Original Article

Linear Morphea – A Report of Two Cases

Sane Roja Renuka[1], G. Sukanya[2], K. Manoharan[3*], V.S. Harsha Rupak[4]

Senior Resident[1], Associate Professor[2], Professor and Head of the Department[3], Junior Resident[4]
Sree Balaji Medical College & Hospital, Chennai

Abstract

Morphea, also known as localized scleroderma, an inflammatory sclerosing disease of the skin and subcutaneous tissue. It is seen equally in children and adults. Multiple morphologies of morphea exist like guttate, plaque, keloidal, linear or pansclerotic. The incidence rate of linear morphea is 2.5/ million children per year. Hereby we report two cases of linear morphea, a15-year-oldand a 21-year-old with classical clinical and histopathological findings.

Corresponding Author: ***Prof. K. Manoharan***, *Head of Department, Dept of DVL, Sree Balaji Medical College & Hospital, Chennai. Email: drkmanoharan1957@gmail.com*

Introduction:

Linear scleroderma is one of the variant of localized scleroderma. It presents in a linear pattern with sclerotic areas of the skin. It is the second most common form of localized scleroderma. It can occur over the head, trunk, or extremities. On the limbs it may lead to contractures of the joints. En coup de sabre is a type of linear scleroderma which involves the frontoparietal area that looks like a scar of a saber cut. Central nervous system and ophthalmic involvement are rare.

Case Series:

Case Report 1:

A 15-year-old male came to the outpatient department with complaints of hyperpigmented linear patch over the right side of forehead for 9 years. The lesions started as a small hyperpigmented patch which progressed in a linear manner to current size. He had no history of photosensitivity, atopy, itching, burning, pain over the lesion. He had no similar lesion elsewhere in the body. No significant family history. On examination – hyperpigmented linear patch of size 2x8cm present over the right forehead encroaching the scalp. Mild induration noted. On seeing the lesion first, we thought it to be a facial melanosis, post inflammatory hyperpigmentation, on palpation we noted a mild induration so, 3.5mm punch biopsy was taken from the patch over the forehead and sent for histopathological examination.

Figure 1 – Clinical picture of hyperpigmented patch over the right side of forehead.

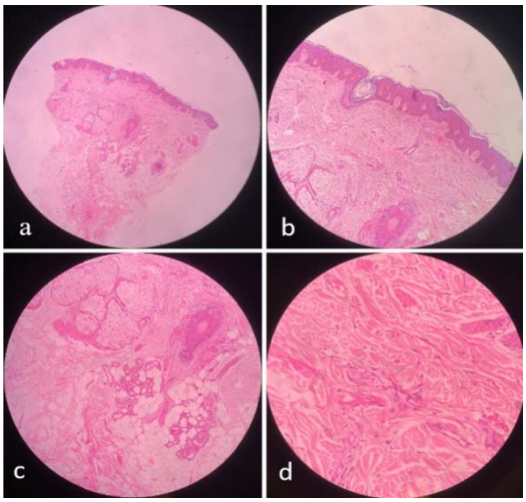

Figure 2 – H and E section
Scanner view (a) – showing mild epidermal atrophy and homogenization of collagen
Low power view (b)- showing epidermal atrophy and hair follicle
High power view (c & d)-showing high uptake of eccrine glands

HPE revealed epidermis showing hyperkeratosis, acanthosis, upper dermis shows sparse mononuclear cell infiltration around blood vessels. Numerous adnexal structures seen in lower dermis extending into fat. There was homogenization of collagen and high uptake of eccrine glands. Subcutaneous fat has large bands of fibrocollagenous tissue.

Case Report 2:

A 21-year-old female came to the outpatient department with complaints of localized scar over the forehead for the past 1 year. She had a history of trauma over the forehead 10 years back. She had no history of itching/burning/ pain over the lesion. She had no similar lesions elsewhere in the body. No significant family history. No history of photosensitivity, atopy. On examination single non tender linear atrophic scar measuring 5x1cm present over the forehead with mild induration was present. A 3.5 mm punch biopsy was taken from the atrophic scar present in forehead and sent for histopathological examination (HPE).

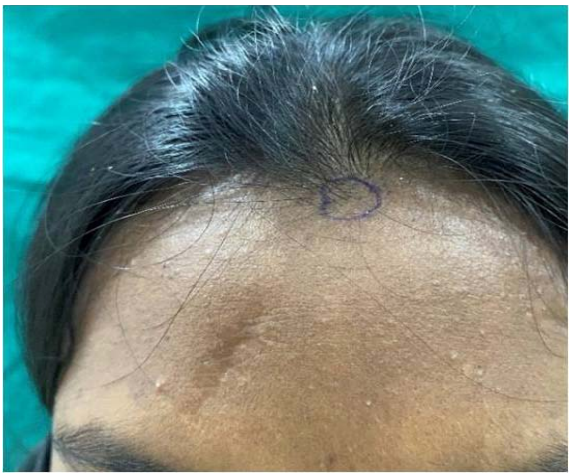

Figure 3 – clinical picture of linear atrophic scar over the forehead.

