Case Report

'Atypical Localized Not Well-Specified Cases' a case study of a rare type of Localized Lichen Myxedematosus

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Abstract

Lichen Myxedematosus (LM) is an uncommon and distinct disease entity characterized by cutaneous mucin deposition in dermis. It is classified in several subtypes depending on many factors including the monoclonal gammapathy, pattern of skin involvement and systemic involvement. We now present a case of LM which is localized in respect of skin involvement but according to recent classification it falls into a diagnosis of 'atypical localized not well- specified cases'. This is a very rare case and to the best of our knowledge, it is the first case report in medical literature until now. We treated the case with a short course of oral Prednisolone and got excellent result. Patient has been followed up in every 6 month for last 13 yrs and observed no progression to scleromyxedema.

Keywords: Lichen Myxedematosus, Scleromyxedema, Mucin Deposition, Atypical Type, Treatment.

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Introduction

Lichen Myxedematosus (LM) is a rare idiopathic cutaneous disorder characterized by an abnormal accumulation of mucin in the dermis. Its classification dates back to 1953, when Montgomery and Underwood distinguished 4 types of LM. Still now, there is no consensus regarding type or classification of mucinosis. Rongioletti and his colleagues[1,2] studied the disease extensively and classified it primarily into three clinico pathologic subsets: (i) a generalized and sclerodermoid form associated with monoclonal gammapathy and systemic, even lethal manifestations (the only one which should be called scleromyxedema) (ii) localized forms, in the absence of monoclonal gammapathy and systemic involvement, and it is subdivided into 4 subtypes: (1) a discrete popular form involving any site; (2) acral persistent popular mucinosis involving only the extensor surface of the hands and wrists; (3) papular mucinosis of infancy, a pediatric variant of the discrete form or the acral form of persistent papular mucinosis; and (4) nodular form. (iii) A third group of atypical or intermediate forms, not meeting the criteria for either scleromyxedema or the localized form, includes cases of (1) scleromyxedema without monoclonal gammopathy, (2) localized forms with monoclonal gammopathy and/or systemic symptoms, (3) localized forms with mixed features of the subtypes, and (4) not well-specified cases. In the literature, many forms of LM are reported. However due to its rarity, it is still a poor documented disease. In this paper authors describe a case of LM which does not fit with any subtypes of localized LM. We categorized it as a'atypical localized non- specified cases' depending the type and distribution of lesions.

Case Report

A 41 years old man presented himself with skin lesions in trunk and extremities for 7 months [Fig 1,2,3].

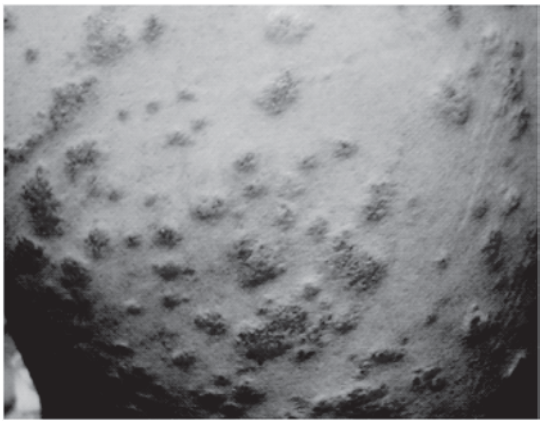

Figure-1: Erythematous Papules and Plaques in left side of trunk.

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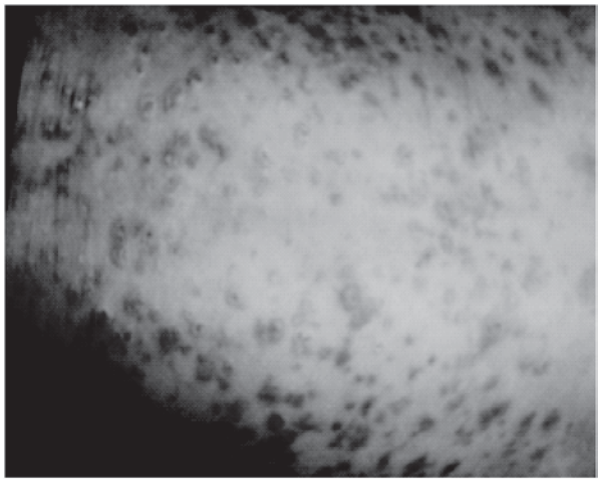

Figure-2: Linear pattern of lesions in back.

