CASE REPORT

SCLEREDEMA DIABETICORUM; A RARE DISEASE

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Scleredema Diabeticorum (SD) is a rare condition characterized by diffuse, symmetrical induration along with non-pitted swelling mostly on the upper back as a result of mucin being deposited in the dermis. It can also involve posterior neck, shoulders, and scalp. We report a case of 48 years old female patient from Pakistan, with uncontrolled diabetes mellites type 2 for the last 15 years, presenting with thickened skin at the back of the neck resulting in difficulty in neck and shoulder movements. This led to decreased functional class from I to II causing her to develop insomnia and depression. Scleredema diabeticorum is a difficult disease to manage as it runs a long and debilitating course with little propensity of remission with available treatments.

Keywords: Scleredema diabeticorum; Diabetes; Cutaneous manifestations; Pakistan.

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INTRODUCTION

As per World health organization (W.H.O.) about 422 million people worldwide are affected from diabetes mellitus and about 1.5 million deaths are directly related to it each year. Longstanding uncontrolled diabetes mellitus is also a source of various cutaneous manifestations in at least 30% to 79% of diabetic patients.[1] This includes acanthosis nigricans (AN), necrobiosis lipoidica (NL), diabetic dermopathy (DD) as well as Scleredema diabeticorum (SD). First recognized in 1970, Scleredema diabeticorum has since been considered a rare cutaneous manifestation in diabetics which due to its low prevalence, 1.4 % as per a study done by Trihan JE *et al.*[3] in France, and often remains unnoticed.[2] This disease can lead to severe discomfort to the patient if ignored and it must be diagnosed earlier so that it could be referred for appropriate management as soon as possible Hence, the need to report this case arises to emphasize the importance of earlier diagnosis.

CASE REPORT

A 48-year-old female patient from Islamabad, Pakistan presented in our dermatology OPD with complain of some skin tightness which she felt on upper back and shoulders causing her to develop moderate to severe restriction of movement at shoulder joints bilaterally as well as at the cervical spine. She also started experiencing depressive symptoms because of her decreased functionality for which she also visited psychiatrist multiple times.

She was a known case of diabetes mellitus type 2, which was poorly controlled with HbA1c of 9.3%, diagnosed 15 years back while she was being investigated for increased lethargy and generalized weakness. Over this period, she developed symptoms of paraesthesia and numbness in feet. There was complain of pain in thigh muscles over walking and swelling over feet was also noticeable. So microvascular complications like nephropathy and neuropathy were identified. Her recent baselines tests showed Haemoglobin 13.1 g/dL, WBC count 5810 /µL, platelets 333000 / µL, creatinine of 1.3 mg/dl, ALT of 45 U/L She also had significant history of coronary artery disease leading to Percutaneous coronary intervention (PCI) to right coronary artery. Her medical history was not significant otherwise. Her surgical history included one caesarean section apart from her angioplasty. She did have a significant family history of uncontrolled diabetes as well as dyslipidaemia. Her current medications included tablet atorvastatin 10mg, tablet aspirin 75mg, tablet bisoprolol 5mg, tablet sitagliptin/metformin 50/500mg.

On physical examination, there was a noticeable swelling of upper back and neck which on palpation revealed symmetric, non-tender skin induration as shown in figure 1. Mild to moderate restriction was also noted in range of motion.

She was investigated for any connective tissue disease. Her autoimmune profile was negative for ANA, anti DsDNA, anti-histone, anti-smith, anti RNP, SCL70, anti Jo1, anti SSA and anti SSB making it unlikely that she was suffering from diseases like systemic sclerosis or dermatomyositis. We ordered X-ray cervical spine which showed muscular spasm only. Other laboratory evaluations included normal blood count, urinalysis, liver, renal and thyroid profile as well as normal CRP levels. For her skin thickening at the back, we initially advised Ultrasonography which showed thickening up to 4mm while no cyst, mass or collection was noted. We then advised biopsy. Figure 2 shows posterior view of the patient post biopsy. Histopathology showed atrophic

epidermis, with fibrosis at dermo-epidermal junction, collagen fibres were thick abnormally separated by clear spaces suggesting mucin deposition. Dermis showed chronic inflammatory infiltrates as shown in figure 3. This, indeed, attributed further to our clinical diagnosis of Scleredema diabeticorum.

She was then counselled in detail regarding the need to control her diabetes. She was referred to phototherapy unit of another hospital for Narrow band UVB phototherapy as first line treatment due to unavailability of the facility in our hospital. She was also sent to physiotherapist to help her control her restricted movement at cervical spine and shoulders. She was also counselled to be compliant to her endocrinologist and to visit nutritionist so that she has better understanding regarding her glycaemic control. Three months after her initial visit she returned with improvement in her symptoms and functionality.

