Association Of Sclerodema With Pelvic Floor Muscle Dysfunction - A Case Report

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DOI: 10.47750/pnr.2022.13.S04.246

Abstract

Scleroderma is a rare multisystemic autoimmune disease characterised by vasculopathy, inflammation and fibrosis of the skin and many other organs. Here we report a case of a 56 year old female who presented with Limited cutaneous scleroderma associated with pelvic floor dysfunction with uterine, rectal and bladder prolapse, recurrent hernias, urinary incontinence and bilateral varicose veins. These associations have been reported in literature but overlooked as an association or manifestation of scleroderma due to its uncommon presentation along with the disease. All these associations should be kept in mind and screened for in all patients presenting with Scleroderma for better management of overall condition of the patient and to also address female sexual dysfunction , thus helping in improving the quality of life of these patients.

KEYWORDS: Scleroderma, Pelvic floor muscle dysfunction, rectal prolapse, bladder prolapse, uterine prolapse, urinary incontinence, limited cutaneous scleroderma.

INTRODUCTION :

Scleroderma is a multisystemic autoimmune disease characterised by vasculopathy, inflammation and fibrosis of the skin and many other organs. There is loss of cutaneous elasticity , tightness followed by thickening and hardening of skin (sclerosis). It can be classified based on skin sclerosis into Limited Cutaneous scleroderma and diffuse cutaneous scleroderma. ANA positivity is seen in majority of the patients. Anti-topoisomerase (Scl70) antibody positivity is commonly seen in diffuse cutaneous scleroderma while Anticentromere antibody is more commonly associated with limited cutaneous scleroderma. Sclerodermal fibrosis can also affect other organs apart from skin such as oesophagus, lung , heart, kidneys, urinary bladder, gastrointestinal tract etc . Here we report a patient diagnosed to have Limited cutaneous scleroderma associated with pelvic floor muscle dysfunction with uterine, rectal and bladder prolapse, recurrent hernias, urinary and fecal incontinence and bilateral varicose veins. In this patient, all the above mentioned associations are seen as a consequence of sclerodermal fibrosis and other manifestations of scleroderma.

CASE REPORT:

A 56 year old female residing in Kanchipuram came to the dermatology OPD with the complaints of salt and pepper discoloration and thickening of skin over upper back, chest [Figure 1a and 1b] and bilateral ear lobe associated with itching over these lesions for the past 5 years. Patient gave history of darkening of face and bilateral lower limb [Figure 2] over the past 5 years and recurrent ulcerations over the medial aspect of bilateral lower limb above the ankle . Patient experienced difficulty in free movement of fingers with difficulty in mixing

food and other activities due to thickening and tightening of fingers. Patient also gave history of urinary incontinence and gastric symptoms such as regurgitation, retrosternal burning sensation and dyspepsia for which patient takes tablet pantoprazole 40mg on and off.

Patient gave past history of recurrent hernias for which mesh repair was done 24 years back, history of varicosities for which radiofrequency ablation/ foam sclerotherapy of bilateral lower limb and surgical management was done 5 years back and history of grade 3 uterine prolapse along with rectal and bladder prolapse were present for which patient underwent Ward Mayo hysterectomy procedure and bladder and rectal prolapses were also repaired.

On examination, salt and pepper pigmentation of skin was present over bilateral ear lobe, chest and upper back and Induration was present over these lesions. Hyperpigmentation of face was present. Thickening of skin was observed over distal phalanx of both hands with movement of fingers slightly restricted. Diffuse Hyperpigmentation over bilateral lower limb and foot with atrophy and thinning giving an inverted champagne appearance was observed [Figure 2] . Mouth opening and protrusion of tongue were normal and no oral ulcers were observed. Nails showed Beu's lines , longitudinal ridges and nail plate thinning. Genital region, palms and soles, scalp and hair were not involved.

Skin biopsy was done and showed skin lined by thin epidermis with melanin incontinence and collagenisation in the upper and deeper dermis with perivascular and periadnexal inflammatory infiltrates and these features were suggestive of Sclerodema [Figure 3] . Laboratory investigations were done and the patient was found to have ANA positivity , positive for Anti centromere antibody and Anti Histones Antibodies and had Hb of 9.6g/dl with normal liver and renal function tests. Bases on the clinical findings, skin biopsy and other laboratory investigations, a diagnosis of Limited cutaneous scleroderma associated with pelvic floor muscle dysfunction with uterine, rectal and bladder prolapse, recurrent hernias, urinary incontinence and bilateral varicose veins was made.

Figure 1a.

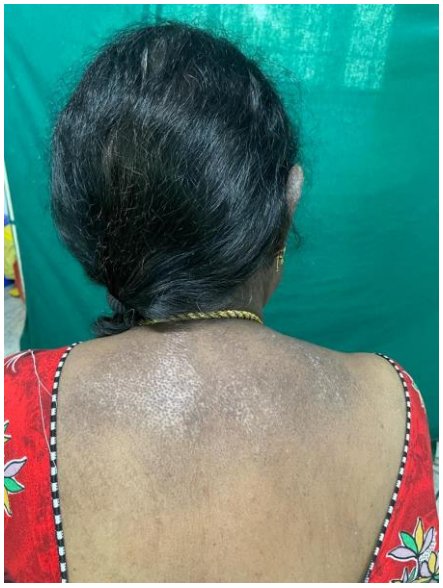

Figure 1b.

