

CASE REPORT

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Morphoea Profunda Presenting with Atrophic Skin Lesions in a 26 Year Old Female: A Case Report

Smith Giri¹, Upama Paudel¹, Abhimanyu Jha², Dipendra Gurung³ and Sudip Parajuli¹

¹Department of Dermatology and Venereology, Tribhuvan University Teaching Hospital, Kathmandu, Nepal. ²Department of Pathology, Tribhuvan University Teaching Hospital, Kathmandu, Nepal. ³Department of Dermatology and Venereology, DI Skin Hospital, Kathmandu, Nepal. Corresponding author email: girismith@iom.edu.np

Abstract: Morphoea Profunda is a rare variant of Morphoea that presents clinically as a solitary fibrotic plaque. Morphoea Profunda presenting with atrophic lesions has rarely been reported in literature. We report the case of a 26 year old Nepalese lady who presented to us with multiple non-inflammatory atrophic lesions on her body without significant skin induration, pigmentation and texture change. The findings on histopathology confirmed a diagnosis of Morphoea Profunda. Hence, Morphoea Profunda should be considered in the differential diagnosis of anyone presenting with asymptomatic atrophy of the skin.

Keywords: atrophy, Morphoea Profunda, multiple lesions

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Introduction

Morphoea is a localized type of scleroderma, which is a connective tissue disorder affecting the microvasculature and loose connective tissue.¹ The etiology of Morphoea is unknown but environmental exposures, immune alterations, microchimerism with autoimmunity, trauma, *Borrelia* infection and familial predisposition have all been suggested as contributing to the pathogenesis of the disease.² Clinically Morphoea comprises of at least five distinct types: Morphoea en plaques, Bullous Morphoea, Linear Morphoea, Nodular Morphoea and Deep Morphoea.³ Four different types of Deep Morphoea have been distinguished: Subcutaneous Morphoea, Eosinophilic Fasciitis, Disabling Pansclerotic Morphoea and Morphoea Profunda.⁴

Here, we report an uncommon case of Morphoea Profunda (MP), a type of Morphoea rarely mentioned in the literature, presenting with multiple isolated non-inflammatory atrophic lesions mimicking Localized Lipoatrophy.

Case Report

A 26 year old female presented to us with asymptomatic atrophy of skin on multiple sites of her body since two years. It was first noticed on her anterior thighs followed in a few months by similar lesions gradually appearing on her buttocks, eventually appearing in her iliac region, back, left upper arm and her face within a year. She did not complain of pain, redness or itching on these areas. There was no history of trauma or any other skin lesions on those sites prior to this. There was no history of any systemic illness, neither a history of similar illness in her family. She was not under any medication.

On examination, there were multiple atrophic areas on her right cheek, left upper arm, both anterior thighs, buttocks, sacral region and scalp (Figs. 1–4). The lesions varied in size from 2 × 3 cm to 8 × 10 cm with ill-defined margins. The skin overlying the lesions showed normal color without any obvious induration or tenderness. Examination of the surrounding skin, hair, nails and other systems did not reveal any abnormality.

Her routine blood examination including hemoglobin, blood counts, and hematocrit, ESR, random blood sugar, urea, creatinine, sodium, potassium and

liver function tests were normal. Anti-nuclear antibodies, Anti dsDNA and serologies for Syphilis and HIV were non reactive. *Borrelia* Serology was not done due to unavailability of laboratory facilities.

