Case Report

A Rare Case of Limited Cutaneous Systemic Sclerosis – A Case Report

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Abstract

Limited cutaneous systemic sclerosis (lcSSc) is a sub-type of systemic sclerosis (SSc) characterized by association of Raynaud's phenomena with skin fibrosis limited to hand, face, feet and forearm. Manifests in females of 40-50 years (female: male – 4:1) with prevalence of 1/12500 adults. Disease originates from idiopathic auto-immune reaction leading to over production of collagen or with the chemical exposure. Prognosis of disease is relatively good with survival of 10 years in 80-90 % patients. Complications of Pulmonary arterial hypertension and lung fibrosis have poor prognosis.

Key words

Limited cutaneous systemic sclerosis, Autoimmunity, Raynaud's phenomena, Pulmonary arterial hypertension (PAH), Lung fibrosis, Anti-centromere antibody.

Introduction

Limited cutaneous systemic sclerosis is a sub type of systemic sclerosis characterized by association of raynaud's phenomenon with skin fibrosis limited to hand, face, feet and forearm. Raynaud's phenomenon is the most common and often first sign of the disease [1]. Other signs usually appear some years later. Skin tautness is absent in abdomen, back and thigh. Sometimes it can present with esophageal dysmotility and provoke gastro-esophageal reflex and dysphagia. In some early stages of the disease cutaneous involvement is not yet present as a consequence the disease is called limited cutaneous systemic sclerosis. The disease usually manifest between 40 to 50 years of age. Women are more affected than men (female: male – 4:1). Prevalence is estimated about 1/12500 adults. Exact cause is unknown, the disease originate from auto-immune reaction leading to over production of collagen or with exposure to chemicals (silica, solvents and hydrocarbons) [1]. Prognosis of Limited cutaneous systemic sclerosis is relatively

good with survival of 10 years in 80-90 % patients. Pulmonary arterial hypertension and lung fibrosis have poor prognosis.

Case report

A patient of 42 year old female was presented to Malla Reddy Institute of Medical Sciences with chief complaints of joint pains since 2 year (Shoulder, elbow, wrist, ankle, inter-phalangeal joints of both hands), tightness and dryness of skin and burning sensation of eyes since 1 year, whitish discoloration of fingers along with pain since 1 year, Recurrent history of cough and cold since 1 year for which she got hospitalized 4 times, dysphagia for solids since 6 months, fever since 2 months and shortness of breath and dry cough since 1 month. No complaints of any morning stiffness or movement difficulty and no variation of joint pain as the day progress. Patient had history of similar complaints in the past 2 years ago. In 2016, patient had similar complaints along with increased fatigability, decreased appetite, weight loss and a swelling in the right axillary area with serous discharge which was diagnosed as Tb lymphadenitis for which she was started on anti-tubercular therapy and she completed course for about 12 months. In November of 2018, patient had history of similar complaints along with severe joint pains which on investigating found out as chikungunya by serology. No history of any Diabetes mellitus, hypertension, epilepsy, coronary artery disease, cerebro vascular accidents, asthma, thyroid disorders.

General Examination

Patient was having dry, scaly and thickened skin with sparse hair (**Figure - 1, 2**) with patches of hyperpigmentation and depigmentation of the skin (**Figure - 3, 4**) on face, neck, arm, legs, ankle and distal to ankle with peri oral skin tightness with small oral aperture (**Figure - 3**).

Figure – 1, 2: Dry, scaly and thickened skin.

Figure – 3, 4: Patches of hyperpigmentation and depigmentation of the skin.

Investigations revealed her Hb was 10.8gm% peripheral smear showed normocytic normochromic picture with ESR of 25 mm/hour. RA factor and CRP were negative, ANA (140.9

U/ml) strongly positive, with Anti-centromere antibody positive on immunobloating (intensity ++++), mantoux test was negative, no erythema and induration after 72 hours at injected site. Pulmonary function tests revealed FEV1/FVC was 60%. High resolution computerized tomography of chest suggestive of pleural thickening and atelectatic bands in bilateral apical and lower lobes, with calcified lymph nodes in peripancreatic region. On echocardiography mild pulmonary arterial hypertension with right ventricle systolic pressure of 42 mm of Hg with mild tricuspid regurgitation was seen, Upper GI endoscopy suggested of mild antral gastritis with mild duodenitis. Ultrasonography of abdomen suggested of minimal ascities. Liver function tests and renal function tests were normal. Patient was kept on low dose steroid (wysolone – 10 mg) with methotrexate 7.5 mg once weekly with folic acid supplementation, proton pump inhibitor (rabeprazole with domperidone) for anti-reflux measures, calcium channel blockers (nicardipine -10 mg/BD) for raynaud's phenomenon.

