Pictorial CME

Morphea and Systemic Sclerosis Coexistant : An Uncommon Association

Tathagata Ghosh[1], Kirtiman Mandal[2], Pranabananda Pal[2], Nandini Chatterjee[3]

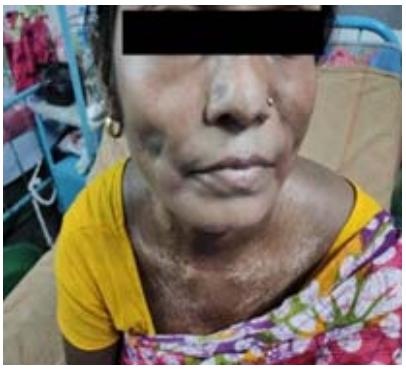

Fig 1 — It shows a depressed scar like lesion at right cheek with hyperpigmentaion of skin over it and salt and pepper like skin pigmentation over the front of chest

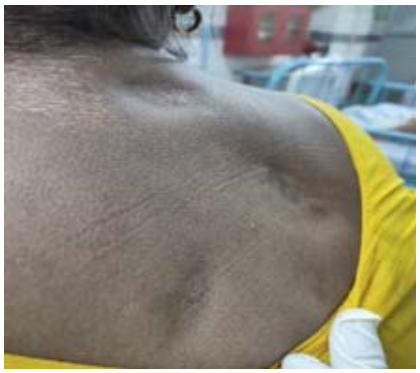

Fig 2 — It shows three discrete depressed scar like lesions at the back of chest with pigmentary changes within two of them

Fig 3 — It shows digital infarcts on left middle finger , ring finger and little finger

A 51 year old female, nondiabetic, hypertensive patient presented with complaints of gradually progressive skin tightening for last 1 year which initially started on fingers and face and gradually involved both upper limbs , neck, upper chest and back along with areas of skin colour changes. She also noted some areas of depressions over right and left cheek and upper back since last 8 months. There was also history of exertional dyspnea for last 6 months. No history suggestive of Raynaud's phenomenon was present.

On examination, patient had pallor, digital pitting scar over fingertips of both hands, salt and pepper like skin over neck, front and back of upper part of chest wall and dorsum both hands. There were 7 sclerotic depressed lesions over both cheeks and back with salt and pepper like changes over some of them. Systemic examination revealed velcro crepitations in both lung fields on chest auscultation and a loud P2 in auscultation of heart.

Investigations showed raised inflammatory markers, Scl70 and ANA positivity, NSIP pattern in HRCT Thorax, moderate PAH in 2D Echocardiography. Diagnosis of Systemic Sclerosis was made in association with Generalized Morphea complicated by Interstitial Lung Disease and Pulmonary Arterial Hypertension. Patient was treated with low dose steroid, PDE 5 inhibitor and planned for monthly injection Cyclophosphamide for 6 months and being followed up.

Morphea, also called as localised scleroderma is a dermatological condition characterized by excessive collagen deposition resulting in thickening of dermis, subcutaneous tissue or may be both. It is caused due to overproduction of collagen mainly type I and III in the affected tissues by fibroblasts probably activated by some immunologic dysregulation. According to clinical features and depth of tissue involvement, Morphea is classified into five subtypes — 1. Circumscribed/Plaque (3 or less discrete lesions) ; 2. Generalized (4 or more discrete lesions); 3. Linear; 4. Mixed and 5. Pansclerotic. Morphea may occur due to infections like Borreliosis, autoimmune causes like scleroderma, radiation therapy, drug induced like bleomycin or D-penicillamine , vaccination like BCG and Tetanus , trauma and chemical exposure. It has a female preponderance. The generalized variant do not have Raynaud's phenomenon or visceral involvement but it is usually linked to a concomitant autoimmune disorder with positive ANA bearing poorer prognosis[1]. Systemic sclerosis is known for its features like Raynaud's phenomenon and widespread visceral involvement. The coexistence of morphea and systemic sclerosis is a very uncommon entity described in 3.2-6.7% cases[2].

This case emphasizes the importance of detailed clinical examination as well as high index of suspicion to detect this type of association between morphea and systemic sclerosis for early intervention and better prognosis.

Department of General Medicine, IPGME&R and SSKM Hospital, Kolkata 700020
[1]MD Student and Corresponding author
[2]MD Student
[3]MD (Gen Medicine), FICP, Professor

References

1 Sapra A, Dix R, Bhandari P, Mohammed A, Ranjit E — A Case of Extensive Debilitating Generalized Morphea. *Cureus* 2020; **12(5):** e 8117doi10.7759/cureus8117.
2 Dilia G, Michele C, Emanuele C, Amelia S, Federica L, Clodoveo F — From localized scleroderma to systemic sclerosis: co-existence or possible evolution. *Dermatol Research & Practice* 2018; 1-5.