International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2024: 7.101

A Rare Presentation of Plasma Cell Disorders - Scleredema: Case Report

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Abstract: ***Background: Scleredema is a rare connective tissue disorder characterized by thickening of the skin and subcutaneous tissue, involving skin of the neck, shoulders, upper limbs and face. Its association with paraproteinemia is very rare. Case Presentation: A 24 - year - old female homemaker presented with progressive skin tightening over her upper limbs, face, back, and chest, along with difficulty swallowing for three years. Examination revealed taut, shiny facial skin with decreased wrinkling, reduced mouth opening and skin tightening extending to the arms. She did not have other symptoms suggestive of systemic sclerosis. Rheumatological workup was negative for systemic sclerosis. Skin biopsy showed collagen fibers deposition separated by clear spaces suggestive of scleredema. Further workup revealed serum electrophoresis suggestive of an IgA lambda peak. Her bone marrow biopsy revealed cellular marrow with approximately 20% plasmacytosis. Flowcytometry found 20% plasma cells in the interstitium with immunophenotypes of CD 138+, confirming the diagnosis of Smoldering Multiple Myeloma. Conclusion: Scleredema is a rare disorder which has an unexplained association with paraproteinemia.***

Keywords: Scleredema, Plasma cell disorders

1. Introduction

Scleredema is a rare connective tissue disorder with a poorly understood pathogenesis. It is characterized by thickening of the skin and subcutaneous tissue. It mainly involves skin of the neck, shoulders, upper limbs and face. It is typically associated with an underlying disease, such as poststreptococcal infection, paraproteinemia, or diabetes mellitus. [1] Association of scleredema with paraproteinemia is rare. [2]

2. Case Report

A 29 - year - old female homemaker presented with complaints of progressive skin tightening affecting both upper limbs, face, back, and chest, restriction of movement at neck and shoulder joint, along with difficulty in swallowing for the past 2 years. She had no joint pain, joint swelling, sclerodactyly, digital ulceration, oral ulcers, alopecia, Raynaud's phenomenon. She had no constitutional symptoms and exposure to any drugs, chemicals or radiation. Her vitals were normal. On examination, the skin over her face appeared tight, taut, and shiny with induration, making it difficult to pinch, associated with decreased wrinkling. The skin tightening extended up to the arms in the upper limbs, upper back and chest. She also had decreased mouth opening and perioral wrinkling. There was restriction of neck movement and overhead abduction of shoulder joint due to skin thickening. The Modified Rodnan Skin Score was 20. [3] The scalp, nail and rest of the systemic examination were normal. Her complete blood count, liver function test and renal function test, serum calcium levels, ASO titres were normal and 24 hour urinary protein was 136mg per 24 hours. Upper gastrointestinal endoscopy could not be performed as patient was unable to swallow the probe. Barium swallow showed normal swallowing, with no strictures. HRCT of the chest showed no evidence of interstitial lung disease. Patients autoimmune antibody profile, thyroid profile, viral markers were negative. Her HbA1c was also normal.

A skin biopsy (Figure 1) was done which showed collagen fibers deposition separated by clear spaces suggestive of scleredema. Further workup revealed serum electrophoresis suggestive of an IgA lambda peak (Figure 2). Her IgA was 1390 mg/dl. Serum free light chains, beta - 2 microglobulin, and other immunoglobulin levels were within normal limits. Her bone marrow biopsy revealed cellular marrow with approximately 20% plasmacytosis. Flowcytometry found 20% plasma cells in the interstitium with immunophenotypes of CD 138+. There was no evidence of any end organ damage. Skeletal survey and Whole-body PET - CT did not reveal any lytic lesions. These results confirmed that she had low a risk smoldering multiple myeloma according to the Mayo Clinic Criteria. [4]

