IRA E-BULLETIN : ISSUE 2 | APRIL 2020

DISEASE IN DISGUISE

A 40-year male presented to us with complaints of inflammatory polyarthritis of 1.5 years duration and recurrent oral ulcerations, alopecia and photosensitive facial rash for the last 6 months. Initially he had been treated with methotrexate for possible rheumatoid arthritis and with the development of the other symptoms for the last 6 months, low dose steroids and HCQs were added by a local practitioner for possible SLE. For anemia with Hb of 6.6, he had also received 2 units of packed cells. He was referred to us and examination showed facial rash, oral ulcers, no arthritis, and mild proximal muscle weakness. Investigations revealed anemia with Hb 9.2, raised liver enzymes with AST/ALT 302/125 and ALP 524. CPK, however, was normal but EMG showed a myopathic pattern. Renal functions were normal. chest X-Ray, ECG, ECHO were normal. Immunological workup showed ANA 3+ nucleolar, ANA profile showed Sm and Ro 52 positivity, low complements with C3 of 57. The renal evaluation showed normal urine routine and 24-hour urinary protein and DCT was positive. He was diagnosed as SLE based on the above presentation and reports. He was started on steroid at 1mg/kg for myositis, HCQs, and oral methotrexate and discharged.

He was advised to review after 2 weeks but was lost to follow up and came after 6 weeks with complaints of ulcers over bilateral knuckles and dorsum of right foot, Raynaud's phenomenon with digital gangrene and mild cough. He was taking prednisolone but at a suboptimal dose. On examination, he had pallor, digital infarcts in both ring fingers and a 3x4cm ulcer over the dorsum of the right foot. There was no history of preceding trauma to the foot.

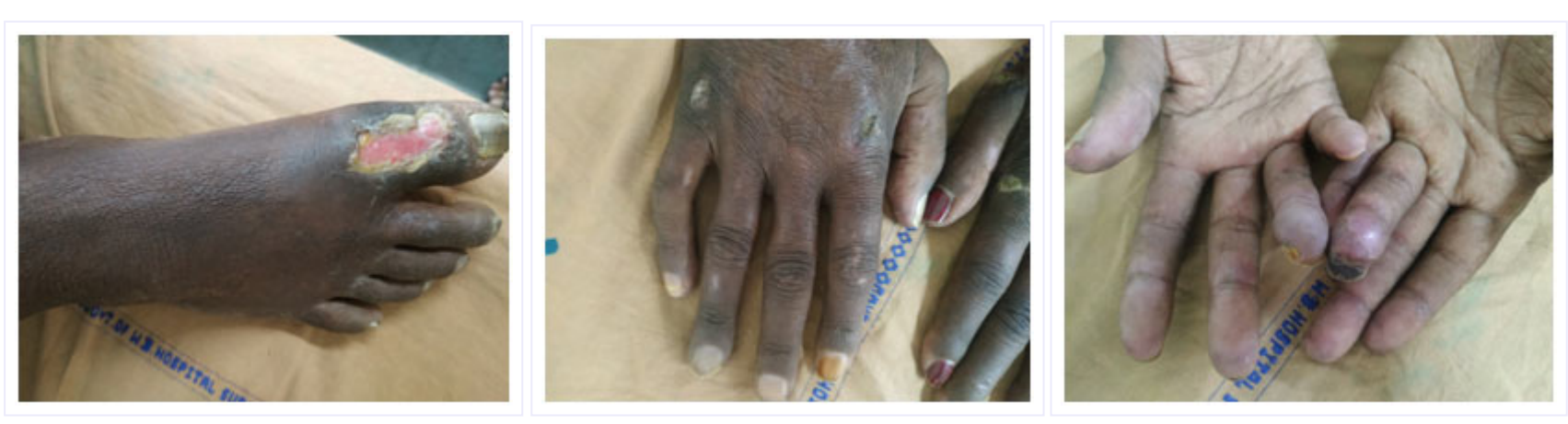

Figure 1 - Ulcer over knuckle, and dorsum of the right foot. Digital infarct over both ring fingers.

