Case Report

A rare case of interstitial lung disease as a presenting feature in Juvenile Systemic Lupus Erythematosus responding to Mycophenolate Mofetil

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ABSTRACT

Background: Symptomatic interstitial lung disease (ILD) is rare in systemic lupus erythematosus (SLE) and there is no established treatment for it. Data is limited to adult lupus patients.

Aim: The aim of this case report is to highlight that ILD can be as a presenting feature in juvenile lupus and Mycophenolate Mofetil can be a promising immunosuppressive agent for lupus associated juvenile ILD.

Case Report: We report a case of a 15-year old girl with polyarthralgia, intermittent fever, weight loss, dry cough and progressive dyspnea of one year duration. Investigations and high resolution CT scan revealed SLE associated progressive ILD which responded well to mycophenolate mofetil.

Conclusion: The highlighting point of this case is that pediatricians and rheumatologists should bear in mind that ILD may be part of the initial manifestation of SLE. Mycophenolate mofetil may be a promising therapeutic choice in juvenile SLE-associated ILD.

Keywords: Juvenile systemic lupus erythematosus, interstitial lung disease, mycophenolate mofetil

INTRODUCTION

Systemic lupus erythematosus (SLE) is a multisystem connective tissue disorder with diverse clinical presentations. The disease has varied organ involvement with cutaneous, muskuloskeletal, renal, neurological, hematological and pulmonary manifestations. Symptomatic Interstitial lung disease (ILD) is not so common in SLE and even much rarer to be the initial manifestation of SLE without any other organ involvement. Limited data exist with regards to association of ILD in juvenile SLE. We report a unique case of subacute progressive ILD as the presenting manifestation of a 15-year old girl with SLE who responded to mycophenolate mofetil (MMF).

CASE DESCRIPTION:

A 15-year-old girl presented with polyarthralgia, intermittent fever, weight loss, dry cough and progressive dyspnea of one year duration. There was no history of skin rash, oral ulcers, skin tightening, sicca symptoms, raynaud's phenomenon or sclerodactyly. Examination revealed wasting, pallor, tachycardia, tachypnea, with oxygen saturation (SpO2) of 90% at room air, diffuse bilateral crepitations with normal heart sounds. Investigations revealed anemia, leucopenia, thrombocytopenia (Hb=8.2 gm/dl, WBC= WBC=2300/cmm; Neutrophils =56%, Lymphocyctes =44%, platelet count=1,20000/cmm) with raised acute phase reactants (erythrocyte sedimentation rate- 76 mm/hr and C-reactive protein -14 mg/L). Direct coomb's test (DCT), mantoux test, and blood cultures were negative. X-ray chest revealed diffuse reticulo-nodular shadows with no hilar lymphadenopathy in both the lung fields. The serum electrolytes, liver and renal function, urine routine, 2D-ECHO, Ultrasonography abdomen, serum Angiotensin-converting enzyme (ACE) levels, schirmer's test, ophthalmology examination were all within normal limits. Immunological examination showed positive anti-nuclear antibody [ANA titres 1:640, (homogenous pattern)], positive anti-double stranded DNA (anti ds-DNA)

and anti-Ro/SSA, low serum complement 3 (C3- 40 mg/dl; range 82-173), low complement 4 (C4- 7.2 mg/dl; range: 13-46 mg/dl), negative Rheumatoid factor (RF) and antiphospholipid antibodies. High resolution CT scan (HRCT) chest showed symmetrical fine reticular opacities with relative subpleural sparing, traction bronchiectasis, mild ground glass opacities in bilateral lungs suggestive of ILD -predominantly non-specific interstitial pneumonitis (NSIP) pattern (Figure 1). Patient refused consent for lung biopsy. Based on constitutional symptoms, polyarthralgia, leucopenia, thrombocytopenia, high ESR, ANA and anti ds-DNA positivity, low complements and HRCT findings; a diagnosis of SLE with ILD-NSIP was given. She was initiated on monthly cyclophosphamide (CYC) at 500 mg/m2 and oral steroids at 1mg/kg/day. A total of 2 doses of CYC were given following which she started developing significant nausea, vomiting, diarrhea and subsequent hospitalization for sepsis. Our patient refused consent for subsequent CYC treatment. Hence, MMF was started at 1200mg/m2/day along with oral steroids. The patient reported marked improvement in dyspnea, and constitutional symptoms which was noticeable between 6-8 weeks after the initiation of MMF and steadily improved. At the end of six months of MMF treatment, her counts and acute phase reactants have normalized and repeat HRCT chest show improvement in the ground-glass opacities and we were able to taper the steroids to 0.5 mg/kg/day.