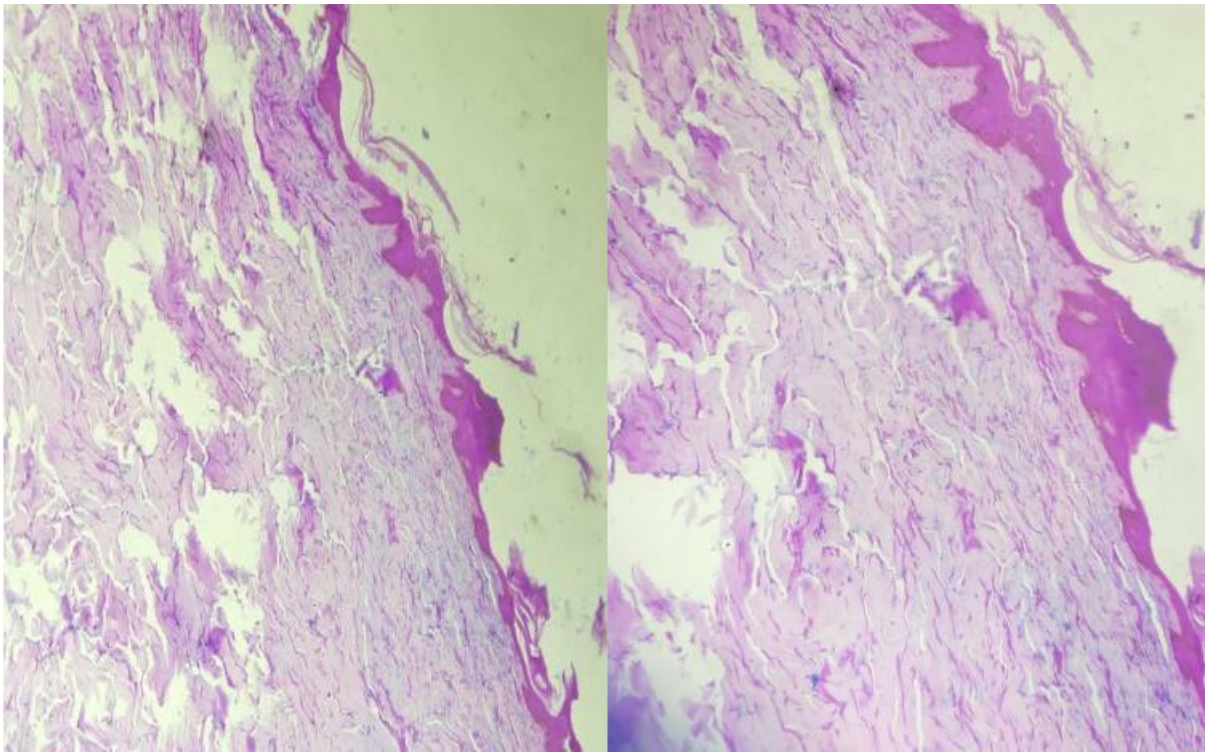

Figure 1: Skin biopsy showing collagen fibers deposition separated by clear spaces

