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Case Report

Multifocal linear morphea - a rare presentation with rapid progression and deformities in a young adult: a case report

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ABSTRACT

Morphea or localized scleroderma, is an uncommon skin disorder characterized by skin thickening and hardening. This case report describes a late 20s married woman working at a spinning mill, who presented with a unique and rare manifestation of morphea. Over two years, she developed a linear black color thickening of the skin on her right upper limb, left lower limb, and abdomen, accompanied by deformities and functional impairments. The patient reported a rapid progression of hyperpigmented lesions and weakness, prompting medical consultation. Cutaneous examination revealed distinctive hyperpigmented, atrophic plaques with xerotic changes and flexion deformities. A biopsy from the chest confirmed localized scleroderma. Basic investigations were within normal limits. The patient was managed with weekly pulse therapy involving corticosteroids, resulting in some relaxation of skin tightness. This case highlights the importance of recognizing and promptly treating rare multifocal morphea presentations to prevent severe deformities and enhance the patient's quality of life. Long-term follow-up is essential to monitor treatment outcomes and disease progression.

Keywords: Linear morphea, Multiple sites, Young adult, Deformities

INTRODUCTION

Morphea also known as localized scleroderma, is a rare skin disorder that is characterized by thickening and hardening of the skin. It has been associated with certain genetic factors, including specific human leukocyte antigen (HLA) class alleles. With the age of onset usually in the second decade of life, it can present as erythematous skin lesions to sclerotic change, and it can be associated with autoimmune diseases. Early diagnosis and intervention can prevent severity and improve the patient's daily life quality.[1]

CASE REPORT

A late 20s married woman, who was working at a spinning mill, presented with linear black color thickening of the skin over the right upper limb for the past 3 years. She also complained of linear black color thickening of the skin over the left lower limb for the past 1 and half years, and a linear black color lesion over the abdomen present for the past 6 months.

On eliciting history, she was normal before 3 years, then she developed a small black color skin lesion over the right hand, which gradually spread towards the forearm, arm, and back over 2 years. Later she developed weakness and inability to use the right ring and little finger for one year. She noticed similar linear hyper-pigmented skin lesions in her left lower limb one and half years back, and over the abdomen 6 months back without any weakness or restriction of movement, for these complaints, the patient consulted a physician and was using topical therapies without improvement.

Cutaneous examination

Hyperpigmented patch about 15×10 cm over the right side of the chest, which extends into the medial aspect of the arm, and over the forearm along with atrophy and xerotic changes (Figure 1a).

Hyperpigmented, atrophic plaque with xerotic changes seen over the right scapular region to the posterior aspect of the arm, forearm, and medial two-thirds of the dorsum of the hand and flexion deformities of the medial 2 fingers with dystrophic changes of the nails (Figures 1b, 2a and b).

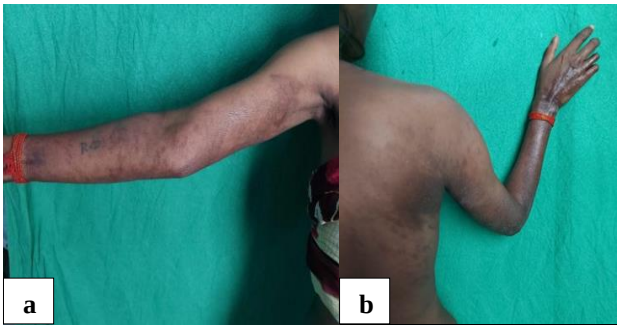

Figure 1 (a and b): Linear morphea affecting the right side trunk and upper limb.

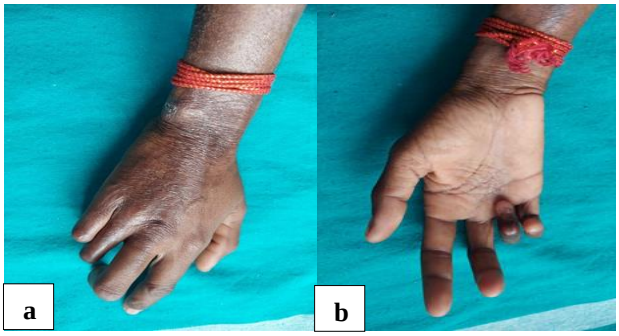

Figure 2 (a and b): Linear morphea involveing right hand with contracture of medial 2 fingers.