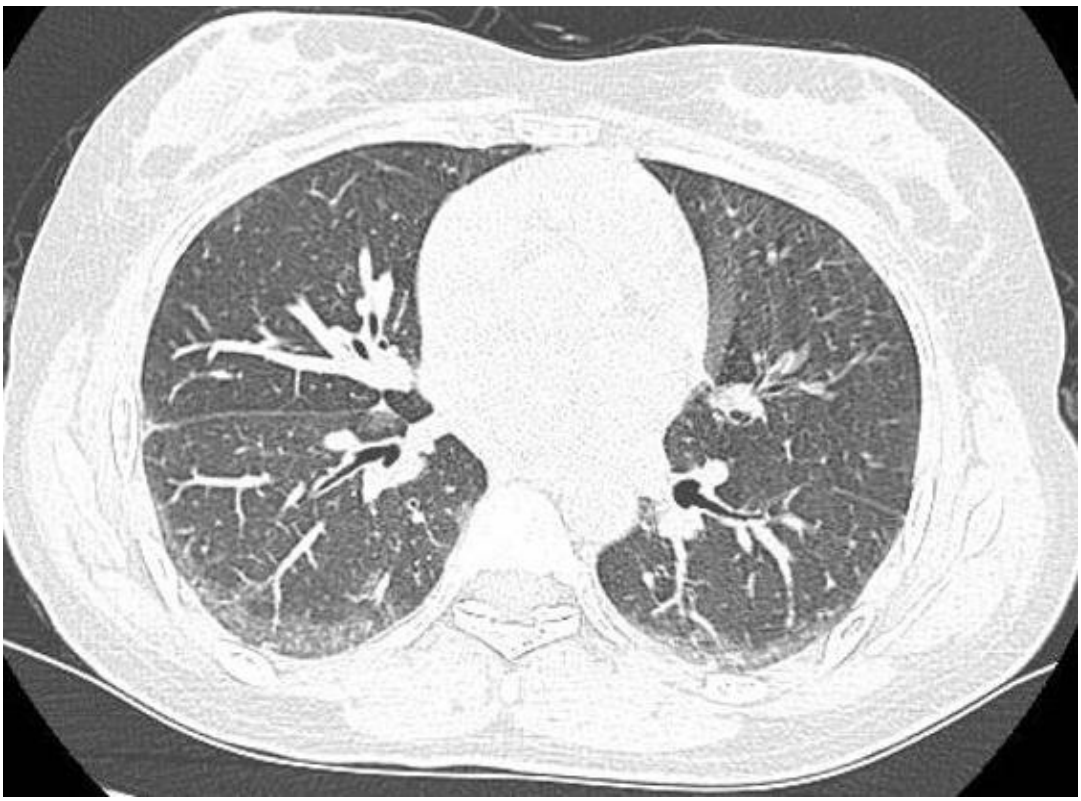

Figure 1: High-resolution computed tomography of the chest reveals bilaterally symmetrical fine reticular opacities with relative subpleural sparing, traction bronchiectasis, mild ground glass opacities (red arrow) with lower lobe predominance suggestive of nonspecific interstitial pneumonia

Author	Age and Gender of patient(s)	Treatment
Hanata N et al; [7] 2020	69 years/female	Corticosteroids, Azathioprine, MMF
Chakrabarti S et al; [8] 2015	40 years/male	Corticosteroids, Cyclophosphamide
Mondal S et al., [9] 2014	40 years/female	Immunosuppressant details not mentioned
Kumar A et al; [10] 2013	40 years/female	Not mentioned
Esmaeilbeigi F et al; [11] 2012	23 years/male	Corticosteroids, Azathioprine

Table 1- Case reports of Interstitial Lung Disease as Presenting Feature of SLE

DISCUSSION:

Pleuro-pulmonary complications of SLE are varied and include acute lupus pneumonitis, diaphragmatic dysfunction, shrinking lung syndrome, cavitating pulmonary nodules, pulmonary hypertension, pulmonary vasculitis, pulmonary embolism, alveolar haemorrhage, chronic interstitial pneumonitis, bronchiolitis obliterans and opportunistic pulmonary infections or drug toxicity from immunosuppressive therapy [1]. Symptomatic ILD is rare in patients with SLE, affecting about 3% of adult SLE [2]. Herein, we probably report the first case from India with regards to ILD and juvenile lupus (Table 1). It has been reported that Anti-Ro/SSA which was positive in our case is a risk factor for development of ILD in SLE [3]. There are no established treatment guidelines for ILD associated with SLE. Empirically, high-dose steroids and immunosuppressive drugs such as Azathioprine and intravenous cyclophosphamide (CYC) are used to treat SLE-associated ILD [4]. MMF has shown good efficacy in myositis and scleroderma-related ILD [5, 6]. MMF is widely used for severe lupus nephritis and other severe manifestations of SLE, whereas its efficacy against SLE-associated ILD has not been validated. Our patient was not able to tolerate CYC and fortunately, her ILD was improved on MMF treatment. Similar cases have been reported but data is limited to adult lupus patients [7].

CONCLUSION

Our case is unique as symptomatic ILD as a part of the presenting feature in juvenile lupus is extremely rare. Further, we highlight that MMF can be a promising immunosuppressive agent for juvenile SLE-associated ILD.

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