Histopathological examination of a punch biopsy taken from a lesion on the left upper arm showed keratinized stratified squamous epithelium with increased basal pigmentation. Underlying papillary and reticular dermis showed thick and thin bands of collagenous tissue. The dermal adnexal glands showed lack of adipocytes. Subcutis revealed increased collagen bands with diminished adipocytes. There was patchy mononuclear infiltrate in the subcutis. (Figs. 5 and 6). A diagnosis of Morphoea Profunda was made based on this typical histological appearance.⁵

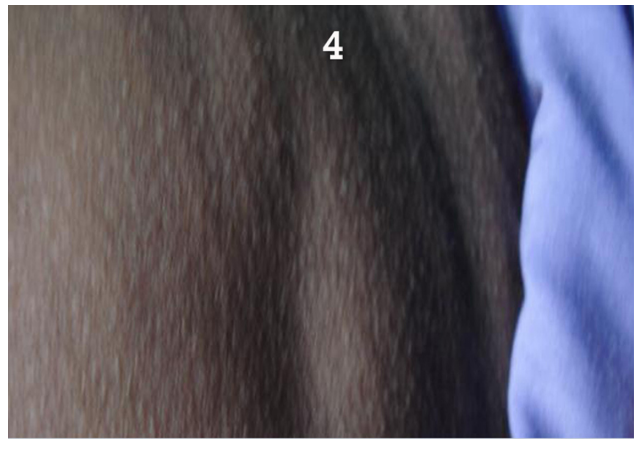
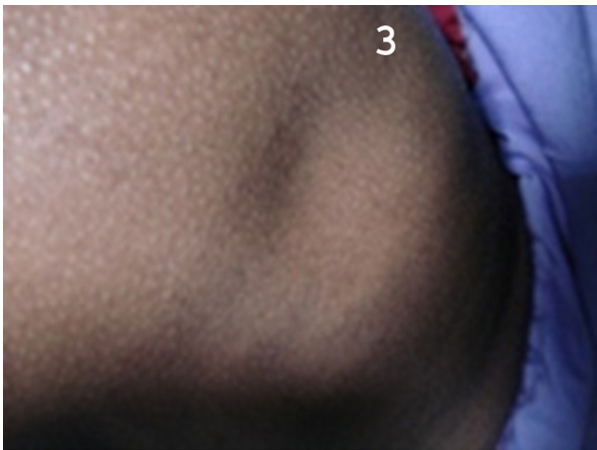
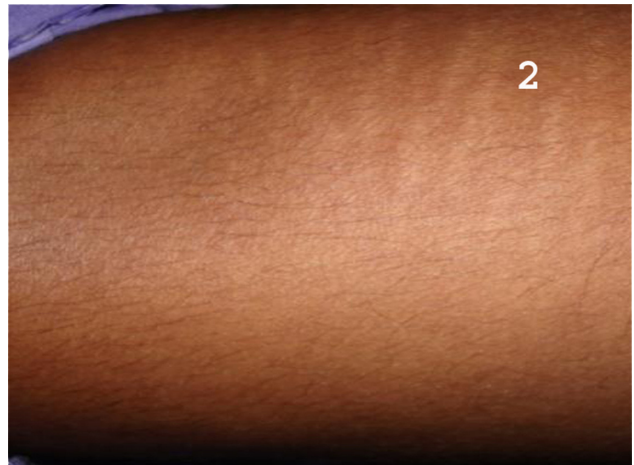
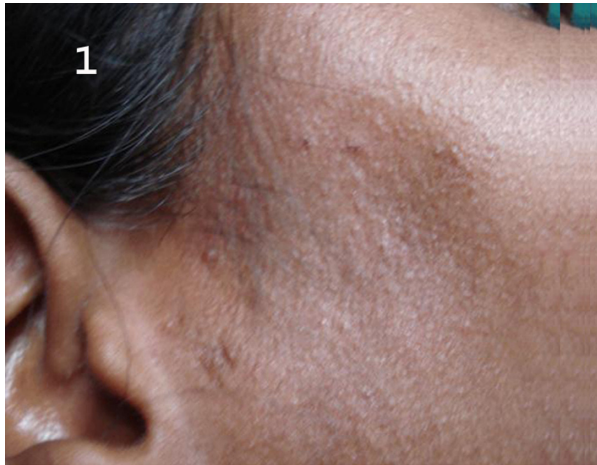
The patient was started on oral Hydroxychloroquine 200 mg twice a day and a follow up visit was scheduled in 3 months. On follow-up, the lesions had been static without any changes. No new lesions were noted during this period. The patient was advised to continue with her medication and the need for regular follow up was emphasized. However, the patient was lost to follow-up thereafter.

