Short Communication

Lymphatic Obstruction as a Rare Complication of Morphea and Response to Intralesional Steroid

Ishmeet Kaur, Archana Singal, Deepak Jakhar, Deepika Pandhi

Department of Dermatology and Venereology and Leprosy, University College of Medical Sciences and Guru Teg Bahadur Hospital, University of Delhi, Delhi, India

Abstract

Morphea is a rare sclerosing disorder of the skin. Linear morphea is commonly seen in children and can affect head, neck, trunk, or limbs. It may extend to involve deeper structures such as muscles, bones, and joints. Involvement of lymphatics or the vasculature is very rare. We report a case of a 20-year-old woman presenting with linear morphea involving the nape of the neck and scalp. The lesion gradually developed two linear thick cord-like structures within the lesion of morphea because of secondary lymphatic obstruction causing restriction of neck movements. The patient was given intralesional steroid that led to softening of the skin lesions including that of cords, resulting in improvement of neck movements.

Keywords: Fibrosing disorder, intralesional steroid, lymphatic obstruction, morphea, scleroderma, steroids

Introduction

Morphea is a rare sclerosing disorder presenting with localized thickening of skin. It may extend to involve subcutaneous tissue, muscles, bones, and joints.[1] Lymphatic obstruction in morphea has been rarely reported as a bullous form of morphea. We report a case of linear morphea causing lymphatic obstruction, presenting as sclerosed lymphatic cords and causing restriction of neck movements

Case Report

A 20-year-old woman presented with a hyperpigmented depressed lesion over the back of the neck for the past 10 years. Over the past 3 months, she had noticed gradual development of two thick, linear cord-like structures within the preexisting lesion. This restricted the neck movements, especially flexion, which hampered her routine activities. No preceding evidence of inflammation, trauma, or any local injection was observed. There was a history of multiple topical treatments but without any relief. On examination, two linear hyperpigmented indurated plaques were present over left side of the posterior neck (3 × 3 cm and 7 × 10 cm) extending into the scalp with associated scarring alopecia. The larger plaque had two parallel thick linear cord-like structures fixed to the plaque [Figure 1]. Surrounding skin was apparently normal and no lymphadenopathy was observed. Severe restriction in the neck movement (lateral and anterior flexion) was reported.

Histopathology of the lesion was consistent with morphea. Ultrasonography showed skin thickening with decreased echotexture as compared to that of the surrounding normal skin. The cord-like structures had decreased echogenicity with few bleb-like structures (dilated lymphatics), suggestive of underlying lymphatic obstruction. On the basis of clinical, histopathological, and radiological features, a final diagnosis of linear morphea with secondary lymphatic obstruction was made.

The patient was started on oral dexamethasone pulse (0.1 mg/kg) along with intralesional triamcinolone acetonide (20 mg/mL) given at 3-weekly interval. In

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Address for correspondence: Dr. Archana Singal, Department of Dermatology and Venereology and Leprosy, University College of Medical Sciences and Guru Teg Bahadur Hospital, University of Delhi, Delhi 110095, India.
E-mail: archanasingal@hotmail.com

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between the injections, she was prescribed topical potent steroids. After 1 month, marked softening of the lesion was observed. After the second intralesional steroid, there was further softening as well as improvement in the neck movements. Ultrasonography showed improvement in the skin thickness as well as echotexture. Neck flexion reached within the normal limit after the 4th month [Figures 2 and 3]. The dexamethasone pulse was tapered off in 3-month time. No relapse was seen in the next 6 months of follow-up.

Discussion

Morphea is a rare sclerosing disorder of the skin. Traditional classification by Peterson *et al.* categorizes morphea into plaque (localized), linear, bullous, generalized, and deep variant, which may be pansclerotic.[1,2] Linear morphea is commonly seen in children and can affect head/neck or trunk/limbs.[1] Because of the tendency of morphea to extend deep, it is often associated with abnormalities in the underlying joints, bones, and muscles, whereas involvement of lymphatics or vasculature is quite rare. Lymphatic obstruction in morphea has been reported in few cases. In most of these, the lymphatic involvement in the lesion is presented in bullous form of morphea.[3-6] Daoud *et al.*[7] described 13 patients with bullous morphea of which 77% had underlying lymphatic obstruction. The pathogenesis of bullous morphea is still unknown, but several mechanisms have been proposed, such as inflammation, lymphangiectasia, and immune-mediated aggression. Many of these reports have suggested that compression because of the sclerotic plaque of morphea is responsible for lymphatic obstruction.[3,4,7] Fiala *et al.*[8] reported a case of linear morphea with unilateral edema. They postulated that the pathogenesis behind this complication could be lymphatic obstruction, secondary to morphea itself.[8] In all these cases, the level of lymphatic obstruction was superficial.

In our patient, the lymphatic obstruction was deep and was seen in the form of sclerosed linear lymphatic cords. To the best of our knowledge, this kind of presentation has never been reported. The lymphatic obstruction in this case is also expected to be secondary to morphea because of the temporal correlation of the onset of lesions of morphea and the appearance of lymphatic cords. The improvement in the plaques of morphea by intralesional steroids was also accompanied by softening of the cords, which strengthened the causal relationship between sclerosis and lymphatic obstruction.

