outcome. Renal biopsy should be considered in patients of SLE having clinical or lab evidence of nephritis.

The principal goal of therapy in lupus nephritis is to normalize renal function, and to prevent progressive loss of renal function. DPGN if left untreated can lead to ESRD within 2 year of diagnosis. Therefore main goal of therapy is to improve renal function, and to prevent the progression to ESRD. All patients with lupus nephritis should receive

therapy with hydroxychloroquine, unless contraindicated. Most patients with episode of severe lupus require many years of maintenance with low doses steroids. It becomes a standard practice to initiate therapy for active potentially life threatening SLE with high dose IV glucocorticoids. In specific situations there might be a role of cytotoxic agents along with glucocorticoids, and trials are underway. In that cyclophosphamide seems to be an acceptable choice for induction of improvement in severely ill patients. In patients whose ISN Grade 3 or grade 4 disease, early treatment with combination of glucocorticoids and cyclophosphamide has exhibited a reduction in the rate of progression to ESRD and in improving survival. Survival is better in people with DPGN treated with high dose daily glucocorticoids (40-60 for 4-6 months). Patients with ISN/RPS 1 and 2 do not require immunosuppressive therapy. Administration of ACE inhibitor or ARBs is required if proteinuria reaches or exceed >0.5gram/24 hours. Blood pressure should be maintained at or below 130/80 mmHg. Patients with ESRD will require dialysis, and are good candidate for renal transplantation.

Conclusion:

All patients with Cushinoid features and deranged renal function test should be properly investigated. Lupus nephritis should be suspected, which needs to be followed up by a renal biopsy. Appropriate treatment and care should be started at earliest once the diagnosis is confirmed.

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