Discussion

This is a case of limited cutaneous systemic sclerosis (lcSSc) presented with tightness and dryness of skin and burning sensation of eyes, raynaud's phenonomena, dysphagia, recurrent history of cough and cold and joint pains. On examination our patient is having dry, scaly and thickened skin with sparse hair with patches of hyperpigmentation and depigmentation of the skin on face, neck, arm, legs, ankle and distal to ankle with peri oral skin tightness with small oral apperture. The face, including the lips and frenulum of the tongue, may also be affected in systemic sclerosis (SSc). However, typical facial features associated with SSc include telangiectasia, a beak-shaped nose and reduced apperture of the mouth (microstomy). There may also be radial furrowing around the mouth. As the skin thickens and hardens, the patient's face develops an expressionless, mask-like stiffness and appears 'mummified'. Patients with lcSSc usually present with raynaud's phenomenon years prior to the development of additional symptoms. Non-pitting edema of the fingers is often an early cutaneous manifestation of SSc after which the skin of the swollen fingers starts to thicken and highly disabling sclerosis of the fingers (sclerodactyly) develops. Pitted scarring of fingertips due to loss of substance from digital pulp, with tapering of fingertips is commonly seen. Painful digital ulcers that occur on the fingertips as a result of local ischemia and vascular insufficiency are a frequent complication[2,3]. Secondary bacterial infection of these slowly healing digital ulcers can lead to gangrene, atrophy and auto-amputation. Calcinosis, the abnormal deposition of calcium in the tissues, is common in lcSSc, and usually occurs over pressure points. Other common skin manifestations of SSc include hypo and hyperpigmented areas of skin (salt and pepper), loss of hair follicles and loss of sebaceous glands (anhydrosis) with resultant dryness. Pruritis associated with dry skin can be intensely irritating and bothersome to SSc patients. Joint contracture is another important cutaneous manifestation of SSc. While some SSc patients have skin lesions that remain largely confined to the extremities, others exhibit skin thickening that extends progressively from the extremities to the trunk. Cutaneous symptoms often associated with or preceded by raynaud's phenomenon (an episodic digital ischemia provoked by cold or emotional stress) and arthralgias of the fingers are common, early signs of SSc, and therefore helpful for establishing a diagnosis [4].

Our patient's investigations revealed anti-nuclear antibody (ANA) strongly positive, with anti-centromere antibody positive, high resolution computerized tomography of chest revealed bilateral pleural thickening and atelectatic bands with echocardiography suggestive of pulmonary arterial hypertension, upper gastro-intestinal endoscopy suggestive of antral gastritis and duodenitis, ultrasound of abdomen shows ascities. Patient was kept on immunosuppressive and symptomatic medical therapy. Majority of patients will have positive ANA with 40% to

90% will have anti-centromere antibody as positive. Patients with lcSSc are less likely to suffer from renal crisis but more likely to suffer from isolated pulmonary arterial hypertension and not associated with interstitial lung disease. Diagnosis of lcSSc depends on disease pattern recognition, not by histopathology. Despite an increased risk of pulmonary arterial hypertension in lcSSc, diffuse cutaneous systemic sclerosis (dcSSc) is considered the more serious of the two clinical subsets being associated with significant morbidity from skin thickening as well as excess mortality due to severe cardiac, pulmonary, gastrointestinal and renal involvement [5]. A link between severe skin involvement and major visceral complications in dcSSc was first established in the 1960s and has been confirmed in more recent studies [6, 7].

Although there has been much debate about how best to classify SSc given its heterogeneity, the separation of SSc patients into one of two principal clinical subsets, dcSSc and lcSSc, is the most commonly used classification today. This classification, which has its origins in criteria proposed by the ACR in 1980 [4], is based on the nature and extent of skin involvement (typical skin thickening and hardening) together with the presence of certain autoantibodies and, particularly, with certain organ involvement. These later criteria, as well as the addition of raynaud's phenomenon and nail-fold capillary microscopy [to separate idiopathic raynaud's phenomenon (primary) from raynaud's secondary to SSc (secondary)], have greatly increased the sensitivity of the original ACR criteria [8, 9]. For example, in dcSSc, there is a high frequency of interstitial lung disease (ILD), whereas in lcSSc, there is a closer association with pulmonary vascular disease and pulmonary arterial hypertension (PAH). Anti-centromere antibodies are most frequently seen in patients with lcSSc, whereas anti-DNA topo I (anti-Scl-70) antibodies are associated with diffuse cutaneous involvement, increased frequency of pulmonary fibrosis and higher mortality [10]. As according to above criteria our patient is having lcSSc with pulmonary arterial hypertension.

Conclusion

Limited cutaneous systemic sclerosis is a disease of middle aged female and a sub-type of systemic sclerosis characterized by association of raynaud's phenomenon with skin fibrosis limited to hand, face, feet and forearm. These patients may experience myriad systemic complications like esophageal dysmotility, interstitial lung disease and isolated pulmonary arterial hypertension, so screening of middle aged female is recommended in suspicion of disease under expertisation. Proper treatment at early diagnosis improved the morbidity and mortality associated with renal crisis.

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