Case Report

Localized morphea profunda: An atypical presentation

Arunima Dhabal[1], Ananya Chandra[1], Sumana Khan[1], Jayanta Kumar Barua[2]

From [1]Resident, [2]Associate Professor, Department of Dermatology, Venereology and Leprosy, School of Tropical Medicine, Kolkata, West Bengal, India

ABSTRACT

Morphea profunda is a variant of localized scleroderma, in which inflammation and sclerosis are found in the deep dermis, panniculus, fascia, or superficial muscle. It generally presents as diffuse, taut, and thickened skin, which is bound to the underlying tissue. Most cases have widespread involvement, but localized lesions have also been reported. We report a case of localized morphea profunda presenting with papular and vesiculobullous lesions on the overlying skin in a 50-year-old female patient who was successfully treated with methotrexate and systemic steroids.

Key words: Localized morphea profunda, Vesiculobullous morphea, Scleroderma

Morphea or localized scleroderma is a connective tissue disease characterized by sclerosis of the skin. Sometimes, the disease also involves the underlying deeper tissues. It has been estimated to have an incidence of about 4–27/million population/year with a prevalence ranging from 0.05 to 0.22% over various age groups [1]. The etiology of morphea is unknown, but environmental exposures, immune alterations, autoimmunity, familial predisposition, trauma, vaccination, and *Borrelia* infection have all been suggested as contributing factors [2]. Clinically, morphea can be classified into five main types: Plaque, generalized, bullous, linear, and deep [1].

Deep morphea encompasses a variety of clinical entities, where inflammation and sclerosis are found in the deep dermis, panniculus, fascia, or superficial muscle. Morphea profunda is a rare variant of deep morphea, with very few cases reported in the worldwide literature. The most common clinical presentation in these cases consisted of solitary or extensive atrophic or fibrotic indurated plaques usually located on the trunk [3-6]. However, we report the case of localized morphea profunda with a previously unreported presentation of papular and vesiculobullous lesions on a background of thickened hyperpigmented skin affecting the left lower leg.

CASE REPORT

A 50-year-old female presented to the outpatient department of our hospital with complaints of darkening and hardening of the skin over the left lower leg, accompanied by some elevated solid and fluid-filled lesions, and discharging ulcers. The lesions had first appeared on the left foot about 6 months back and had gradually progressed since then to involve the entire left lower leg below the knee. There was associated swelling of the left leg. The lesions were initially pruritic but later, the patient experienced pain over the affected area on walking. There was no history of preceding trauma, vaccination, or radiotherapy. The patient did not report any visible granules in the discharge. Constitutional symptoms were absent. There was no history of any systemic illness, neither a history of similar complaints in the family. The patient was not on any medication.

On examination, her vitals were within normal limits. The general and systemic survey did not reveal any abnormality. No lymphadenopathy was noted. The skin on the left lower limb from below the knee up to the dorsum of the foot, excluding the toe tips, was hyperpigmented, thickened, and indurated. There were several papules, vesicles, and bullae on the affected area. Few of these lesions had ruptured to form erosions and well-demarcated ulcers with necrotic base and seropurulent discharge. Some of the ulcers had healed with post-inflammatory hypopigmentation (Fig. 1). The affected area, especially the dorsum of the foot, was edematous. The lesions were non-tender and temperature over the left leg was normal. Peripheral pulse on the left arteria dorsalis pedis was palpable. There was no restriction of movement of the ankle joint. Examination of the surrounding skin, hair, and nails revealed no abnormality.

Her routine blood examination, including complete blood counts, erythrocyte sedimentation rate (ESR), random blood sugar, urea, creatinine, sodium, potassium, and liver function tests, was normal. Anti-nuclear antibodies, anti-double-stranded DNA, anti-topoisomerase antibodies, rheumatoid factor, and

Access this article online	
Received - 28 July 2020 Initial Review - 17 August 2020 Accepted - 19 August 2020	Quick Response code
DOI: 10.32677/IJCR.2020.v06.i08.019	

Correspondence to: Dr. Ananya Chandra, BE-36, Bidhannagar, PO - Midnapore, PS - Kotwali, Paschim Medinipur - 721 101, West Bengal, India. E-mail: ananyachandra12@gmail.com

HIV serology were non-reactive. Radiological examination and Doppler studies of the affected limb excluded muscle or bone involvement and vascular pathologies.

A deep punch biopsy was taken from the edge of an ulcerated lesion and sent for bacterial and fungal culture, both of which yielded no growth. Acid-fast stain and periodic acid-Schiff stain were also non-contributory. Histopathological examination revealed the squaring of the tissue section. The epidermis was absent as biopsy was taken from ulcerated skin. The dermis showed dense homogenized collagen bundles extending up to the subcutis, with thickened fibrous septa in the subcutaneous fat. There was a paucity of appendageal structures in the dermis. The proliferation of fibroblasts and a moderately dense infiltrate of lymphocytes, histiocytes, and plasma cells were seen on higher magnification (Fig. 2). A diagnosis of morphea profunda was made based on the typical histological appearance.

