Salt and Pepper Pigmentation – Skin Manifestation of Systemic Sclerosis

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A 50 year old male presented with progressive difficulty in swallowing both liquid and solid food with no history of Raynaud’s phenomenon. A general examination revealed skin changes in the form of thickening, hyperpigmentation and tightening of skin of fingers, hand, forearm and legs. The patient had painless skin induration over the legs, forearm and hand. Salt and pepper pigmentation was seen on the upper back (Figure 1a), over mastoid process (Figure 1b) and the concha of pinna (Figure 1c). Anti-Scl 70 was positive. Anti-centromere antibodies were negative. Pulmonary function testing (PFT) revealed very severe restrictive lung disease. Barium swallow study was normal. Despite being advised to undergo oesophageal manometry test in view of dysphagia, patient was not willing for the same. Diagnosis of systemic sclerosis was made.

Systemic sclerosis is a disease in which extensive fibrosis, vascular alterations and autoantibodies against various cellular antigens being the principal features with a female to male ratio of 4:1. Skin pigmentation changes among other features of skin involvement include a salt-and-pepper appearance due to diffuse hyperpigmentation with sparing of the perifollicular areas. This may be due to the richer capillary network that may warm the perifollicular skin and preserve melanogenesis producing the perifollicular pigment retention in systemic sclerosis.¹ ² Both cellular and humoral immune factors in combination with external factors such as trauma or inflammation may trigger the destruction of melanocytes.³ Moreover, various physical factors like temperature changes as well as genetic, hormonal factors may influence pigment formation. Such changes in pigmentation is also seen during repigmentation around hair follicles in vitiligo. Clinically, both vitiligo and depigmented lesions of systemic sclerosis present as chalk-white macules with well-defined borders. However, mucosal involvement is commonly seen in vitiligo while depigmented lesions are usually not observed involving the lips or oral cavity in systemic sclerosis.

References


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