ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 11, Issue, 02, pp.1181-1182, February, 2019
DOI: https://doi.org/10.24941/ijcr.34412.02.2019

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

RESEARCH ARTICLE

RARE MANIFESTATION OF LIMITED SCLERODERMA: ONLY FACIAL SKIN INVOLVEMENT

***[1]Dr. Dipesh Patel, [2]Dr. Hetal Pandya, [3]Dr. Hiral Barot and [1]Dr. Dhruv Kotecha**

[1]Junior Resident, Department of General Medicine, SBKS MI&RC, Piparia, Vadodara
[2]Professor and Head, Department of General Medicine, SBKS MI&RC, Piparia, Vadodara
[3]Senior Resident, Department of General Medicine, SBKS MI&RC, Piparia, Vadodara

ARTICLE INFO

Article History:
Received 14th November, 2018
Received in revised form
17th December, 2018
Accepted 20th January, 2019
Published online 28th February, 2019

Key Words:

Scleroderma, limited scleroderma, rare presentation

ABSTRACT

Scleroderma is a multisystem autoimmune disease in which there is increased fibroblast activity resulting in abnormal growth of connective tissue. Scleroderma is classified into two main types: Diffuse cutaneous and limited cutaneous scleroderma. Commonest presentations of limited scleroderma were Calcinosis, Raynaud's phenomenon, oesophageal dysmotility, Sclerodactyly, Telangiectasia and only facial skin manifestation is very rare presentation. We are presenting a case report of rare involvement of facial skin only as an initial manifestation of limited scleroderma.

Citation: Dr. Dipesh Patel, Dr. Hetal Pandya, Dr. Hiral Barot and Dr. Dhruv Kotecha. **2019.** "Rare manifestation of limited scleroderma: only facial skin involvement", *International Journal of Current Research*, 11, (02), 1181-1182.

INTRODUCTION

Scleroderma is a multisystem autoimmune disease in which there is increased fibroblast activity resulting in abnormal growth of connective tissue. This causes vascular damage and fibrosis. The Early Stage of disease is associated with Prominent Inflammatory features, progressed to Functional and structural alterations in multiple vascular beds and progressive visceral organ dysfunction. The disease is more common in women with 4:1 female to male ratio[2]. Scleroderma is classified into two main types: Diffuse cutaneous and limited cutaneous scleroderma. Commonest presentations of limited scleroderma were Calcinosis, Raynaud's phenomenon, oesophageal dysmotility, Sclerodactyly, Telangiectasia. We are presenting a case report of rare involvement of facial skin only as an initial manifestation of limited scleroderma.

CASE

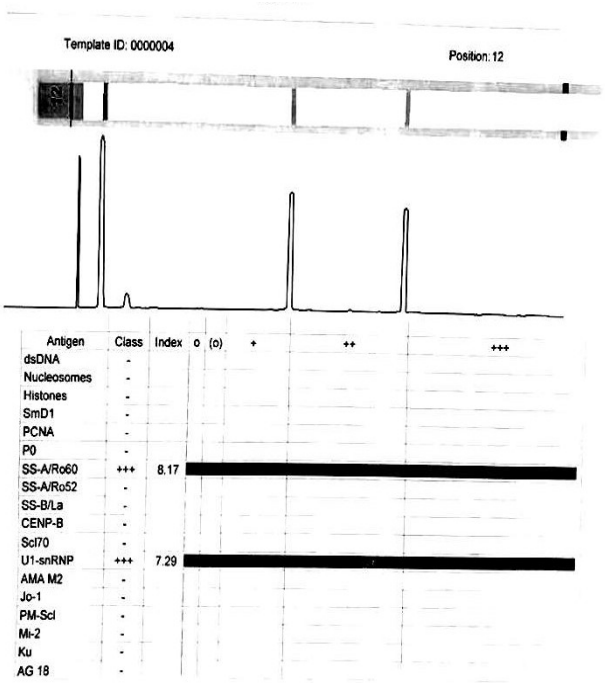
A 40 year old female patient noticed tightening of skin over Right face leads to difficulty in speaking since last 6 months and complain increased since last 1month. She does not have complain of Dryness of mouth and eyes, low grade fever, pain, difficulty in swallowing, thickening of skin over fingers or toes, rashes on skin or mucous membrane. No past history of any major medical illness. On local and systemic examination no positive findings were noted. Her Haematological and Biochemical investigation profile was as follow: CBC: Hb-11.4 gm/dl , TC - 11500/ cumm , Platelet – 4.05 lacs/ml , ESR- 51 mm/hr , Urea -33 mg/dl , creatinine- 0.7 mg/dl , Bilirubin-0.5 mg/dl , SGPT-62 U/L , SGOT-90 U/L , TSH- 1.82 mlU/L , RBS -104 mg/dl. EGC and 2D Echo did not show any abnormality. On clinical suspicion of limited cutaneous presentation of connective tissue disease, her ANA profile was done. ANA Profile positive for U1-snRNP antibody with +3 intensity and SS-A/Ro60 antibody with +3 intensity which suggestive of limited scleroderma. Patient was treated with oral steroid and methotrexate after which patient felt better and was relived of her symptoms.