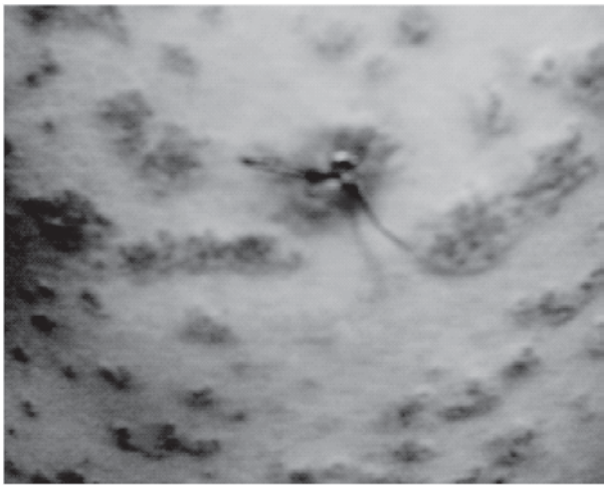

Figure-3: Site of biopsy.

He complained that the lesions were static for last five month without any discomfort except occasional pruritus. He also informed that he had no systemic disease.

Cutaneous examination showed multiple, well defined skin coloured to erythematous papule and plaques scattered over the trunk, thigh and arms. The lesions are few millimeter to few centimeter in diameter and confluent in trunk. Linear patterns of lesions were also noted in back of trunk [Fig:2].

On palpation, the lesions were non-tender, waxy and slightly hard in consistency. In general, thickening of skin, face involvement or systemic symptoms were not apparent. Laboratory evaluation showed a normal blood count, blood chemistry, HIV serology and thyroid hormones. Ig G paraprotein was absent.

A skin biopsy Revealed normal epidermis but moderate deposit of mucinous material in the upper and middle dermis separating the collagen bundles [Fig:5,6].

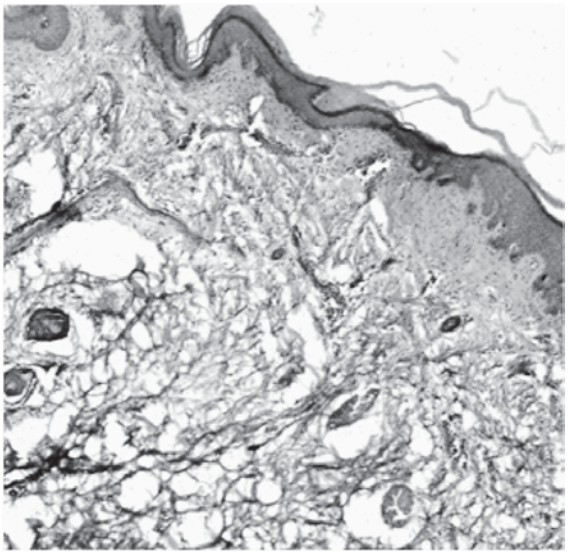

Figure-5: Collagen fibers were separated in upper dermis without epidermal changes (H&E100x).

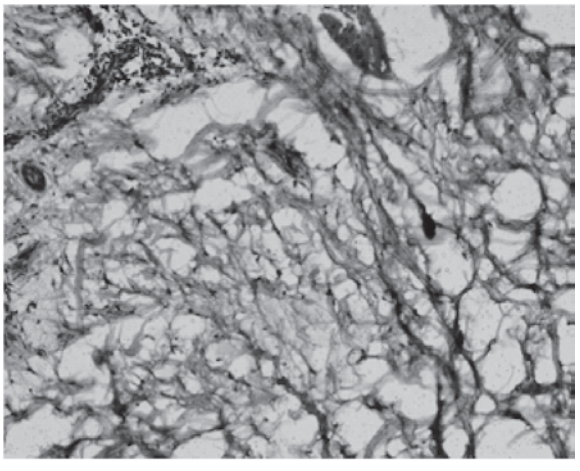

Figure-6: Mucin deposits between collagen fibers with proliferation of fibroblasts (H&E 400x).