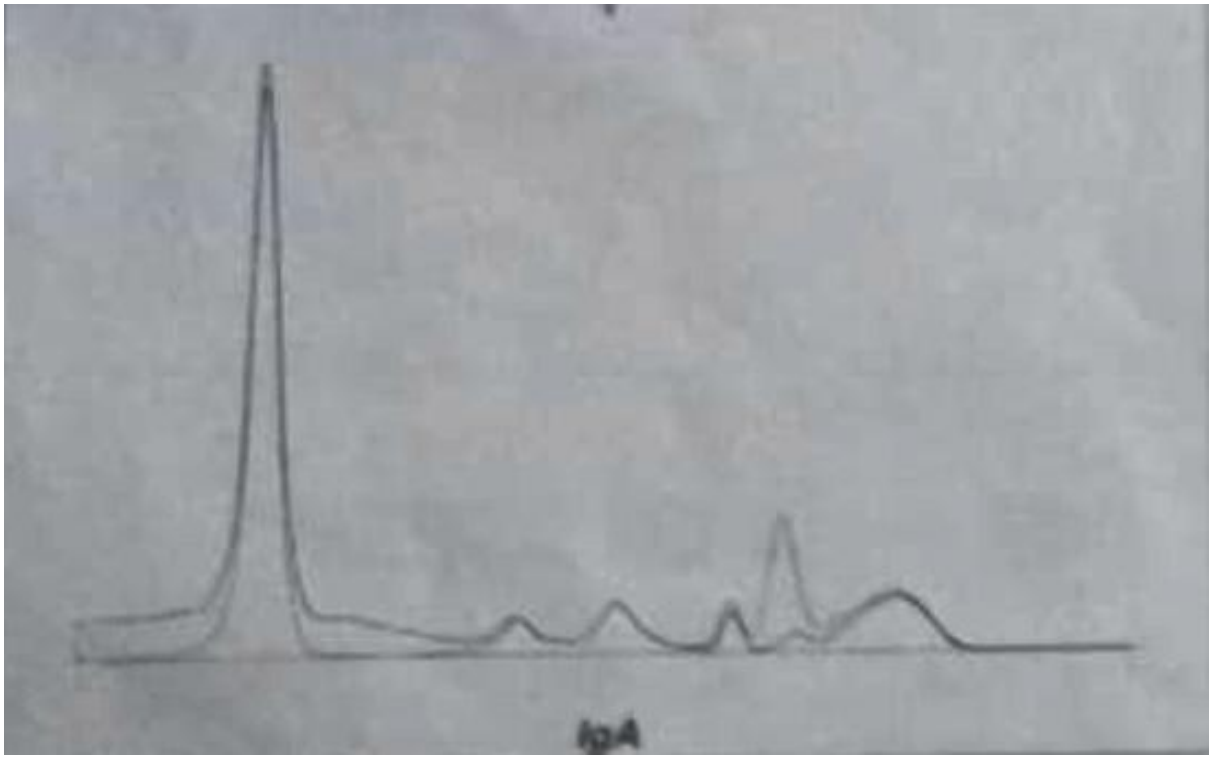

Figure 2: IgA lambda peak on serum electrophoresis

The patient was treated with ultraviolet light therapy in consultation with dermatology department and showed only slight improvement. Patient was counselled for chemotherapy; however, patient did not give consent. The patient was discharged and advised to follow up at 6 months interval and will be monitored for the evolution of Multiple Myeloma.

3. Discussion

Scleredema is a rare connective tissue disorder causing thickening and tightening of the skin, often associated with diabetes, infections, or paraproteinemia. It is classified into three groups. Type 1 usually occurs in children, post infectious, commonly associated with group A beta hemolytic streptococcus. It is acute in presentation, develops after two to three weeks after a febrile illness and resolves spontaneously in months to years. Type 2 is rare, occurs is

middle aged adults and often associated with paraproteinemia. It is chronic and progressive. Type 3 is associated with patients with poorly controlled diabetes. It is chronic and slowly progressive. [5]

The pathogenesis of scleredema is not well understood. It is associated with the increased collagen and glycosaminoglycans production. Various systemic treatments have been explored for scleredema, such as corticosteroids, pentoxifylline, cyclosporine, methotrexate, hydroxychloroquine, and intravenous immunoglobulin. However, these therapies lack universal approval and frequently yield limited success. [6] [7]

The majority of information on scleredema is derived from individual case reports. A comparable large - scale study dates back to 1984 and includes 33 patients documented by the Mayo Clinic. We found a multicentric study published 2015, including 44 patients and Monoclonal gammopathy was identified in five patients, including two with the IgG lambda type and three with the IgG kappa type. Among them, two were diagnosed with multiple myeloma, while the remaining three had monoclonal gammopathy of undetermined significance. While treatment reports remain inconsistent, our data suggest UVA1 phototherapy as the first - line option for severe cases. Systemic corticosteroids and immunosuppressants should be reserved for refractory cases or scleredema linked to multiple myeloma. [6] [8]

Various physical therapies, including UVA, UVB, PUVA, localized electron beam therapy, photopheresis, thalidomide, and intravenous immunoglobulin (IVIg), have shown potential benefits in managing the disease, as reported in various case series and individual reports; however, reports on treatment approaches appear inconsistent and lack strong supporting evidence. [7] [9] [10]

According to International Myeloma Working Group (IMWG) risk stratification system, our patient was stratified as Low risk and kept on observation.

4. Conclusion

Scleredema should be included in the differential diagnosis for patients presenting with diffuse skin thickening lacking the hallmark features of systemic sclerosis. Additionally, evaluating for monoclonal gammopathy and multiple myeloma is crucial in such cases.

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