Discussion

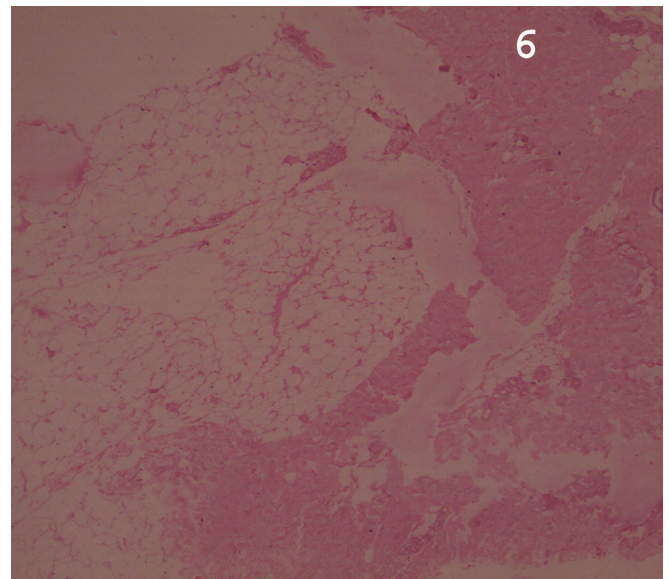
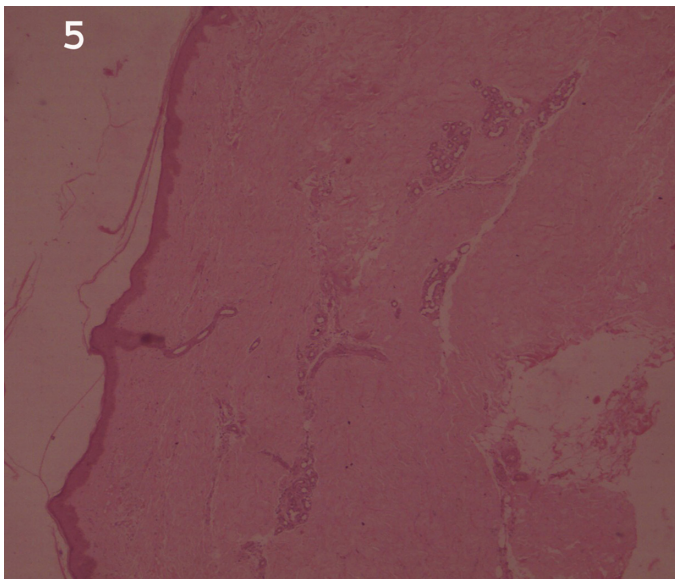
Scleroderma is a chronic disease of unknown etiology that affects the microvasculature and loose connective tissue. It is characterized clinically by fibrous deposition and obliteration of vessels in the skin, lungs, gastrointestinal tract, kidneys and the heart.¹ Scleroderma may be localized or generalized. Localized types are: Morphoea en plaques, Bullous Morphoea, Linear Morphoea, Nodular Morphoea and Deep Morphoea.³ A rare variant of Morphoea is Morphoea Profunda, first described by Whittaker et al in 1989 as a solitary fibrotic plaque.⁶

Patients with MP are usually middle aged with an approximately equal sex distribution. Most patients who are affected present with a single fibrotic plaque usually located over the back, shoulder and neck or paraspinal area involving the skin and deeper tissue.⁷

Our patient was unusual because she presented with deep atrophic lesions on multiple sites of her body without preceding inflammatory changes. Also, unlike the previous case reports of MP, the skin lesions showed no obvious changes in pigmentation, texture or induration. Such a presentation was similar to cases



Figures 1–4. Ill defined atrophic lesions without induration, abnormal pigmentation, or signs of inflammation in right cheek, left arm, right scapula, buttock, and respectively.



