

Linear morphea with secondary mucinosis

*Sujay Khandpur, Saurabh Singh, Vinod K. Sharma, Ruchika Gupta¹,
Manoj K. Singh¹*

Departments of Dermatology
and Venereology and
¹Pathology, All India Institute
of Medical Sciences, New
Delhi, India

Address for correspondence:
Dr. Sujay Khandpur,
Associate Professor,
Department of Dermatology
and Venereology, All India
Institute of Medical Sciences,
New Delhi - 110 029, India
E-mail:
sujaykhandpur@yahoo.co.in
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ABSTRACT

Secondary mucin deposition in the skin is a common feature of lupus erythematosus and dermatomyositis. In scleroderma, it occurs uncommonly or in small amount. We describe a 7-year-old boy with progressive, linear, bound-down plaques involving the thighs, lower abdomen and back with no systemic involvement. Histopathology showed features of scleroderma with abundant mucin deposition in the reticular dermis. This report highlights excessive mucin deposition in lesions of morphea.

Key words: Morphea, Scleroderma, Secondary mucinosis

INTRODUCTION

Mucin deposition is a common histological feature of various connective tissue diseases such as lupus erythematosus and dermatomyositis. In scleroderma, small and interstitial deposits of mucin are occasionally seen in the deep dermis and interlobular septa of subcutis.^[1] There are only isolated reports of scleroderma associated with abundant mucin deposits, manifesting as papules within or occurring beyond the indurated plaque. We describe an unusual case of linear morphea evolving into generalized morphea with excessive mucin deposition, without any systemic involvement.

CASE REPORT

A 7-year-old boy developed a linear, slowly progressive, bound-down plaque on the right thigh at 18 months of age preceded by MMR (Measles Mumps Rubella) vaccine. Subsequently, he developed similar plaques on the other thigh, lower abdomen and back. He had no systemic complaints. Dermatological examination revealed linear, light brown, well- to ill-defined, bound-down indurated plaques with overlying mild hypertrichosis in isolated plaques only, on lateral aspect of both thighs, lower abdomen and back [Figures 1 and 2]. A diagnosis of

linear morphea evolving into generalized morphea was made. Skin biopsy done from outside was reported as cutaneous mucinosis. A repeat skin biopsy performed at our institute showed epidermal atrophy, hyalinization of collagen throughout the dermis and mucin deposition in upper reticular dermis [Figure 3a]. On alcian blue-Periodic Acid Schiff stain, abundant mucin deposition was demonstrated in the upper reticular dermis [Figure 3b]. Investigations including hematological, biochemical, serum and urine protein electrophoresis, thyroid function tests and antinuclear antibody (ANA) were normal.

A diagnosis of morphea with secondary mucinosis was made and oral mini-pulse steroid therapy with betamethasone, 3 mg every Saturday and Sunday initiated. Six months of treatment showed no response and repeat skin biopsies from right thigh and abdominal plaque showed features compatible with scleroderma. The dose of betamethasone was increased to 4 mg but with no improvement.

DISCUSSION

This is an unusual case of linear morphea with abundant mucin deposition. Mucin deposition is a constant feature in both morphea and systemic

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Figure 1: Linear, hyperpigmented, bound-down plaque of morphea over right thigh and lower abdomen. Scars are of biopsy site.



Figure 2: Hyperpigmented bound-down plaques of morphea over left thigh extending to abdomen

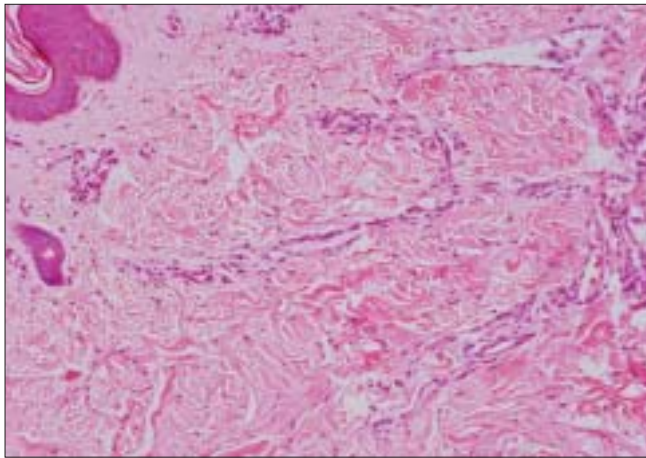


Figure 3a: Photomicrograph showing hyalinization and sclerosis of the dermal collagen with mild perivascular inflammatory infiltrate (H and E, x400)

