ORIGINAL RESEARCH PAPER

Volume-7 | Issue-1 | January-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

AN UNUSUAL MANIFESTATION OF LOCALIZED SCLERODERMA - A CASE REPORT

Dermatology

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ABSTRACT

Scleroderma presenting as localized sclerosis of skin is popularly known as morphoea. Cutaneous manifestations of which may vary clinically. Linear localized scleroderma is less commonly reported and the nodular variant of a linear localized scleroderma is even rarer. In nodular scleroderma, patients develop lesions that are clinically indistinguishable from a keloid; however, the histopathological findings are more variable. Here in we describe a 17-year-old male with a linear raised plaque on the central forehead of 6 years duration. The histological examination revealed findings suggestive of a hypertrophic scar beside localized scleroderma.

KEYWORDS

Nodular scleroderma, Linear localized scleroderma, Keloidal localized scleroderma

INTRODUCTION:

Localized scleroderma or morphoea is a chronic disease of an unknown etiology which manifests as sclerotic areas of induration of skin, often surrounded by a pigmented lilac halo.[1] Of many types of morphoea classified, linear morphoea also known as linear localized scleroderma is often noted as sclerotic areas of skin usually arranged in a linear pattern, often following the Blaschko's lines.[2]

Linear morphoea (linear localized scleroderma) is commonly reported in childhood and is seen commonly over the limbs. When involves the fronto-parietal region of face, it is famously known as en coup de sabre. Apart from atrophy of the skin, it involves the underlying muscles and bones, eyes and central nervous system. Association with Parry-Romberg syndrome (Progressive facial hemi atrophy) is common and it is difficult to clinically differentiate either. Lack of sclerosis of skin and coursing along the branches of the trigeminal nerve in latter can provide clues to the diagnosis. Some suggest that both the conditions are variants of linear morphoea and share a common pathogenesis.[3]

Nodular scleroderma also erroneously known as keloidal morphoea, is a rare presentation of morphoea. It presents as, well-defined sclerotic papules and nodules, which usually can occur with systemic sclerosis, generalised and localized morphoea.[4] Even rarer is nodular morphoea reported to occur in a linear pattern, 1st reported by Sylvia Hsu et al in the year 1999.[5]

We report a case of linear nodular morphoea involving the central forehead in a 17-year-old male, of 6 years duration, who also had development of keloid in a remote area over a tattoo mark.

CASE REPORT:

A 17-year-old male reported to the OPD with complaints of dark raised skin lesion, relatively asymptomatic, of 6 years duration over the central forehead. It was initially small in size and a flat lesion, later increased in its length to reach the frontal hair line with an increase in thickness. Examination revealed a single linear pigmented plaque of size 4 cm long and 1 cm wide, over central forehead, extending between the glabella and the frontal hair line. [Figure 1] Palpation revealed a firm indurated nodule, confined within the lower 2/3rd of the pigmented plaque. The nodule was mobile in side to side direction and the overlying skin was not pinchable. No extension to the scalp, loss of hair and underlying bony defect was noted. Facial symmetry and oral examination were normal. A keloid over right arm following tattooing was present. Differentials considered were morphoea and nevus lipomatous superficialis.

The general and systemic examination were normal. Anti- Nuclear antibody and Anti- Topoisomerase 1 antibody was negative. Skin biopsy revealed, mildly atrophic epidermis. [Figure 2.A] Superficial dermis showed increase in cellularity with whorls of homogenous collagen bundles. [Figure 2.B] The deeper dermis showed homogenous, hyalinized, hypertrophied, oedematous and compact collagen replacing the subcutis. The eccrine gland, is in a falsely higher position, giving an 'oasis in the desert' appearance. A few peri-eccrine gland infiltrate was noted. [Figure 2.C] Special stain for mucin was done and was found to be negative. [Figure 2.D]Thus the histopathology was conclusive to the diagnosis of localized scleroderma developing a hypertrophic scar.

Currently, the patient is on a course of doxycycline 100 mg twice daily along with pyridoxine 40 mg daily. Latter, we are planning for intralesional steroids.