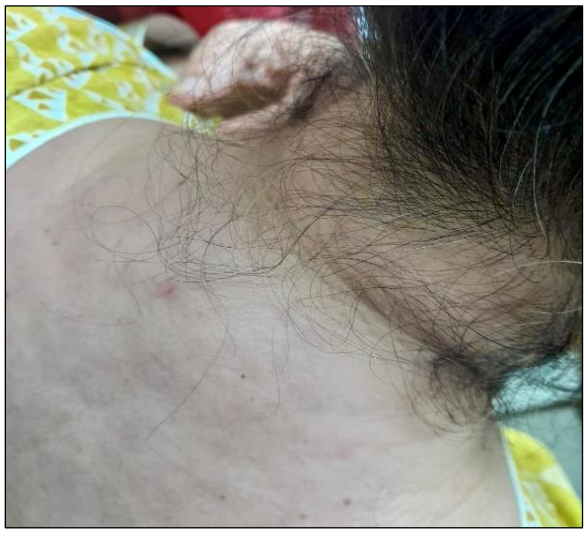

Figure-1: Lateral view of back of neck showing symmetrical induration of skin

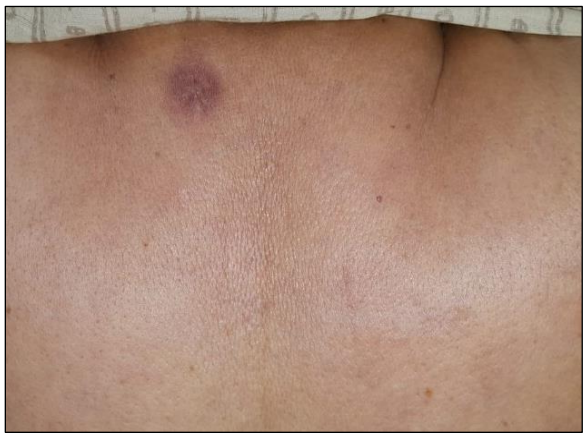

Figure-2: Swelling of upper back and neck with post inflammatory hyperpigmented patch formed after taking biopsy sample.

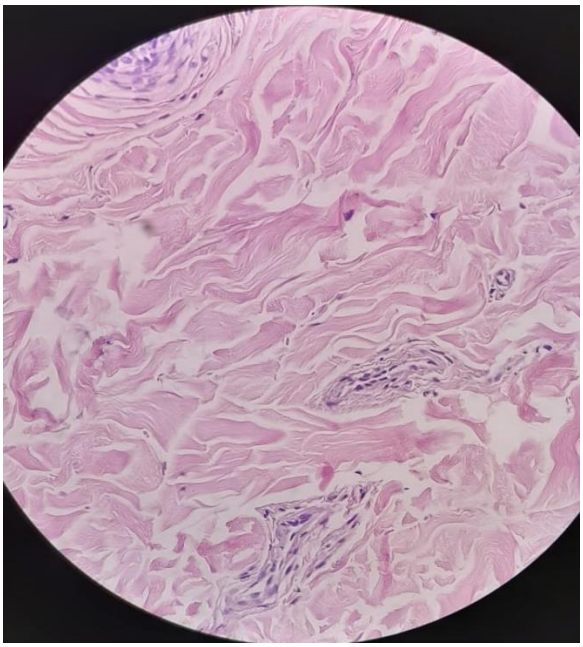

Figure-3: Histopathology (H & E) showing atrophic epidermis with abnormally thick collagen fibres separated by clear spaces suggesting mucin deposition. Dermis showing chronic inflammatory infiltrates.

DISCUSSION

Scleredema diabeticorum is a variant of disease called Scleredema Adultorum (SA) which is classified in to three types depending upon the association with monoclonal gammopathies, infection and diabetes. Scleredema diabeticorum, being the most common variant, is the one associated with diabetes.[1]

It causes non-pitted induration of skin symmetrically because of the thickening of the dermis with excessive mucin deposition among large collagen bundles.[4] Scleredema diabeticorum can occur with longstanding diabetes of both types and doesn't have any racial predilection; however, it is noted that it occurs in men predominantly.[5] It mostly involves the skin over the neck, shoulders and upper limbs. It is usually asymptomatic but erythema, pruritis and hypoesthesia can be accompanying symptoms.[1] It also causes limitation of the range of motion resulting in difficulty in daily life activities.[2] A case was reported in Spain by García-Arpal M *et al*.[6] where patient suffered from Scleredema diabeticorum in a 'cuirasse' pattern causing inability to fold the skin in the scapular retraction movement hence, resulting in restrictive lung disease.

The exact pathophysiology behind Scleredema diabeticorum is unknown but it has been noted that poor glycaemic control can be the culprit behind the nonenzymatic glycosylation process damaging the collagen fibres, resulting in more mucin

produced by fibroblasts which is then deposited in dermis.[2]

The treatment options that have been experimented with, include pituitary extract, thyroid hormone, immunosuppressant, antibiotics even corticosteroids but none showed effective results.[5] Although it is seen in many cases that phototherapy (particularly psoralen plus UVA) or electron-beam therapy have showed good results, we still do not have any standard treatment protocols and algorithms.[1] In one case series by Sun M *et al* [4] three patient with Scleredema diabeticorum were treated with tranilast (a tryptophan metabolite analogue), given for 3 months duration which ultimately caused reduction in skin thickness on ultrasound findings and improvement in patient's symptoms.

Due to its chronicity, it can hinder daily life activities and can lead to depression. Pereira M *et al*.[7] in their case report also found that their patient was suffering from depression from the last 10 years, just like in our case report, however its not life threatening.

CONCLUSION

Scleredema diabeticorum is a difficult disease to manage as it runs a chronic and enfeebled course with hardly any remission with available treatment. Hence, we realise the need of highlighting this chronic condition which has rarely been reported in South Asian, Pakistani population in particular, to establish the prevalence so that more research can be done to look for newer treatment options.

Consent: Verbal and written consent about publication has been given by the patient.

Conflict of interest: Nil

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