Respiratory system examination revealed a few crepitations bilaterally and muscle power was normal this time. Investigations showed anemia, normal liver enzymes, CPK, renal functions. Complements were near normal with C3/C4- 82/15. Chest X-ray revealed bilateral reticular changes that were not seen on the previous X-Ray. HRCT chest showed features of ground glassing suggestive of ILD. (Figure 2)

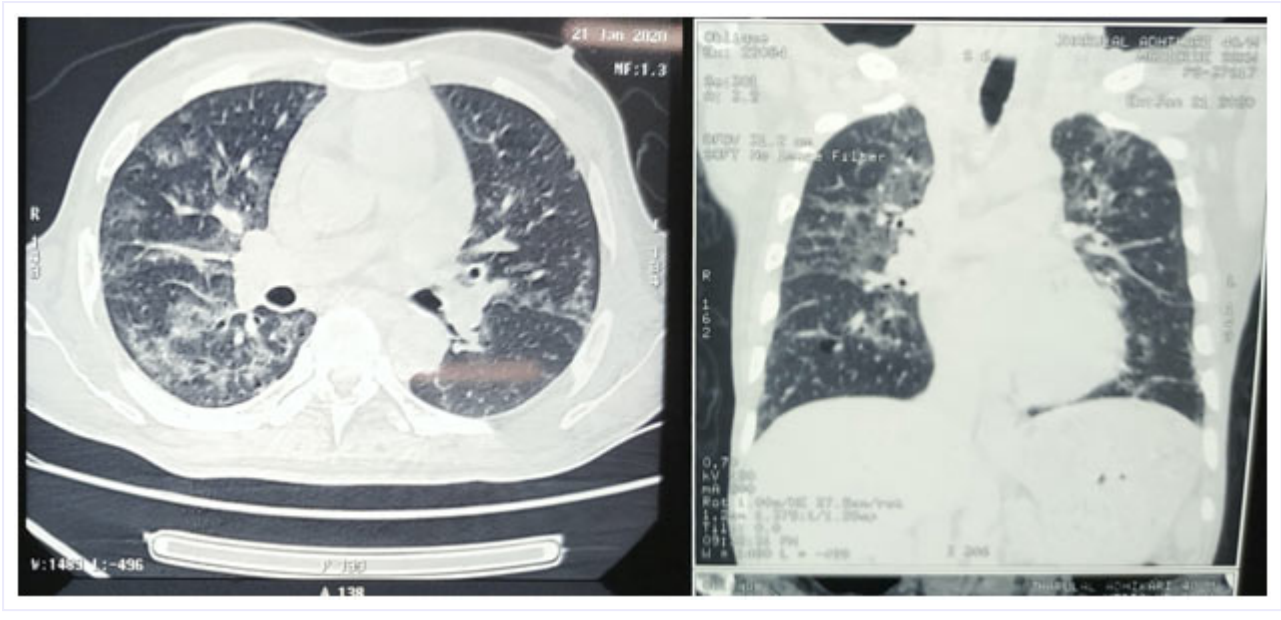

Figure 2 - HRCT chest showed features of ground glassing and reticulations suggestive of ILD.

PFT showed an FVC of 44%. ECHO was repeated which showed mild PR, moderate TR with PASP of 55mm suggestive of PAH. With the findings of cutaneous ulcers, amyopathic/hypomyopathic dermatomyositis, rapidly progressive ILD and facial rash, the diagnosis was revised to Dermatomyositis- probably anti MDA5 related. Lupus- dermatomyositis overlap was also a possibility in view of low complements and Smith positivity. Myositis profile was negative but anti MDA5 could not be checked due to financial constraints. He received two doses of Rituximab 1gm 15 days apart for the ILD. He has come for follow up once after that and is doing better with no muscle weakness or respiratory symptoms and resolving ulcers.

This case was of a 40-year male who initially presented with polyarthritis, oral ulcers, rash, DCT positive anemia, and myositis with low complements and positive ANA and was treated as SLE. Soon after, he came with cutaneous ulcers and ILD which rapidly progressed to cause PAH. The diagnosis was then revised to dermatomyositis, treatment of which was initiated with Rituximab and steroids.

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