CASE STUDIES

Extensive oral telangiectasias in systemic sclerosis

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Abstract

The word telangiectasia is derived from tel- end, angos- vessels and ectasis- dilatation. Telangiectasia can be primary or secondary. Connective tissue diseases like systemic sclerosis, dermatomyositis and systemic lupus erythematosus may be associated with secondary telangiectasias. Systemic sclerosis presents with mat-like telangiectasias over face, hands and rarely oral mucosa. The present study discusses a case of extensive oral telangiectasias without significant sclerosis over face.

Keywords: Telangiectasias, systemic scelrosis, sclerodactyly

Introduction

Telangiectasias are permanent/temporary dilatations of pre-existing vessels with no proliferation. Depending upon the underlying vessels, the colour may vary. The disease may involve the skin or mucosal surfaces and it can be congenital or acquired. The acquired forms may or may not be associated with atrophy of the supporting structures.[1]

Case report

A 62-year-old male was admitted with shortness of breath and was on treatment for interstitial lung disease. He was referred to dermatology OPD for lesions in the oral cavity, which were present for the past 6 months. There was no history of pain, epistaxis, bleeding from any other site of body, xeropthalmia or xerostomia. There was history suggestive of Raynaud's phenomenon for 6 months. On examination of the oral cavity, there was blanching of mucosa of hard and soft palate with multiple discrete macular telangiectasias (Fig.1). On the vermilion of the lips and dorsum of the tongu, similar telangiectasias were present with no evidence of atrophy or pigmentary alteration (Fig. 1).

Gingival mucosa was normal according to the patient's age and mouth opening was normal (47mm). Telangiectasias were present over both the cheeks with no evidence of facial sclerosis. Sclerodactyly with pigmentary changes and digital pitted scars were present. There was no evidence of sclerosis on any other part of skin or any other systemic involvement. Routine investigations were within normal limits. Antinuclear antibodies test on hep 2 cells was positive (1: 160, speckled pattern), Scl-70 was positive. Radiology of chest ill defined opacities in the lower zones bilaterally (Fig. 2). Ziehl Neelsen staining for sputum and CBNAAT (cartridge based nucleic acid amplification test) for AFB were negative. A final diagnosis of diffuse cutaneous systemic sclerosis with interstitial lung disease was made.

Discussion

The word telangiectasia is derived from tel- end, angos- vessels and ectasis- dilatation. They may appear on the skin or mucous membrane. Morphologically they can be classified as macular or papular. The macular variant may be primary or secondary. Various common causes of secondary telangiectasias without atrophy are seen in photosensitive disorders, rosacea, mastocytosis, HIV/AIDS associated and drug induced.[2] Connective tissue diseases, poikilodermas and sarcoidosis have telangiectasias associated with atrophy. Connective tissue diseases associated with telangiectasias are seen in systemic sclerosis, systemic lupus erythematosus, dermatomyositis, overlap syndrome, morphea, etc.[2] Telangiectasias in systemic sclerosis are usually described as mat like (polygonal in shape and 1-2mm across).[1] Oral telangiectasias in systemic sclerosis have been reported by Bajraktari *et al.* in 20% cases.[3] The current patient had no binding down of the skin over the face or perioral region, however skin over the palate was sclerosed. Literature search did not find any case of mucosal sclerosis

Fig.1: (a) Sclerosis (arrow) and telangiectasias (arrow) in oral cavity, (b) Telangiectasias over lips and tongue, (c) Sclerodactyly

