SLE in a Male Patient Presented Initially as Rowell’s Syndrome

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Abstract
A 22 year old male Indian patient presented with high grade fever, multiple joint pain, low back pain, generalized body ache since 6 months and erythematous pruritic rashes and atypical annular target like lesions over face, arm, leg and back and ulcers on hard palate and buccal mucosa for 2 months. Laboratory investigations showed a speckled pattern anti-nuclear antibody with a titer >1:160 and positive SS-A, dsDNA auto-antibodies and Rheumatoid factor. Diagnosis of Rowell’s syndrome was made based on clinical and laboratory finding and the patient was treated with oral prednisolone (50 mg/day), hydroxychloroquine (200 mg q12h) and pulse cyclophosphamide (700 mg) chemotherapy. Majority of skin lesions and oral ulcerations subsided after 4 weeks of therapy. Till date only 11 male patients out of the total 71 cases of Rowell’s syndrome were reported in the world’s literature.

Introduction
SLE (Systemic lupus erythematosus) is a systemic autoimmune disorder which may present with several skin manifestations like malar rash, discoid rash, photosensitivity, subcutaneous lupus erythematosus, oral ulceration etc. But SLE patient presenting with erythema multiforme like skin lesions is very unusual. Rowell first described this association in 1963. Rowell’s syndrome is a rare presentation of lupus erythematosus (LE) with erythema multiforme like lesions associated with positive antinuclear antibody (ANA), anti-La (SS-B)/anti-Ro (SS-A) antibodies and rheumatoid factor.¹ Till date only 11 male patients out of the total 71 cases of Rowell’s syndrome were reported in the world’s literature.² Here we describe a male patient whose clinical picture is consistent with so-called Rowell’s syndrome.

Case Report
A 22 year old non-smoker, non-alcoholic unmarried male Indian patient without any history of high risk sexual behavior was admitted to our hospital with high grade fever, multiple joint pain, low back pain, and generalized body ache since 6 months and erythematous pruritic rashes throughout the trunk, extremities and face since 2 months (Figures 1 and 2). Sometimes fever was associated with chill and rigor without burning sensation during micturition. Patient also had oral ulceration during the course (Figure 3). There was no similar type of past illness or history of prolonged medication. There was no history suggestive of chilblain too.

On clinical examination patient was normotensive, had mild pallor, multiple soft, non-tender, non-matted discrete cervical, axillary and inguinal lymph nodes. Skin examination revealed well-defined, erythematous papules and plaques, mostly annular, some with scales that coalesced into large, confluent lesions which were present on the nose, malar areas, ears, back, arms, legs, palms and soles. Patient had few atypical annular target like or erythema multiforme-like lesions over face, arm, leg and back. Patient also had ulcers with surrounding erythema on hard palate and buccal mucosa. Musculo-skeletal system examination revealed swollen and tender small joints of hands, wrists and knees and tenderness over arms and thighs with proximal muscles weakness.

Laboratory investigations showed mild anemia (Hb-10.4 gm %). Liver function test was normal except elevated enzymes: ALT - 246 IU/L, AST - 469 IU/L and Alkaline Phosphatase - 514 IU/L. Anti-nuclear antibody test was found to be positive - titer was >1:160 with a speckled pattern. Subsequent investigations revealed positive dsDNA (++, signal intensity-30) and SS-A (+++, signal intensity-87) auto-antibodies. Rheumatoid factor was also positive. C3 level was decreased (52 mg/dl) and C4 level was normal. A comprehensive metabolic
Discussion

The first described association between LE and erythema multiforme was made by Scholtz in 1922. In 1963, Rowell et al. reported a new syndrome characterized by LE, erythema multiforme-like lesions, a positive test for rheumatoid factor, speckled ANA and a saline extract of human tissue (anti-SJ/T) which is now regarded as similar to Ro (SSA).3,5 Rowell defined this association as a distinct entity upon discovering different clinical and immunologic findings in four patients during his study including 120 discoid lupus erythematosus (DLE) patients. Since the first report of Rowell’s syndrome, a total 71 cases have been reported in the literature.2 However, a recent review demonstrated that most of the reported cases did not fulfill all the diagnostic criteria of Rowell’s original description, especially the presence of RF and anti-La antibody.2 In 2000 Zeitouni et al. redefined Rowell’s syndrome with major and minor criteria.3 Major criteria included any form of LE, Erythema multiforme like lesions (with/without involvement of the mucous membranes) and speckled pattern of ANA. Minor criteria were chilblains, positive rheumatoid factor and anti-Ro or anti-La antibody. All three major criteria and at least one minor criterion are required for the diagnosis of Rowell’s syndrome.3 A review of 18 case reports of Rowell’s syndrome between 1963 and 2000 showed that the speckled ANA pattern was the most consistent feature of Rowell’s syndrome and was described in about 88 percent of the cases, whereas rheumatoid factor was the least preserved feature and is present in only 41 percent.4 Anti-Ro/La antibodies were detected in 53 percent of the cases.4 Although chilblains had been described in all four of Rowell’s original cases, this feature was found in only five of the 15 cases reported between 1982 and 2008.4 A more clinically relevant question and resulting controversy concern whether Rowell’s syndrome truly merits distinction as a unique clinical entity.3 The speckled ANA pattern, which correlates with antibodies to various ribonucleoproteins, is not unique to Rowell’s syndrome; it is also positive in SLE, mixed connective-tissue disease, and scleroderma.4,5 Similarly, anti-Ro/La antibodies can be detected in SCLE (70%), Sjögren syndrome (80%), SLE (20–60%), rheumatoid arthritis, and scleroderma; they are strongly associated with photosensitivity and vasculitis in SLE.4,5 These two antibodies contribute to the formation of the ANA speckled pattern. Therefore, the concomitant appearance of this pattern with positive anti-Ro or anti-La antibody can be expected. Also, rheumatoid factor positivity can occur in DLE (17%), SLE (40%), scleroderma (40%), SCLE, and Sjögren syndrome.4 In view of the lack of specific features Kuhn et al. suggested that Rowell syndrome is probably not a distinct entity and is now widely considered to be a variant of SCLE.6 The therapeutic regimen, responses, and prognosis in Rowell’s syndrome are similar to those of SLE or DLE that occur alone.4 Most of the reported cases showed good responses to mid-to-high doses of prednisone with azathioprine or antimalarials, such as chloroquine or hydroxychloroquine.1 However, male patients with these characteristic clinical and immunological features very rarely reported in the world literature and here we have described a male patient whose clinical picture was consistent with so-called Rowell’s syndrome.

Conclusion

SLE is rare in male and initial presentation with erythema multiforme is even more rare. So high degree of suspicion is clue to the diagnosis of SLE in this situation. We like to report the case to make the physicians aware about the possibility of SLE in a case of erythema multiforme with systemic features like fever and joint pain.

References