

Case Report

Scleromyxedema in an HCV-positive male

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Abstract Scleromyxedema is rare chronic cutaneous mucinosis usually associated with a monoclonal gammopathy and underlying systemic disease.

We report a 42-year-old male who developed scleromyxedema and was also HCV positive.

Key Words

Scleromyxedema, lichen myxedematosus, HCV

Introduction

Scleromyxedema is a rare chronic disease with an unknown etiology.¹ It is characterized clinically by generalized papular eruption and diffusely thickened plaques over the face, trunk and acral areas. Histopathologically there is mucin deposition in the dermis.² Many patients reveal a monoclonal gammopathy on serum electrophoresis. The cutaneous involvement in scleromyxedema is often associated with underlying systemic pathology. Hepatitis C is one of the rare associations.³ We report a case of scleromyxedema who had HCV infection.

Case report

A 42-year-old diabetic, alcoholic, HCV-positive male reported with a 2-year history of asymptomatic papular lesions over his natal cleft, fingers of right hand, face and nape of neck. He had developed a feeling of

tightening and hardening of skin over the hands, lower trunk and perineum for the last 6 months. There was also complaint of polyuria and generalized weakness for the past 5 months. Cutaneous examination revealed skin-colored firm papules and nodules coalescing to form plaques over the natal cleft and adjoining buttock area bilaterally (**Figure 1**). Similar lesions were present over the index finger of right hand (**Figure 2**), right periorbital region and over nasal bridge (**Figure 3**). There were multiple diffusely thickened, indurated, firm, skin-colored papules and plaques over the anterior chest and posterior and lateral aspect of trunk (**Figure 4**). The plaques were about 5 to 6 cm in size having diffuse margins and prominent follicular openings over the surface. The sensations over the lesions were normal. On systemic examination there was hepatosplenomegaly.

Laboratory investigations revealed normocytic hypochromic anemia, raised blood sugar levels and normal renal function tests. Liver function tests showed markedly raised SGPT and SGOT and normal alkaline phosphatase levels. The patient was anti-HCV positive. The HIV test was negative.

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Figure 1 Skin-colored papules and nodules coalescing to form plaques in the natal cleft.



Figure 2 skin-colored firm papules and nodules on fingers.



Figure 3 skin-colored papules on nose.



Figure 4 Skin-colored papules and nodules coalescing to form plaques.

Serum electrophoresis was normal. Histopathology revealed alcian blue-positive homogeneous deposit in the upper and mid dermis along with fibroblast proliferation.

Discussion

Lichen myxedematosus is a rare chronic disease.¹ It usually appears between 30 and 70 years and has no sex predilection. A spectrum of disease is seen with localized less severe papular forms and a generalized form known as scleromyxedema.^{4,5} Scleromyxedema, also known as Arndt-Gottron (S-AG) syndrome^{6,7} is characterized clinically by generalized papular eruption often linearly arranged and coalescing into diffuse scleroderma-like thickened, indurated plaques. Large areas of the body may be affected including the face, giving the appearance of leonine facies, upper

limbs, trunk and less frequently the lower limbs. The diagnostic criteria for scleromyxedema include generalized papular and sclerodermoid eruptions, mucin deposition, fibrosis and fibroblast proliferation, monoclonal gammopathy and absence of thyroid disease.⁸ Apart from skin, various other organs may be involved by the disease. Proximal myopathy, polyarthritis, hoarseness, dysphagia, ectropion and lagophthalmus have been observed.² Lung disease, cardiovascular involvement and nervous system may also be involved. Endocrine abnormalities have not been reported so far.¹ Although our patient was found to be diabetic for past 5 months. The disease may also be associated with underlying malignancies including hepatocellular carcinoma. Lo and Tzung reported an HCV positive male with scleromyxedema and underlying hepatocellular carcinoma.⁹ The disease has also been associated with chronic hepatitis C.¹⁰ Hiroyuki and colleagues reported a case of 55 years old female with scleromyxedema and HCV.³ They reported that HCV has commonly been observed in Japanese patients¹¹ of lichen myxedematosus but it has rarely been seen outside Japan. The study of literature reveals that the case we report is the first case of a patient with HCV and scleromyxedema in our setup. The case was referred to a gastroenterologist for his liver problem. We recommend HCV screening in cases of scleromyxedema.

References

1. Sarkany RPE, Breathnach SM, Seymour CA *et al.* Metabolic and

- nutritional disorders. In: Burns T, Breathnach S, Cox N, Griffiths C. *Rook's Textbook of Dermatology*. 7th edn. London: Blackwell Science; 2004. p. 57.1-83.
2. Bari AU, Rashid M, Rahman SR. Acral persistent mucinosys: a rare variant of cutaneous mucinosis. *J Pak Assoc Dermatol* 2004; **14**: 253-6.
3. Banno H, Takamo H, Nitta Y *et al.* Lichen myxedematosus associated with chronic hepatitis C. *Int J Dermatol* 2000; **39**: 212-4.
4. Rongioletti F. Lichen myxedematosus (papular mucinosis): new concepts and perspective for an old disease. *Semin Cutan Med Surg* 2006; **25**: 100-4.
5. Rongioletti F, Rebora A. Updated classification of papular mucinosis, lichen myxedematosus and scleromyxedema. *J Am Acad Dermatol* 2001; **44**: 273-81.
6. Korber A, Frankson T, Graffe S, Dissemmond J. Successful therapy of scleromyxedema with low dose intravenous immunoglobulin. *J Eur Acad Dermatol Venerol* 2007; **21**: 553-4.
7. Rampino M, Garibaldi E, Ragona R, Ricardi U. Scleromyxedema: treatment of widespread cutaneous involvement by total skin electron beam therapy. *Int J Dermatol* 2007; **46**: 864-7.
8. Lin YC, Wang HC, Shen JL. Scleromyxedema. An experience using corticosteroid and review of the published work. *J Am Acad Dermatol* 2006; **33**: 207-10.
9. Lo P-Y, Tzung T-Y. Lichen myxedematosus in a patient with hepatocellular carcinoma. *Br J Dermatol* 2000; **143**: 452-3.
10. Rongioletti F, Rebora A. Worsening of lichen myxedematosus during interferon alfa-2a therapy for chronic active hepatitis C. *J Am Acad Dermatol* 1998; **38**: 760-1.
11. Masaki S, Nishio Y. Lichen myxedematosus associated with liver cirrhosis. *Hihubyou-Shinryou* 1994; **16**: 29-32 (in Japanese).