Hyperpigmented, atrophic plaque extending from left lumbar to iliac fossa and pubic symphysis along Blaschko's line (Figures 3a and b).

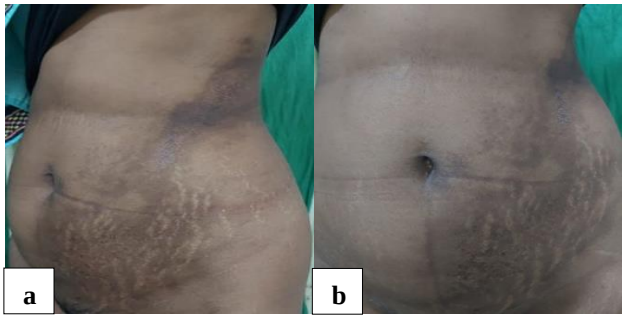

Figure 3 (a and b): Linear morphea involving abdomen left side.

Hyper-pigmented atrophic plaque with xerotic changes extends from the anterior aspect of the upper third of the left thigh, the anterior aspect of the leg to the dorsum of the foot including a tip of the medial aspect of the first three toes (Figures 4a and b). All basic investigations were within normal limits except erythrocyte sedimentation rate raised and an X-ray right hand showed bony deformities (resorption of distal phalynx of medial 2 fingers). A skin biopsy was done on the chest lesion which revealed features of morphea (Figure 5). We started on oral corticosteroids, topical emollients, and physiotherapy showed better improvement patient is now under follow-up without any progression of disease.

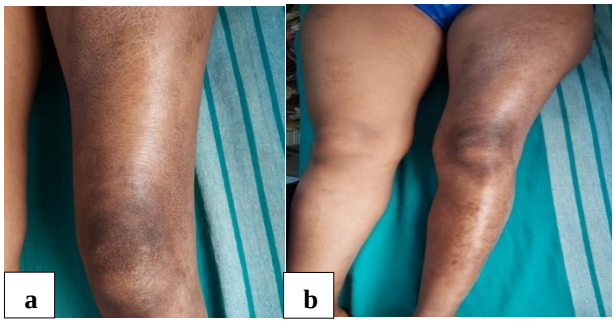

Figure 4 (a and b): Linear morphea close-up view affecting thigh left side linear morphea affecting entire length of left limb.

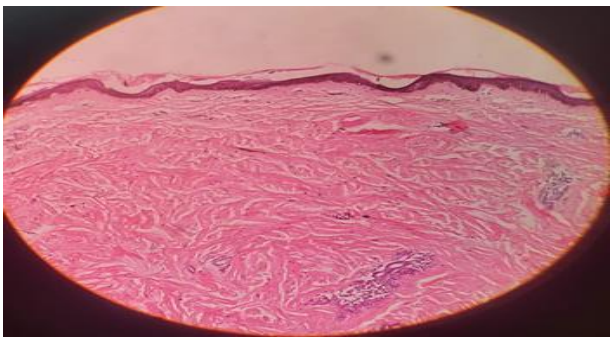

Figure 5: HPE features of linear morphea, absence of adnexal structures and hair follicles and dermis fully covered with collagens.

DISCUSSION

Localized scleroderma, is a rare skin disorder that is characterized by thickening and hardening of the skin. The European dermatology forum (EDF) classification system, as of 2017, divides morphea into various subtypes to help categorize and describe the different clinical presentations. They are generalized morphea that involves widespread skin lesions affecting multiple areas of the body. Linear morphea is characterized by band-like cutaneous lesions that typically follow Blaschko's lines. These lesions often appear on the head, neck, and limbs. Linear morphea can further be divided into two subtypes. En coup de sabre, is characterized by a linear sclerotic band on the fronto-parietal region of the scalp, which can lead to alopecia. It

typically appears as a unilateral, atrophic, and depressed groove on the forehead. Progressive facial hemiatrophy (Parry-Romberg syndrome) involves progressive facial atrophy, typically affecting one side of the face. It can lead to significant facial asymmetry.[2-5] Deep morphea involves deeper tissue involvement, including the muscles and underlying structures. Mixed Morphea occurs when both linear and plaque morphea are present simultaneously in a patient. Limited Morphea typically involves smaller, localized patches of thickened skin.[1-5]