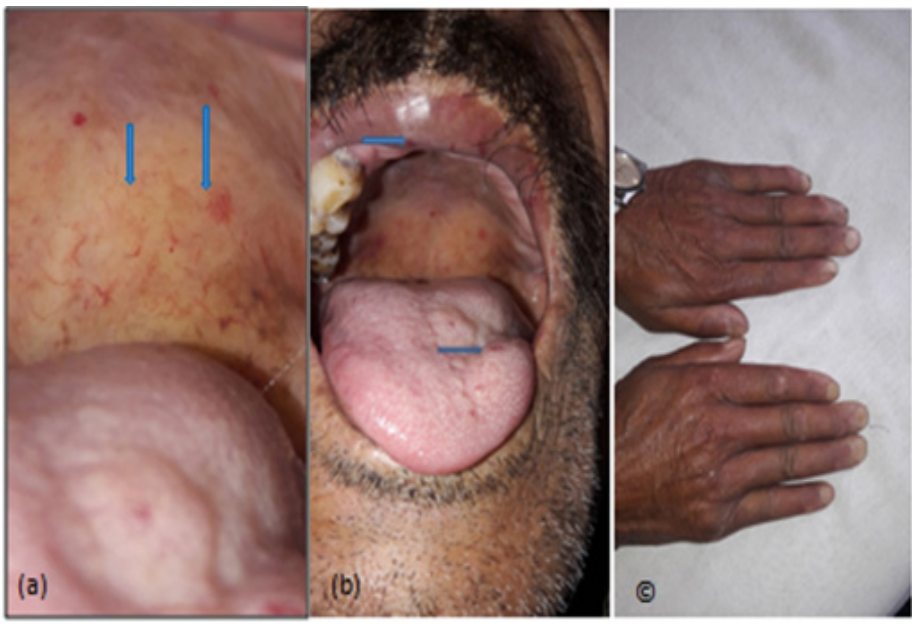
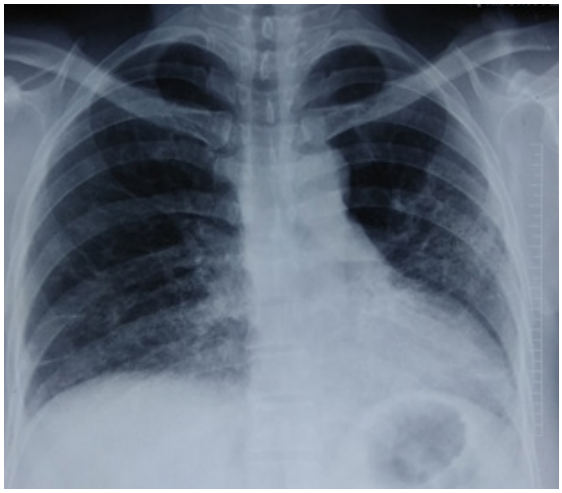


Fig.2: X ray chest showing ill-defined opacities over bilateral lower zones



preceding facial sclerosis.

Telangiectasias in scleroderma are usually post capillary dilatation of the upper horizontal plexus. Vasculature of endothelium of hands, face and mucosa are more prone for endothelial damage. The exact mechanism and predisposition of these sites is not known. Endothelial damage and alterations in the perivascular supportive tissues may lead to the formation of telangiectasias.[4] Oral telangiectasias are usually seen in Osler-Rendu-Weber syndrome (ORWS). Unlike scleroderma, telangiectasias in the ORWS is an autosomal dominant disease manifesting at earlier age without atrophy, with recurrent bleeding episodes and nose being the commonest site. In ORWS, similar to systemic sclerosis, primary intrinsic defect in endothelium and perivascular supporting connective tissue leads to vascular dilatation.[5]

In ORWS, there is a mutation in gene coding for endoglin and ALT-1 components of TGF β . This growth factor is not only important for vascular architecture but regulates matrix and basement membrane formation. Similarity in sites, morphology and microscopy of lesions suggests that there is an acquired TGFβ receptor defect in scleroderma

also. Sclerosis , which may cause only endothelial damage, is not responsible for the formation of telangiectasia. In the current case, though sclerosis was seen in oral mucosa. there was no facial sclerosis and he had extensive oral and a few facial telangiectasias.

Competing interests
The authors declare that they have no competing interests.

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