The patient was started on oral methotrexate 15 mg once a week and prednisolone 20 mg once daily, tapered by 5 mg every 2 weeks. At follow-up after 4 weeks, the lesions were found to have healed considerably with residual hypopigmentation (Fig. 3).

DISCUSSION

As mentioned earlier, morphea profunda is a rare variant of localized scleroderma, in which the inflammatory and sclerotic process primarily involves the deep dermis, subcutaneous tissue, fascia, or superficial muscle. It mostly affects middle-aged patients, with an approximately equal gender distribution [7].

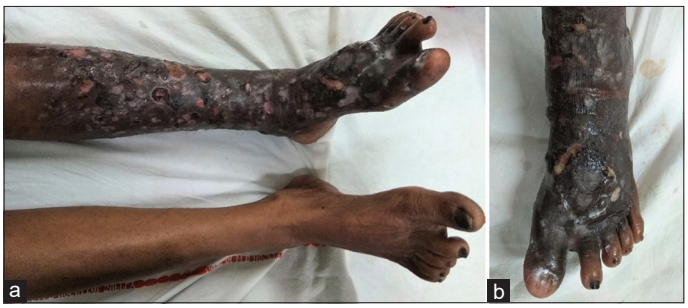

Figure 1: Left lower leg (a) and dorsum of left foot (b) showing hyperpigmented skin, with vesicles, erosions and well-demarcated ulcers, some of which have healed with post-inflammatory hypopigmentation

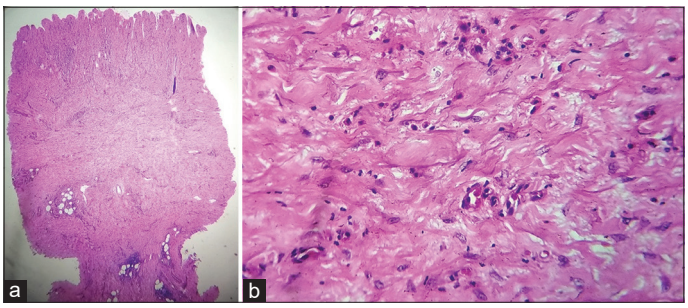

Figure 2: Squaring of the tissue section; dense homogenized collagen bundles in the dermis, extending into the subcutis with fibroblast proliferation, and inflammatory infiltrate composed mainly of lymphocytes (hematoxylin and eosin, ×40 [a] and ×400 [b])

Although a definitive etiology is unknown, morphea profunda has been reported to be associated with trauma, vaccination, radiation, and *Borrelia* infection [8-11]. No such etiology could be identified in our patient, although *Borrelia* serology was not done due to the unavailability of test facilities.

Clinically, morphea profunda most commonly presents as asymptomatic atrophic indurated plaques with hyperpigmentation. The term "morphea profunda" was first introduced in 1981 by Su and Person, who reported the occurrence of generalized inflammatory sclerosis of the panniculus or fascia in 22 out of the 23 patients that they studied [3]. These patients presented with diffuse taut, bound-down, and deep cutaneous sclerosis. Another report from 1985 described a patient with symmetrical non-tender subcutaneous indurations with brownish hyperpigmentation involving the inner thighs and lower abdomen [4].

Multiple asymptomatic hyperpigmented indurated atrophic patches in a generalized distribution in association with neurofibromas and lipomas have been described in a recent report from India [5]. Giri *et al*. also reported similar generalized atrophic lesions in a 26-year-old female [12]. On the other hand, Whittaker *et al*. reported a localized form of morphea profunda, where they described a series of patients having solitary ill-defined, indurated, and deeply tethered plaques with a peau d'orange appearance, located on the upper trunk [6]. They termed it as "solitary morphea profunda." Localized lesions were also described by Khelifa *et al*. who reported a patient presenting with non-inflammatory cupuliform depressed plaques, without any surface change or induration, on the left shoulder at the site of a previous intramuscular vaccination [13].

In our patient, the unilateral distribution of lesions localized to the left lower leg, as well as, the vesiculobullous morphology was in stark contrast to the above reports. Although there are no previous reports of morphea profunda presenting with vesiculobullous lesions, bullae have earlier been reported on lesions of plaque morphea, where it has been attributed to lymphatic obstruction [14]. A higher frequency of bullae on the lower extremities has been observed, suggesting that increased hydrostatic pressure may also play a key role [15]. This may also explain the appearance of vesiculobullous lesions in our patient.

Blood investigations in morphea profunda may show peripheral eosinophilia and raised ESR [16]. The presence of anti-nuclear antibodies is not uncommon in patients with linear and

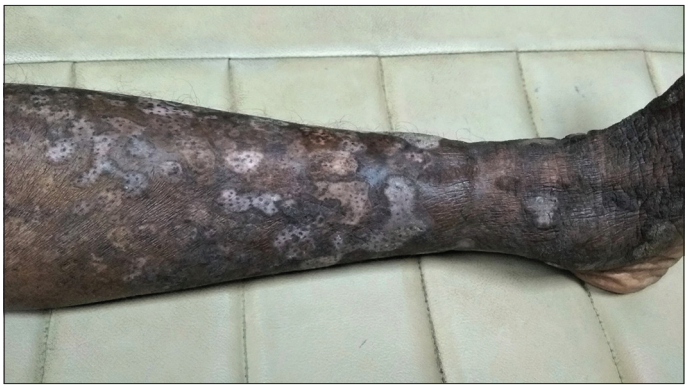

Figure 3: Healed lesions with residual areas of post-inflammatory hypopigmentation on follow-up at 4 weeks

deep morphea. Positive rheumatoid factor has been described as a risk factor for joint affection in these patients. In our patient, these investigations did not reveal any abnormality.