DISCUSSION

Scleroderma was first described in 1753 by Carlo Curzio of Naples, Italy. Scleroderma is a connective tissue disease characterized by excessive fibrosis, microangiopathy and immune dysregulation, including autoimmunity and chronic low-grade inflammation[3].Limited cutaneous scleroderma[4] is the more common type of scleroderma. Areas of skin affected include only the face, forearms and lower legs up to the knee. Abnormal production of type 1 collagen by fibroblasts leads to accumulation of glycosaminoglycan (GAG) and fibronectin in

****Corresponding author:*** **Dr. Dipesh Patel,**
Junior Resident, Department of General Medicine, SBKS MI&RC, Piparia, Vadodara.

extra cellular matrix. The older term for limited scleroderma is CREST syndrome (Calcinosis, Raynaud's phenomenon, oesophageal dysmotility, Sclerodactyly, Telangiectasia). Diffuse cutaneous scleroderma[6] is less common and Associated with Extensive skin In duration, Starting in Fingers ascending from Distal to proximal limbs and trunk. There is higher risk of mortality in diffuse scleroderma. Multiple genes were contributed to disease susceptibility, but environmental exposures are likely to play a major role in causing and progressing the disease. The majority of disease-associated autoantibodies in Scleroderma patients are anti-nuclear antibodies (ANAs) that target proteins that play essential roles in transcription, splicing, and cell division. Currently, various ANA specificities specific to scleroderma have been reported and well characterized. These include anticentromere, anti-topoisomerase I (topo I), anti-RNA polymerase (RNAP) III, anti-U3 ribonucleoprotein (RNP), anti-Th/To, anti-U11/U12 RNP, anti-PM-Scl, anti-Ku, anti-RuvBL1/2, anti-U1 RNP antibodies. In our patient U1 RNP and SS-A/Ro60 antibody was found positive. As U1 RNP antibody also found in multiple connective tissue disorder, this patient might develop multiple system involvement but yet there is no other involvement occur in patient at present.



The disease might have been picked up in very initial stage, where patients might progress to develop other involvement, yet to have facial skin involvement is very rare phenomenon.

The main focus of treatment is slow the progression of disease. As there are no specific treatments for scleroderma itself, but organ system–specific complications are treated.[5,6]Main treatment of autoimmune disorders is immunosuppressant like corticosteroid, Methotrexate. Prednisolone is an immunosuppressant used for treatment of autoimmune disorders. It may decrease inflammation by reversing increased capillary permeability and suppressing inflammatory cell activity.

Outcomes are generally good for limited scleroderma of the skin when there are no lung complications. They are worse for those with the diffuse skin disease, particularly in older age and males. Death occurs most often from lung, heart and kidney complications. In diffuse cutaneous disease, five-year survival is 70% and 10-year survival is 55%. As there is no definite curative treatment for this disease, it is always better to pick up early diagnosis and start treatment at initial stage of disease to slow down the progression of disease and lower complications to have better quality of life. In our case, the diagnosis was made very early stage, when patient was having minimal involvement. Starting treatment at this early stage will be beneficial for the patient to have prolonged disease free life.

Conclusion

We conclude that if a patient presents with facial skin tightening it should not be neglected as a normal or hysterical complain as on further investigation it can turn out to be an atypical presentation of limited scleroderma.

REFERENCES

Gabrielli A, Avvedimento EV, Krieg T. 2009. "Scleroderma". *New England Journal of Medicine*. 360 (19): 1989–2003.

Harrison's principle of internal medicine/19th edition, chapter 382-page: 2154

Harrison's principle of internal medicine/19th edition, chapter 382 -page: 2155

Klippel, John H. Primer On the Rheumatic Diseases 11ED. Atlanta, GA: Arthritis Foundation.

Mayes MD, Lacey Jr JV, Beebe-Dimmer J, et al. Prevalence, incidence, survival, and disease characteristics of systemic sclerosisin a large US population. *Arthritis Rheum.* 2003; 48(8):2246–55.

Varga J, Abraham D: Systemic sclerosis: a prototypic multisystem fibrotic disorder. *J Clin Invest*., 117: 557-567.

****