DISCUSSION:

Development of nodular lesion in morphoea is widely misinterpreted as a keloidal morphoea and there is no clear line of demarcation. Attempts have been made to delineate the two as the 'nodular' type and 'keloidal' type with some pros and cons.[6,7] Keloids, by definition is an area of overgrowth of fibrous tissue which usually can occur spontaneously or follows a skin injury and which often extends beyond the original defect. Histologically, they are characterized by nodules and whorls of dense, thickened, glassy, eosinophilic collagen bundles with decrease in cellularity and vascularity and by presence of mucin in the dermis. On the other hand, the hypertrophic scar usually is confined within the site of original defect and histopathologically characterised by been more cellular and vascular than keloids with absent mucin deposition.[8] Dermal mucinosis in a nodular linear morphoea along the Blaschko's lines is reported and its occurrence may be due to the mucin deposits seen in the keloids.[9]

Development of keloid or hypertrophic scar with in the morphoea lesion is thought to be as a keloidal response to an early inflammatory component of scleroderma in patients who are either genetically at the risk for developing a keloid or in the areas of skin that have a high predilection for the development of keloid (such as face, chest and shoulders). High levels of tenascin, TGF-β and connective tissue growth factor has been observed in keloidal scleroderma lesions, suggesting an active fibroblastic activity, which is analogous to that seen in scleroderma.[4,6]

Clinically, nodular scleroderma is more common in female gender and the onset of nodular lesions is reported to occur usually after the development of localized scleroderma. Cases with pre-existing keloidal tendency and external triggers are reported.[4,6]

Treatment options include use of systemic methotrexate, intralesional and systemic steroids, D- penicillamine, azathioprine and PUVA therapy, with varied outcomes.[4,6]

In our case, the patient neither had keloid like clinical findings nor had histopathological findings consistent to that of a keloid. Rather, both clinically and histologically features were suggestive of hypertrophic scar with a background of linear morphoea.

CONCLUSION:

This case of linear nodular localized scleroderma is reported for its rarity and emphasizes the need for a correlation of clinical to histopathological findings. Further insight is required to understand the pathogenesis and treatment of this rare variant of scleroderma.

ACKNOWLEDGEMENT

None

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

LEGENDS TO FIGURE:

Figure1: Clinical photograph showing linear nodular localized scleroderma on the central forehead.

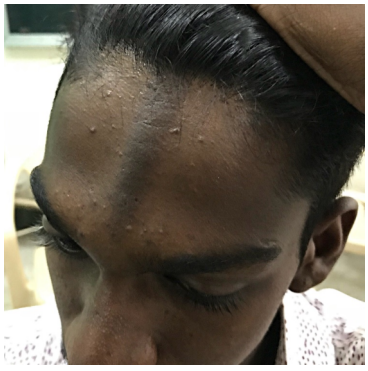
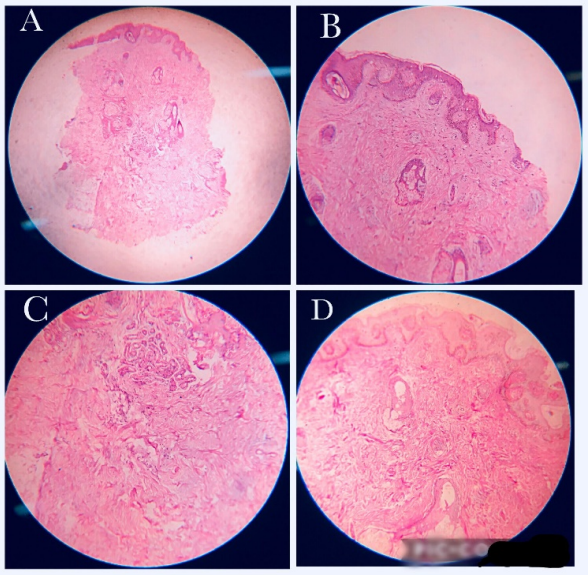


Figure 2: A. Photomicrograph showing mildly atrophic epidermis and dense collagen bundles in both superficial and deep dermis. B. Low power view (10 X) showing whorls of collagen bundles in superficial dermis with increase in cellularity. C. Low power view (10 X) showing newly formed homogenized, hyalinized, oedematous and compact collagen in deep dermis with peri-eccrine gland infiltrate. D. Special stain (PAS), showing no mucin deposits.



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