A slight increase in fibroblasts with accompanying inflammatory infiltrate were also observed. According to Rongioletti and his colleagues, above mentioned histopathologic criteria are diagnostic for LM. He opined that neither collagen deposition nor sclerosis are typical features of LM[3].

The type and distribution of lesions of our patient did not match with any subtypes of localized form of LM. So in the light of Rongioletti classification, this LM case was categorized as 'Atypical Localized Not Well specified Cases'[1].

The patient was treated with oral prednisolone 20 mg for 10 days with tapering dose of 10mg for another 4 days. The patient was examined after 1 month and it was found that lesions were cured absolutely. Patient returned back after 3 month with a minor relapse and again he was treated with same doses of prednisolone.

In follow up examination after 1 month, [Fig:4] it was found that lesions were almost cured and left behind scars only.

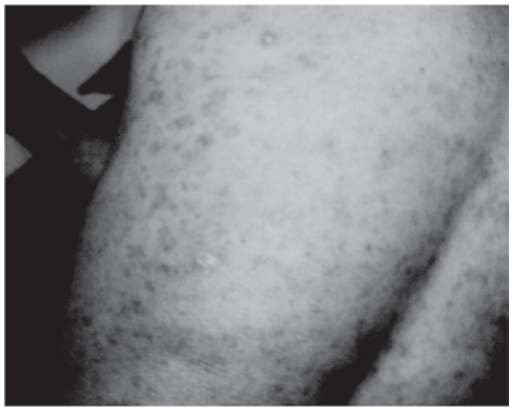

Figure-4: After 1 month of treatment.

The patient has been followed-up every 6 months for last 13 years. We did not observed any relapse and also there was no progression of the disease to sclerormyxedema or other systemic disease.

Discussion

Lichen Myxedematosus is one of the rare disease in dermatology. Until now less than 200 cases are documented in medical literature. Among the three major form, localized variants are benign in nature. Localized LM is further subdivided into the following four subtypes: (1) discrete LM, which has firm, smooth, waxy or flesh-colored papules of 2 to 5 mm in size, numbering a few to hundreds on the limbs and trunk in a symmetric pattern. The papules are isolated or form confluent nodules or plaque and rarely resolve spontaneously[1]; (2) acral persistent papular mucinosis, which is characterized by a few to multiple, ivory to flesh colored papules of 2 to 5mm in size on the back of the hands, wrists, and occasionally the distal aspect of the forearms. Papules persist or increase without spontaneous resolution and occur predominantly in females[4] (3) cutaneous mucinosis of infancy, characterized by firm, opalescent papules located on the upper arms, especially the elbows, and the trunk without spontaneous resolution[1], and (4) nodular-type LM, characterized by multiple nodules on the limbs and trunk, with a mild or absent papular component[2].

Due to its rarity, only few numbers of studies are available which focus on pathogenesis, clinical variants and management of the disease. Our case did not match with any subtypes of localized LM and according to Rongioletti classification it categorized into 'Atypical Localized Not Well-specified Cases'. In a Medline search, there is no single report is found on 'atypical localized non specified cases' of LM.

To the best of our knowledge, this is the first case report of the condition in medical literature until now. Treatment of LM is always a challenge to physician. Different drug or treatment modalities are tried. But still there are no consensus regarding its treatment. However, localised form of LM is benign in nature and many drugs are successfully tried. There was no documentation or guideline for the treatment of atypical localized case.We treated our case successfully with systemic prednisolone.

Conclusion

Lichen Myxedematosus is a rare but a distinct entity with wide varieties of clinical feature. It is not well documented in medical literature and may be under diagnosed. Different types of disease are not well studied. There are also lacking of knowledge in cause and treatment of the disease. We have to work a lot to unveil the many facts of this distinct disease entity.

Acknowledgement

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