Figures 5 and 6. Punch biopsy of affected lesion showing hyalinization of the collagen bundles in the deep dermis and subcutaneous tissue associated with sparse inflammatory infiltrate that were suggestive of Morphoea Profunda in low power (Fig. 5) and high power (Fig. 6).



of Morphoea Profunda reported by Jablonska et. al. and more recently by Khelifa et al.^{8,9} However, in the former, the involvement was linear and band like and there was involvement of muscles as opposed to our patient. In the latter, there were multiple asymptomatic atrophic lesions in the left arm of a 53 year old woman as opposed to more widespread lesions in our patient.

The histopathological changes in MP are different from widespread Morphoea because the former have deeper involvement and more pronounced inflammatory infiltrates. Also the collagen in the reticular dermis and subcutis are hyalinized and sclerotic.⁸ Histopathologically, collagen deposits in the dermis and the subcutis, atrophy of the appendages, diminished adipocytes along with an inflammatory cellular infiltrate are considered pathognomonic of Morphoea; all of which was present in our case.

The cause of Morphoea is unknown. Studies have shown abnormalities in fibroblasts from patients with Morphoea including increased expression of a number of growth factors and their receptors.⁷ Other factors that have been implicated in the etiology are infection with *Borrelia*, genetic susceptibility, trauma and drugs including Bromocriptine and L-tryptophan.² Our patient did not have an obvious precipitating factor. However, *Borrelia* serology could not be done due to unavailability of laboratory facilities.

Morphoea Profunda should be differentiated from Eosinophilic Fasciitis and Atrophoderma of Pasini and Pierini. These diagnoses were ruled out based on the histopathological and clinical findings. Eosinophilic Fasciitis clinically presents as edema, skin induration with a “peau d’orange” appearance which was not present in our case.¹⁰ Atrophoderma of Pasini and Pierini was differentiated based on histopathological findings, as sclerosis was more prominent in our case. However, the two conditions can be regarded as a spectrum of the same disease process; Atrophoderma being a more abortive form of the atrophic process resulting in Morphoea.¹¹ Clinically two different conditions: Localized Involutional Lipoatrophy (LIL) and lipoatrophic scars of Lupus Panniculitis, can present with deep atrophic lesions similar to our case. However, in the former, the lesions are usually solitary and hypopigmented as opposed to multiple lesions and normal skin color in our case. Histologically in LIL, there are small attenuated fat lobules composed

of small adipocytes embedded in a well vascularized and hyalinized background.¹² These conditions were ruled out on the basis of histopathological findings.

We conclude that Morphoea Profunda can rarely present as deep atrophic lesions of the skin without preceding inflammatory changes in the skin as seen in typical cases. It can mimic Localized Involutional Lipoatrophy and other causes of deep atrophic lesions of the skin. Hence, it should be considered as one of the differential diagnosis in any patient presenting with deep atrophic lesions of the skin.

Author’s Contributions

SG wrote the first draft of the manuscript. UP and SG gathered the clinical information, took consent from the patient and did literature review. SP, DG and AJ contributed to the writing of the manuscript. DG provided technical assistance and critically reviewed the content of the article. DJ and AJ reviewed the histopathology slides. All authors read and approved the final version of the manuscript.

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Disclosures and Ethics

As a requirement of publication, the authors of this article have provided to the publisher signed confirmation of compliance with legal and ethical obligations including but not limited to the following: authorship and contributorship, conflicts of interest, privacy and confidentiality and protection of human and animal research subjects. The authors have read and confirmed their agreement with the ICMJE authorship and conflict of interest criteria. The authors have also confirmed that this article is unique and not under consideration or published in any other publication, and that they have permission from rights holders to reproduce any copyrighted material. Any disclosures are made in this section. The external blind peer reviewers report no conflicts of interest. A written consent was obtained from the patient to reproduce information or photographs appearing in this work.



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Supplementary Data

Figures 1–4 as a multi-panel photograph, and Figures 5 and 6 separately as another multi-panel photograph as JPG files.