Conclusion

Morphea can also lead to lymphatic obstruction presenting as sclerosed lymphatic cords, which can be associated with restriction of movement and significant morbidity. In such cases, lesion should be evaluated for

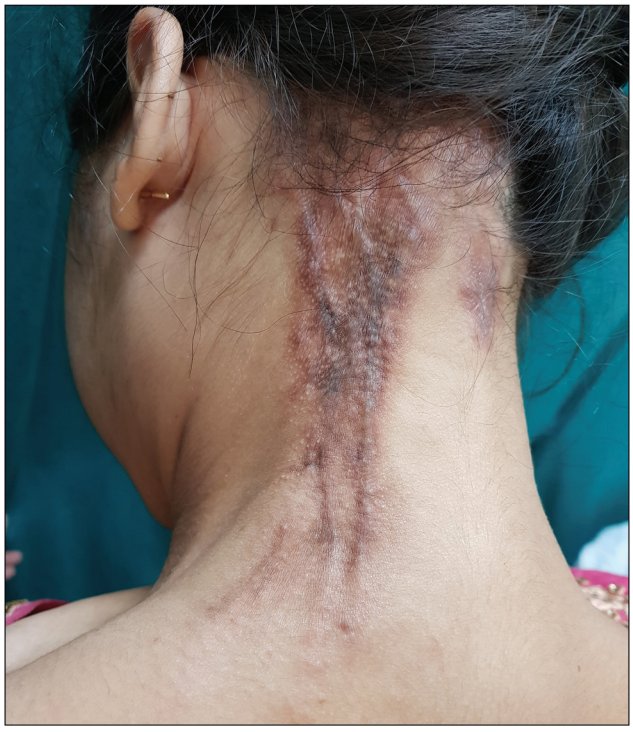

Figure 1: Two linear hyperpigmented indurated plaques over left side of back of neck with scarring alopecia. The plaque on the left side also shows linear thickened cord-like structures restricting the neck movement

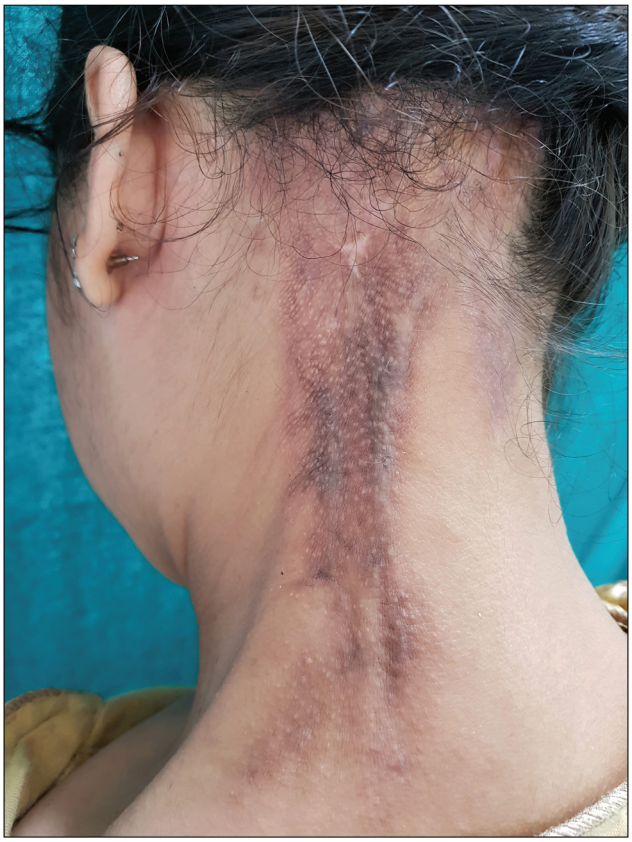

Figure 2: Lesion after 1 month of treatment showing softening of the plaque as well as the cords

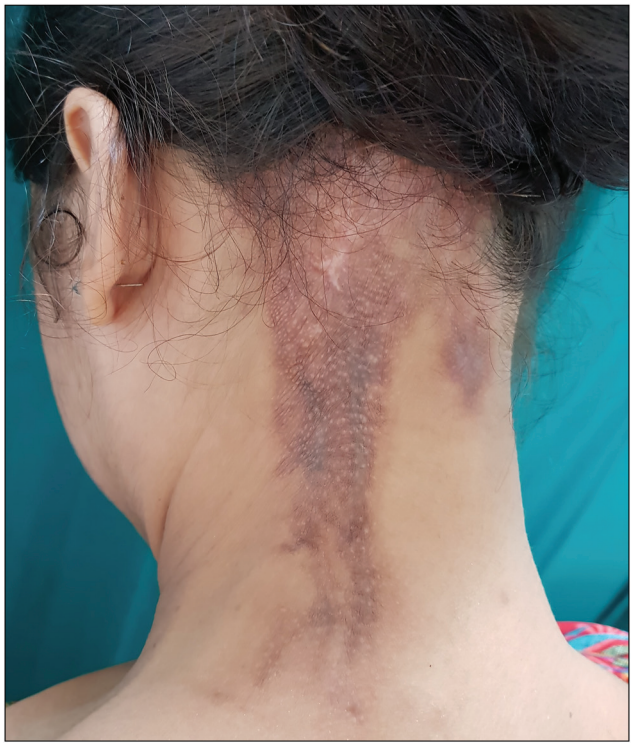

Figure 3: Further improvement in the lesion seen at 4th month of treatment with normal range of motion of the neck

underlying lymphatic involvement. Intralesional steroids may have a role in the management of the lesion as well as the lymphatic obstruction.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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