It has multifactorial causes, morphea has been associated with certain genetic factors, including specific HLA class alleles. The following alleles have been linked to an increased risk of morphea: DRB104:04, HLA B37, HLA DR5, HLA DR8, and HLA DR11. These genetic factors suggest a genetic predisposition to the condition. Morphea also exhibits a familial tendency, with a 1.6% frequency in first-degree relatives and a higher frequency (4.7%) in twins. Additionally, there is a female preponderance, with a higher incidence in females than males, at a ratio of 2 to 3:1. Other factors may contribute to the development of morphea, which includes autoimmune mechanisms, infections like Borrelia burgdorferi (associated with Lyme disease), radiation exposure, and trauma, as seen in Parry-Romberg syndrome. It's important to note that these factors may not be the sole triggers but could contribute to the development of morphea in susceptible individuals.[1,6]

The proposed pathogenesis of morphea involves various mechanisms, depending on the triggering factor.

Radiation

Radiation exposure can lead to increased stimulation of interleukin-4 (IL-4) and interleukin-5 (IL-5), which, in turn, can promote TGF-beta-mediated fibrogenesis. This results in the accumulation of collagen and the characteristic skin thickening seen in morphea.

Autoimmune mechanisms

In autoimmune conditions, there is an upregulation of molecules like VCAM-1 and ICAM-1, which play a role in recruiting CD4 T helper cells. These CD4 T cells can further stimulate the production of interleukin-4 and interleukin-5, leading to the upregulation of fibrotic factors such as TGF-beta, PDGF, CTGF, and INLGF. This cascade ultimately results in fibroblast activation and the deposition of collagen in the extracellular matrix. It's important to keep in mind that while these mechanisms have been proposed, the exact cause and pathogenesis of morphea can vary among individuals, and further research is needed to fully understand the condition

In the early inflammatory stage, there is an erythematous skin lesion with itching and tenderness which progresses into lesions with a sclerotic center and violaceous border, followed by hyperpigmentation or hypopigmentation with atrophy of the skin and subcutaneous tissues. The active phase of this disease lasts for 2 to 5 years.

En coup de sabre (a sword stroke-like lesion) is slowly progressive and starts with contractions and stiffness in the affected area followed by a depressed groove extending into the frontoparietal region of the scalp causing linear alopecia. It is associated with bone deformities and neurological complications like seizures, trigeminal neuralgia, learning disability, and ophthalmological issues like enophthalmos, myopathy of eye muscles, anterior uveitis, keratopathy, perforation and absence or loss of eyebrows or eyelashes, if it extends into the maxilla, it may cause dental abnormalities like malocclusion of teeth, and root atrophy, temporomandibular joint involvement and also hemiatrophy of the ipsilateral tongue.[6] Progressive facial hemiatrophy also known as Parry-Romberg syndrome, is a rare neurological disorder that primarily affects the skin, underlying tissues, and sometimes the nervous system on one side of the face. It was first described by Caleb Hillier Parry in 1825 and later by Moritz Heinrich Romberg in 1846. It typically presents with gradual, asymmetric atrophy of the skin, subcutaneous tissues, muscle, and bone on half of the face. The atrophy can range from mild to severe. Minimal skin changes, such as hyperpigmentation or hypopigmentation, may be present. In some cases, it can involve the nervous system, particularly the trigeminal nerve. This involvement may result in sensory disturbances or pain on the affected side of the face. The condition progresses slowly over 2 to 20 years. After this progressive phase, the disease usually becomes stationary, and atrophy and symptoms do not worsen. En Coup de Sabre and Parry-Romberg syndrome can coexist in some individuals.[1-3,6,7]

Approximately 22% of patients with linear morphea experienced extracutaneous manifestations. These include ocular involvement, arthritis, alopecia, Raynaud's phenomenon, restrictive lung diseases, and musculoskeletal involvement. These can result in reduced growth of extremities and shortening. Neurological involvement can occur in 75% of the 22% of patients with extracutaneous manifestations. There can be an increased incidence of autoimmune diseases like type 1 diabetes mellitus, Hashimoto's disease, Graves' disease, ulcerative colitis, vitiligo, lichen planus, and pemphigus vulgaris.