The characteristic histopathological features of morphea profunda described by Su and Person include thickening and hyalinization of collagen bundles in the deep dermis and subcutis, fewer appendageal structures, and the presence of a mononuclear inflammatory infiltrate consisting of lymphocytes, and plasma cells around small vessels and in the interstitium [3]. Histopathological examination in our patient revealed all these features, thereby confirming the diagnosis of morphea profunda.

Based on the clinical presentation, the differential diagnoses of mycetoma, botryomycosis, vasculopathy, and panniculitis were considered. However, these could easily be excluded by specific stains and culture from biopsied tissue, histopathological findings, and Doppler studies. Eosinophilic fasciitis, although a close histopathological differential for morphea profunda, was ruled out by the absence of both peripheral eosinophilia and involvement of muscle or fascia on magnetic resonance imaging.

The treatment of morphea profunda is not well established due to limited case reports and conflicting views regarding effective and appropriate therapeutic options. Different treatment regimens that have been discussed for all types of morphea include oral steroids, D-penicillamine, methotrexate, cyclosporine, mycophenolate mofetil, phototherapy, and extracorporeal photochemotherapy [16]. In this report, the patient responded well to a combination of oral steroids and methotrexate.

CONCLUSION

Morphea profunda is a rare condition with only a few reported cases worldwide. Our report highlights a unique presentation of this entity in the form of vesiculobullous lesions localized to an extremity. Hence, morphea profunda should be kept in mind as one of the differential diagnoses in such patients and histopathological examination should be done to either exclude or confirm the diagnosis.

REFERENCES

1. Peterson LS, Nelson AM, Su WP. Classification of morphea (localized scleroderma). Mayo Clin Proc 1995;70:1068-76.
2. Fett N, Werth VP. Update on morphea: Part I. Epidemiology, clinical presentation, and pathogenesis. J Am Acad Dermatol 2011;64:217-28.
3. Su WP, Person JR. Morphea profunda. A new concept and a histopathologic study of 23 cases. Am J Dermatopathol 1981;3:251-60.
4. Sayama K, Chen M, Shiraishi S, Miki Y. Morphea profunda. Int J Dermatol 1991;30:873-5.
5. Budamakuntla L, Malvankar D. Extensive morphea profunda with autoantibodies and benign tumors: A rare case report. Indian Dermatol Online J 2012;3:208-10.
6. Whittaker SJ, Smith NP, Jones RR. Solitary morphoea profunda. Br J Dermatol 1989;120:431-40.
7. Azad J, Dawn G, Shaffrali FC, Holmes SC, Barnetson RJ, Forsyth A. Does solitary morphoea profunda progress? Clin Exp Dermatol 2004;29:25-7.
8. Touloei K, Wiener A, Glick BP. Solitary morphea profunda following trauma sustained in an automobile accident. Cutis 2015;95:32-6.
9. Torrelo A, Suárez J, Colmenero I, Azorín D, Perera A, Zambrano A. Deep morphea after vaccination in two young children. Pediatr Dermatol 2006;23:484-7.
10. Kreft B, Wohlrab J, Radant K, Danz B, Marsch WC, Fiedler E. Unrecognized radiation-induced localized scleroderma: A cause of postoperative wound-healing disorder. Clin Exp Dermatol 2009;34:e383-4.
11. Büchner SA, Rufli T. Morphaea profunda. Hautarzt 1990;41:155-7.
12. Giri S, Paudel U, Jha A, Gurung D, Parajuli S. Morphoea profunda presenting with atrophic skin lesions in a 26 year old female: A case report. Clin Med Insights Case Rep 2012;5:57-62.
13. Khelifa E, Masouye I, Chavaz P, Hauser H, Grillet JP, Borradori L. Primary atrophic solitary morphea profunda. Dermatology 2008;217:207-10.
14. Tuffanelli DL. Letter: Lymphangiectasis due to scleroderma. Arch Dermatol 1975;111:1216.
15. Rosato E, Veneziano ML, Di Mario A, Molinaro I, Pisarri S, Salsano F. Ulcers caused by bullous morphea: Successful therapy with N-acetylcysteine and topical wound care. Int J Immunopathol Pharmacol 2013;26:259-62.
16. Careta MF, Romiti R. Localized scleroderma: Clinical spectrum and therapeutic update. An Bras Dermatol 2015;90:62-73.

Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Dhabal A, Chandra A, Khan S, Barua JK. Localized morphea profunda: An atypical presentation. Indian J Case Reports. 2020;6(8):478-480.