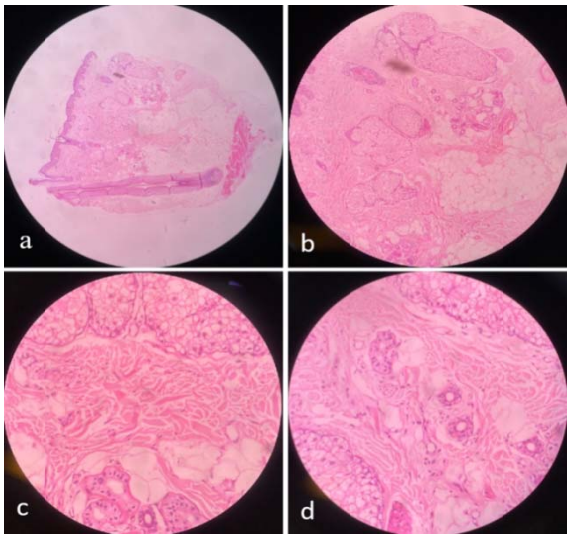

Figure 4 –H and E section
Scanner view (a) - epidermal atrophy with homogenization of collagen and prominent hair follicle.
Low power view (b) – showing hypertrophic and homogenized collagen with high uptake of eccrine glands.
High power (c & d)-showing prominent eccrine glands amid hypertrophied collagen bundles showing oasis in desert appearance.

HPE revealed epidermis with mild hyperkeratosis, acanthosis, large hair follicles

with shaft, numerous pilosebaceous units and adnexal structures near papillary dermis. Collagen bundles are seen distributed between the adnexal structures extending from the dermo epidermal junction, dermis and homogenization of collagen seen. There wereprominent eccrine glands amid hypertrophied collagen bundles showing oasis in desert appearance.

Discussion:

Morphea has different types of clinical phenotypes and EDF classification (European Dermatology Forum) has mentioned the predominant subtypes as generalized, limited, linear, deep, and mixed type[1,2]. Major causes of Morpheaincludes genetic predisposition and autoimmune dysregulation. HLA class II allele DRB104:04 and class I allele HLA–B37 are the strongest genetic associations with morphea. Environmental factors such as trauma, radiation, and frictionalso play a role in morphea. TGF-B mediated fibrogenesis is induced through localized secretion of interleukin 4 and 5 by radiation[3,4]. Linear morphea presents as band-like lesionsover the head, neck and limbs. The sclerosis is localized to the dermis. Deeper tissues can also be involved causing complications like weakness of muscle, contracture of joints, and deformities of limbs in children. Morphea en coup de sabre is a type of linear scleroderma that primarily affects the head and neck areas. The name is derived from its linear shape and its to a wound from a saber. The involved skin and tissues have an excessive deposition of collagen and sclerosis may be present. The involved areas may progressively atrophy. It may present as a hyperpigmented orhypopigmented linear, indurated plaque with atrophic, depressed and ivory appearance[5]. In scalp and eyelids, it may lead to alopecia. It can also progress into deeper tissues causing atrophy of muscle, nerve damage,bone and teeth demineralization[6]. Inorder to confirm morphea some patients may require a skin biopsy, including subcutaneous fat. If deeper skin tissues are involved an incisional biopsy can be done to get an appropriate sample. There is no specific antibodies for morphea but there are some cases reported with positive Anti Histone ab, ANA and SS DNA. Laboratory investigations such as peripheral eosinophilia and elevated inflammation markers may help in the diagnosis. Patients with positive ANA and Raynaud's symptoms, should be screened for systemic sclerosis. Ultrasound is good for monitoring the regular disease, in case of severe lesions Magnetic Resonance imaging (MRI) can be used to look for the depth of the lesions and findings of any involvement of the fascia, tenosynovitis, or myositis that can cause any deformities. In treating this condition for superficial lesions topical corticosteroids are the first line of management. In cases of widespread involvement phototherapy with UVA1 can be used. For severe and rapidly progressing lesions systemic corticosteroids can be used. For relapsing conditions combination therapy of corticosteroids and Methotrexate can be used as first line. Surgical treatments like autologous fat grafting can be used in case of en coup desabre[7]. Before considering it to be a morphea one must rule out other conditions such as lipodermatosclerosis, scleromyxedema and post irradiation morphea.

Conclusion:

Morphea is an inflammatory, sclerosing disease of the skin and subcutaneous tissues.

Here we are presenting a case series of morphea to emphasise the need to consider any case of atrophic scar or hyperpigmented lesion as of case of morpheaand thorough clinical and histopathological examination is required for ruling it out. If left untreated, morphea lesions can lead to cosmetic complications which needs expensive cosmetic corrective procedures. In children it can cause severe atrophy of the extremities with contractures of joint. To limit the disease progression and its consequences early intervention is necessary.

References:

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