(Figure 1a and 1b)
Multiple hypopigmented and hyperpigmented macules present over anterior aspect of neck and upper back suggestive of salt and pepper pigmentation.

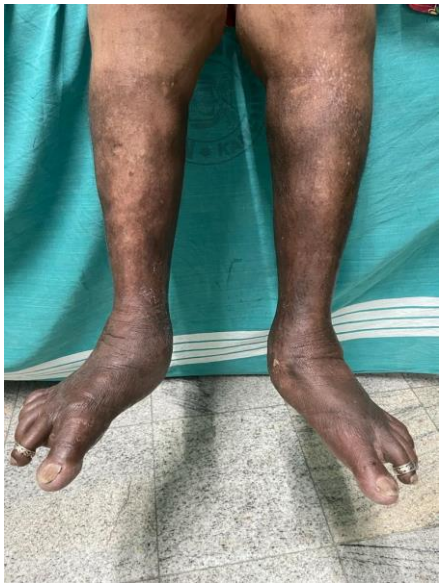

Figure 2. Diffuse Hyperpigmentation over bilateral lower limb and foot with atrophy and thinning giving an inverted champagne bottle appearance.

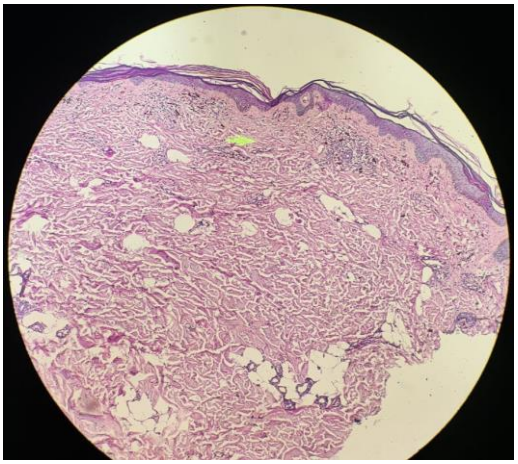

Figure 3. HPE - H and E ,10X , showing atrophied epidermis and homogeneous, hyalinised , hypertrophied collagen.

DISCUSSION:

Scleroderma is a multisystemic autoimmune disease characterised by vasculopathy, inflammation and fibrosis of the skin and many other organs. Skin, oesophagus, lung , heart and kidneys are the most frequently affected organs. It can be classified according to skin sclerosis into:

LIMITED CUTANEOUS SCLERODERMA: Skin sclerosis of fingers (sclerodactyly) with or without mild sclerotic lesions at the neck, face and armpits.

DIFFUSE CUTANEOUS SCLERODERMA : Skin sclerosis extends to proximal limbs, trunk and face.[1]

On further literature search , we found that there are associations of scleroderma with pelvic floor muscle dysfunction, female sexual dysfunction and pelvic organ prolapses like uterine, rectal and bladder prolapse as seen in this patient.There is a significantly more impairment of sexual and pelvic floor muscle function in women with scleroderma when compared to that of their age matched healthy controls and it was more commonly found when there is increased systemic inflammation, presence of interstitial lung disease, lung involvement, muscleweakness , myositis, altered collagen synthesis,etc.[2]

Another article showed association of rectal prolapse with scleroderma. Gastrointestinal tract affection is seen in around 90% of patients with systemic sclerosis and Anorectal involvement was reported in about 50-70% of scleroderma patients. Pathophysiology of rectal prolapse and concomitant fecal incontinence is closely related to disordered collagen synthesis, sphincter atrophy, rectal fibrosis, gut dysmotility resulting in constipation, dystrophic muscle, damage during child birth and neuropathy as suggested by absent rectal inhibition reflex.[3] [4]

There is also an association of limited cutaneous scleroderma and /or patients with Anti Centromere Antibody positivity (as seen in this Patient) with urinary incontinence . Changes in bladder volume and function can be found in more than two third of patients. Fibrosis affects the bladder wall and/ or the urethra in these patients . The Pattern of deposition of fibrosis results in two morphological extremes of bladder presentation in Systemic sclerosis patients: A small , thick and not compliant bladder and a large hypoactive bladder. Urinary incontinence in these patients can also be due to other comorbidities, prevalence of other rheumatological diseases, concomitant gastrointestinal tract involvement, presence of pulmonary arterial hypertension or worsening of dypsnoea, autonomic nervous system dysfunction, antibodies inhibiting parasympathetic neurotransmission and many other causes. In this Patient, though the cause of urinary incontinence can be attributed to bladder prolapse along with other pelvic floor organ prolapses, even after repair of the prolapses, patient still continues to suffer from urinary incontinence. Urinary incontinence in such a presentation can also be considered as a consequence of sclerodermal fibrosis affecting the bladder or urethra .[5][6]

CONCLUSION:

Scleroderma is a rare multisystemic autoimmune connective tissue disorder that affects different organs. The association of scleroderma with pelvic floor muscle dysfunction presenting as uterine, rectal or bladder prolapse and other associations such as urinary and fecal incontinence are commonly overlooked as a manifestation of scleroderma or as a complication of other manifestations of scleroderma. All the above mentioned associations should be kept in mind and be recognised as an association with scleroderma. All these associations should be considered, screened for and evaluated while treating a patient with scleroderma to improve the overall quality of life of the patient.

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