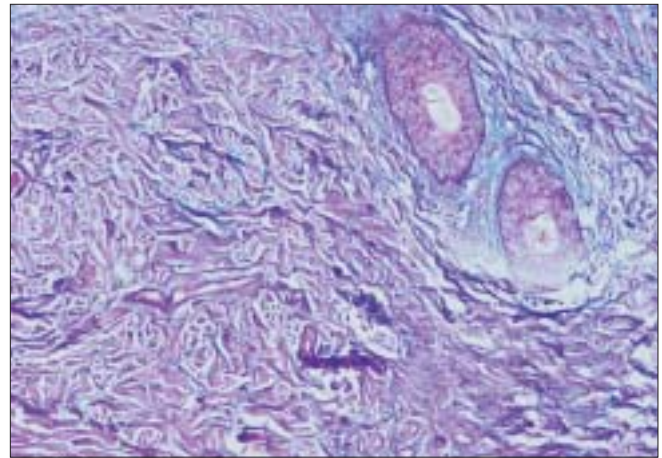


Figure 3b: Photomicrograph demonstrating abundant dermal mucin (Alcian blue-Periodic Acid Schiff stain x400)

sclerosis as shown in some case series.^[1,2] In such cases, the amount of mucin is usually mild, rarely evident on routine stains and located mainly in the deep reticular dermis and interlobular septa of subcutis. The deposits chiefly consist of dermatan sulfate and hyaluronic acid.

However, there are only isolated reports of localized (linear) or generalized morphea with remarkable mucin deposition.^[3-5] In fact in a few cases, this led to development of papules, nodules and plaques of cutaneous mucinosis both within and distant from the sclerotic areas. None of them had systemic involvement.

Pathogenic mechanisms for the production of mucin in scleroderma implicate the role of transforming growth factor- β , which stimulates fibroblasts to produce

increased amount of glycosaminoglycans, fibronectin and collagen, decreases extracellular matrix break down and reduces fibroblast susceptibility to apoptosis.^[6] Similarly, the role of CD4+ T cells, plasma cells and histiocytes, all of which may stimulate dermal fibroblasts, has also been postulated.

Our case must be differentiated from four patients reported by Prendiville *et al.* who presented with hyperpigmented and hypertrichotic plaques, which were also indurated in two cases, over the lower trunk, buttocks, genitalia and lower limbs with associated multisystemic involvement in the form of proptosis of the eyes, hepatosplenomegaly, paraortic lymphadenopathy, diabetes, dysmorphic features and cardiovascular abnormalities and histopathology showing benign dermal perivascular and interstitial infiltrate composed of lymphocytes, histiocytes

and plasma cells with mild subcutaneous infiltrate, hyalinization of collagen in reticular dermis including fibrosis and increased mucin in between collagen bundles.^[7] POEMS syndrome can also present with cutaneous changes of diffuse hyperpigmentation, thickening and sclerodermoid changes, diffuse hypertrichosis more prominent on face, chest and limbs, plethora, hemangiomas (glomeruloid hemangioma) and white nails. Diagnosis of POEMS syndrome requires two major features which include polyneuropathy and monoclonal plasma cell proliferative disorder and one minor feature such as sclerotic bone lesions, Castleman's disease, organomegaly (splenomegaly, hepatomegaly or lymphadenopathy), edema (edema, pleural effusion or ascites), endocrinopathy (adrenal, thyroid, pituitary, gonadal, parathyroid, pancreatic), skin changes or papilledema.^[8] Our patient showed linear bound-down plaques over lower trunk and thighs with mild hypertrichosis in isolated plaques only, no systemic involvement and histopathology showing abundant mucin deposition in upper reticular dermis with diffuse sclerosis of collagen.

We also excluded other cutaneous mucinosis such as scleredema and scleromyxedema. Scleredema manifests as sudden onset symmetric induration of skin usually preceded by prodrome of fever, body aches and an identifiable focus of infection. Histology reveals collagen bundles separated by large interfascicular space due to mucin deposition. In scleromyxedema, there are small clustered papules and diffuse plaques with constant facial involvement and usually associated with paraproteinemias. In contrast to scleroderma, skin is pinchable and moveable over the

subcutis. Deposition of mucin occurs in the papillary dermis.

Other dermatoses with secondary mucinosis include connective tissue diseases like lupus erythematosus (LE) and dermatomyositis, malignant atrophic papulosis and certain skin tumors like neural, mesenchymal and epithelial tumors.^[3]

Our patient did not respond to long-term steroid therapy. In the case of generalized morphea with papular mucinosis reported by Rongioletti *et al.*, the sclerotic plaques did not improve and new ones continued to appear despite being on prednisone 25 mg daily.^[5]

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