Lichen sclerosis, radiation dermatitis, acrodermatitis chronica atrophicans, scars, mycosis fungoides, eosinophilic fasciitis, lipodermatosclerosis, scleromyxedema, linear atrophoderma of Moulin, restrictive dermatopathy, muckle wells syndrome, reflex sympathetic dystrophy, cheiroarthropathy due to diabetes, pseudo scleroderma conditions caused by – polyvinyl chloride, epoxy resins, pesticides, dry cleaning solvents, and silica dust are considered as differentials for morphea.[8]

Investigations usually done are: complete blood count (eosinophilia), ESR (increased in active disease), serum IgG, IgM increased in severe cases, antibodies – serum

ANA, anti-centromere antibody (elevated in crest syndrome), anti-single-stranded DNA antibody, antihistone antibody (50%), RA factor positive in 40% patients. Skin biopsy, high-frequency B scan to assess depth and extent of sclerosis. X-ray skull and long bones, computed tomography (CT) brain, magnetic resonance imaging (MRI) brain, stereophotogrammetry (head and neck involvement), and electroencephalography (EEG) wherever indicated, periodic eye examinations.[1,9-11]

In histopathology of an early stage of morphoea, the epidermis appears normal, but later there is flattening of rete ridges. There will be perivascular and interstitial infiltrations with various immune cells, including lymphocytes, plasma cells, eosinophils, and macrophages. Peri adnexal fat is reduced or disappears, indicating changes in the surrounding adipose tissue. The epithelium becomes edematous in the dermis and hypodermis. In the sclerotic stage, there is the formation of multiple collagen bundles extending into the reticular dermis. Collagen encloses the eccrine glands and blood vessels. Adipose tissue is replaced with sclerotic collagens.[12]

Treatment

In early inflammatory stages

Emollients to reduce dryness. Topical steroids to reduce inflammation and control the disease process. Tacrolimus 0.1% ointment an immunosuppressant used topically to reduce inflammation. Topical or bath PUVA- Psoralen plus ultraviolet A (PUVA) therapy can be used to treat inflammation.

For active lesions

Topical or intralesional corticosteroids, topical 5% imiquimod cream, tacrolimus 0.1% ointment twice a day for 12 to 16 weeks. Calcitriol and calcipotriol 0.005% (vitamin D analogs) can be used along with emollients to manage the disease and prevent dry skin. Oral prednisolone: systemic corticosteroid therapy may be effective in the inflammatory stage.

In the fibrotic stage

Methotrexate (15 mg per week) is an immunosuppressant used to manage autoimmune diseases. High-dose methylprednisolone may be administered for a short duration (1000 mg 3 days per month) in combination with other treatments. Methotrexate along with methylprednisolone can be used in severe cases. Mycophenolate mofetil 600 mg to 1200 mg/mt^2/day two times daily for a maximum of 6 to 36 months showed good improvement. The mean duration of MTX treatment before the start of MMF was 6.6 (range 4–15, median 7.5) months topical vitamin D analogs (calcipotriol twice a day) along with UVA1 therapy can be used to manage the fibrotic stage. Topical application of imiquimod (5% ointment/once a day for 5 days for 4 months) gives good improvement. Intralesional interferon-gamma is now under study. Phototherapy – NUVB also shows good results. Laser therapy also reduces cosmetic disfigurement.[13]

Palliative measures

Physiotherapy can be given in linear morphea associated with skeletal and muscular abnormalities like contractures. Severe facial atrophy in Parry Romberg syndrome can be corrected by palliative facial reconstructive surgeries. Some studies and clinical observations have suggested that linear morphea may have a higher risk of recurrence in individuals who develop it at a younger age (before 18 years), and compared to those who develop it later in life. In childhood localized scleroderma, a multidisciplinary approach including a rheumatologist, dermatologist, and pediatrician was necessary to prevent complications.[14,15]

CONCLUSION

Morphea or localized scleroderma, characterized by skin thickening and hardening in a late 20s married woman working at a spinning mill, who presented with a unique and rare manifestations of morphea, a linear black color thickening of the skin on her right upper limb, left lower limb, and abdomen, accompanied by rapid progression of hyperpigmented lesions, deformities and functional impairments, highlights the importance of recognizing and promptly offering the treatment for prevention of severe deformities and enhance the patient's quality of life.

