Crusted Scabies in Systemic Sclerosis with Plasma Cell Dyscrasia

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Sir,

A 45-year-old female presented with generalized scaling, tightening of skin, difficulty in swallowing, breathlessness, joint pain and deformities, with severe itching, and crusted scaly lesions over scalp and all over the body [Figures 1a, b and 2a] since 3 months.

On cutaneous examination, she had salt and pepper pigmentation [Figure 2b], pinched nose, fish mouth, and facial lines of expression were smoothed out suggesting classical systemic sclerosis [Figure 3a]. There was severe tightening of skin leading to flexion contractures at finger and toe joints as well as restricted movement of elbow, shoulder, and knee joints. Grade 2 microstomia was present and chest expansion was reduced to 1 cm at the level of T4. The patient did not have Raynaud's phenomenon. In addition, she had foul smelling thick asbestos like scaling all over the trunk and extremities. Intermittent areas of oozing crusting and echthyma like pyodermas were also noted. Her other family members were also affected with scabies.

The diagnosis of crusted scabies was confirmed on potassium hydroxide (KOH) skin scrape which revealed abundant mites and eggs of Sarcoptes scabiei [Figure 3b]. Skin biopsy was taken from forearm which showed dermal sclerosis characterized by thick hyalinized collagen fibers arranged parallel to epidermis. The blood investigation revealed low hemoglobin 7.7 g/dl raised erythrocyte sedimentation rate 80 mm at the end of 1h, total leukocyte count was 19700/cumm. The rheumatoid factor was positive. Antinuclear antibody (ANA) was negative. Urinalysis revealed albuminuria. Serum calcium level and renal function tests were normal. Chest X-ray showed fibrosis at right basal zone. High-resolution computed tomography (HRCT) showed interstitial lung fibrosis. Pulmonary function test revealed findings consistent with restrictive lung disease, barium swallow showed typical rat tail appearance, and echocardiography demonstrated features consistent with pulmonary hypertension. According to European League Against Rheumatism (EULAR) criteria her score was 19.The patient's human immunodeficiency virus (HIV) and mental status were normal. Serum protein electrophoresis revealed hypergammaglobulinemia and M band in gamma-globulin region. Immunofixation electrophoresis study showed immunoglobulin G (IgG) kappa positive. Bone marrow aspiration study demonstrated 15% plasma cells, Dutcher bodies, and Mott cells.

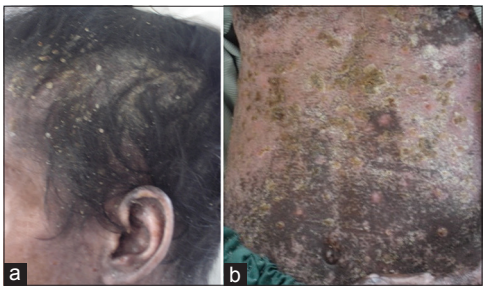

Figure 1: (a) Scaly plaques over scalp. (b) Sclerotic abdominal skin with scale-crusts

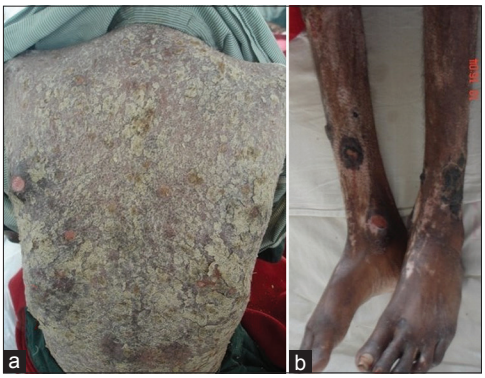

Figure 2: (a) Crusted scaly plaques over back. (b) Salt and paper pigmentation

How to cite this article: Nikam BP, Patil SJ, Jamale V, Kale M. Crusted scabies in systemic sclerosis with plasma cell dyscrasia. Indian Dermatol Online J 2018;9:197-8.

Received: July 2017. **Accepted:** October 2017.

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Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.IDOJ_212_17

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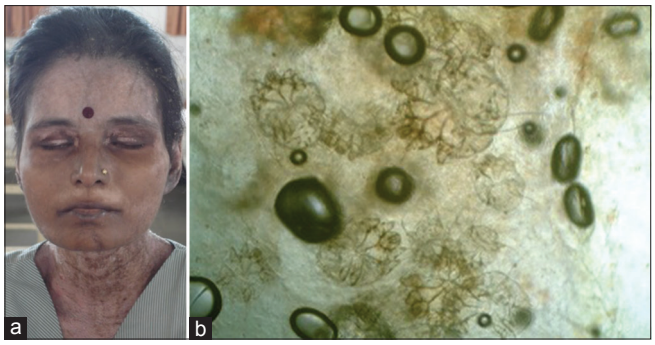

Figure 3a: Pinched nose and fish mouth. (b) Numerous mites and eggs

The patient and her family were treated with two doses of oral ivermectin 12 mg at 2 weeks interval and topical 5% permethrin cream every third day for 3 weeks with good resolution of scabies. The patient did not receive any systemic steroids and immunosuppressive agents, and was referred to oncology department for further treatment.

In our case, crusted scabies and diffuse scleroderma were easily diagnosed. Previous literature shows that crusted scabies is also seen with underlying malignancies like multiple myeloma.[1,2] Hence, patient was subjected to serum electrophoresis and bone marrow biopsy which diagnosed the hidden plasma cell dyscrasia. Thus, the triad of etiopathogenesis gets completed. In addition to the physical morbidities, immunosuppressed state produced by both diffuse systemic sclerosis and multiple myeloma led to the development of crusted scabies in the present case.

We have reported this case, as crusted scabies associated with systemic sclerosis is rare. The association of systemic sclerosis with multiple myeloma although rare, is well described in literature. Finding these three conditions together has not been reported in literature.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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