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REFERENCES

1. Fett N, Werth VP. Update on morphea: part I. Epidemiology, clinical presentation, and pathogenesis. J Am Acad Dermatol. 2011;64(2):217-30.
2. Tollefson MM, Witman PM. En coup de sabre morphea and Parry-Romberg syndrome: a retrospective review of 54 patients. J Am Acad Dermatol. 2007;56(2):257-63.
3. Marzano AV, Menni S, Parodi A, Borghi A, Fuligni A, Fabbri P, et al. Localized scleroderma in adults and children. Clinical and laboratory investigations on 239 cases. Eur J Dermatol. 2003;13(2):171-6.
4. Christen-Zaech S, Hakim MD, Afsar FS, Paller AS. Pediatric morphea (localized scleroderma): review of 136 patients. J Am Acad Dermatol. 2008;59(3):385-96.
5. Pace C, Ward SE, Pace A. A rare case of frontal linear scleroderma (en coup de sabre) with intra-oral and dental involvement. Br Dent J. 2010;208(6):249-50.

6. Fledelius HC, Danielsen PL, Ullman S. Ophthalmic findings in linear scleroderma manifesting as facial en coup de sabre. Eye (Lond). 2018;32(11):1688-96.
7. Zulian F, Athreya BH, Laxer R, Nelson AM, Feitosa de Oliveira SK, Punaro, et al. Juvenile Scleroderma Working Group of the Pediatric Rheumatology European Society (PRES). Juvenile localized scleroderma: clinical and epidemiological features in 750 children. An international study. Rheumatology (Oxford, England). 2006;45(5):614-20.
8. Kreuter A, Wischnewski J, Terras S, Altmeyer P, Stücker M, Gambichler T. Coexistence of lichen sclerosus and morphea: a retrospective analysis of 472 patients with localized scleroderma from a German tertiary referral center. J Am Acad Dermatol. 2012;67(6):1157-62.
9. Ullman S, Danielsen PL, Fledelius HC, Daugaard-Jensen J, Serup J. Scleroderma en Coup de Sabre, Parry-Romberg Hemifacial atrophy and associated manifestations of the eye, the oral cavity and the teeth: a Danish follow-up study of 35 patients diagnosed between 1975 and 2015. Dermatology. 2021;237(2):204-12.
10. Schanz S, Fierlbeck G, Ulmer A, Schmalzing M, Kümmerle-Deschner J, Claussen CD, et al. Localized scleroderma: MR findings and clinical features. Radiology. 2011;260(3):817-24.
11. Abbas LF, O'Brien JC, Goldman S, Pezeshk P, Chalian M, Chhabra A, et al. A cross-sectional comparison of magnetic resonance imaging findings and clinical assessment in patients with morphea. JAMA Dermatol. 2020;156(5):590-2.
12. Chiu YE, Abban CY, Konicke K, Segura A, Sokumbi O. Histopathologic spectrum of morphea. Am J Dermatopathol. 2021;43(1):1-8.
13. Martini G, Ramanan AV, Falcini F, Girschick H, Goldsmith DP, Zulian F. Successful treatment of severe or methotrexate-resistant juvenile localized scleroderma with mycophenolate mofetil. Rheumatology (Oxford). 2009;48(11):1410-3.
14. Constantin T, Foeldvari I, Pain CE, Pálinkás A, Höger P, Moll M, et al. Development of minimum standards of care for juvenile localized scleroderma. Eur J Pediatr. 2018;177(7):961-77.
15. Zulian F, Culpo R, Sperotto F, Anton J, Avcin T, Baildam EM, et al. Consensus-based recommendations for the management of juvenile localised scleroderma. Ann Rheum Dis. 2019;78(8):1019-24.

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