Case report

Coexisting morphea with lichen sclerosus et atrophicus in a single lesion- a rare case report

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Abstract:

Background: Lichen sclerosus et atrophicus (LSA) is an inflammatory dermatitis of unknown etiology affecting usually genital region with rare involvement of extragenital regions like face, neck, shoulder etc. Coexistence of lichen sclerosus et atrophicus and morphea is a very rare finding. Although LSA at early stage and morphea can be differentiated clinically and histologically, both diseases at late stage present diagnostic difficulties. ***Case presentation:*** We reported a 4 year old female child presented with a non pruritic hypopigmented sclerotic patch over left shoulder. Histopathology showed features consistent with both LSA and morphea in the same lesion. ***Conclusion:*** Coexistence of both LSA and morphea in same patient suggests that these lesions represent a spectrum of diseases which may reflect similar events or closely related pathologic process.

Keywords : LSA , Morphea , Scleroderma, Hypopigmented patch

Bangladesh Journal of Medical Science Vol. 15 No. 01 January'16. Page : 145-147

Introduction:

Lichen sclerosus et atrophicus (LSA) consists of flattened white macules which tend to coalesce to form white patches[1]. It is an inflammatory dermatitis of unknown etiology, usually affecting genital regions like vulvar, perianal and perineal skin of prepubertal, perimenopausal and postmenopausal women. Extragenital involvement though rare, includes most commonly face, neck, shoulder and upper part of trunk[2]. Histologically the lesion consists of pronounced edema and homogenization of collagen in the upper dermis [3]. On the other hand morphea is characterized by a circumscribed, sclerotic plaque with an ivory coloured centre and histologically by sclerosed collagen bundles in the dermis[4]. Morphea and LSA have been described separately both clinically and histologically, however there are very few reports in the literature quoting simultaneous occurrence of both these diseases, hence proposing a link between them [3]. We report a rare case of extragenital LSA along with morphea in a single lesion of a patient with some peculiar presentations.

Case report:

We report a case of 4 year old female presenting with a linear non pruritic, slightly indurated hypopigmented to depigmented sclerotic patch over left shoulder gradually enlarging and extending to involve index finger. Clinical examination revealed contracture of the left elbow and areas of cigarette paper wrinkling suggestive of atrophy of the epidermis. Genital areas were normal. Routine examination including hemogram, urine and stool examination were within normal limits. Biopsy was done from the lesion. Histopathology showed coexistent features of both LSA and morphea in the same lesion. Microsections from the lesion showed trilaminar appearance consisting of 1) Compact hyperorthokeratosis, 2) Atrophic epidermis with basal vacuolar degeneration and follicular plugging 3) Pale upper dermis with edema and loss of elastic lamina. These features were consistent with LSA (Fig 1a,1b,1c,1d) whereas lower reticular dermis showed thickened hypocellular collagen bundles, with focal subcutaneous involvement (Fig 2a, 2b) .Special stain masson trichrome and van gieson

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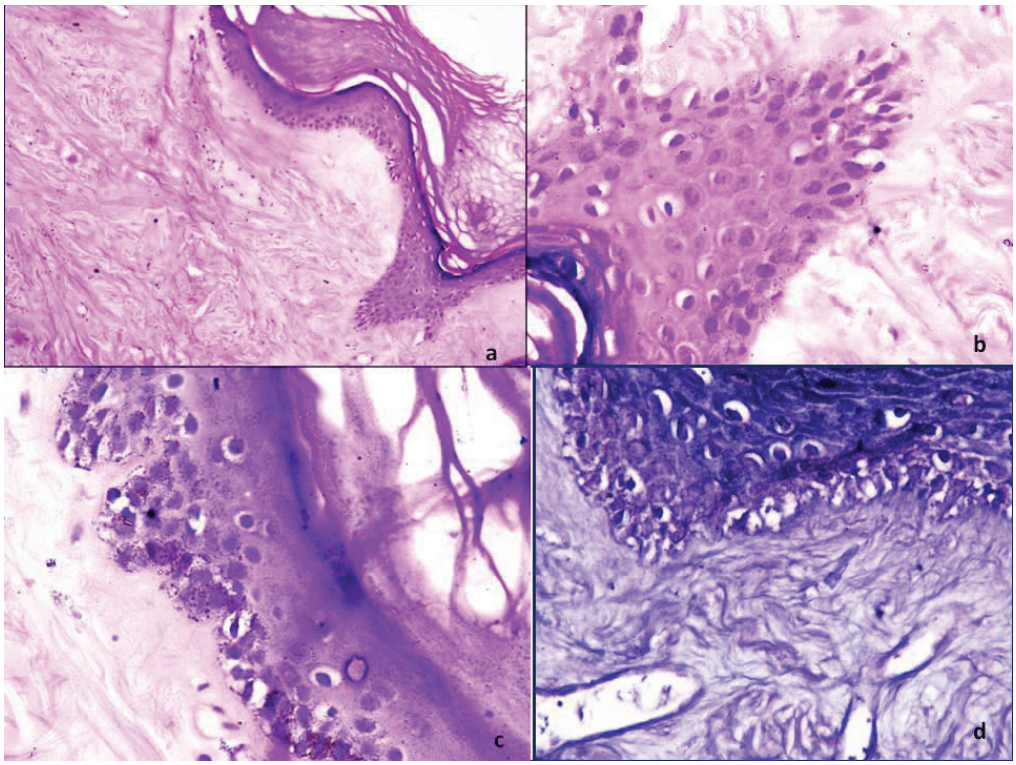

Figure 1 a) shows trilaminar morphology at the right upper part (orthokeratosis, atrophic epidermis & upper dermal pallor) along with loss of rete ridges(H & E ;40x). b) shows basal vacuolar degeneration in the same lesion (features of LSA) (H & E; 100x). c)shows basal vacuolar degeneration (H & E ;400x). d)Loss of elastin fibres in the upper dermis (VGE ; 100x)

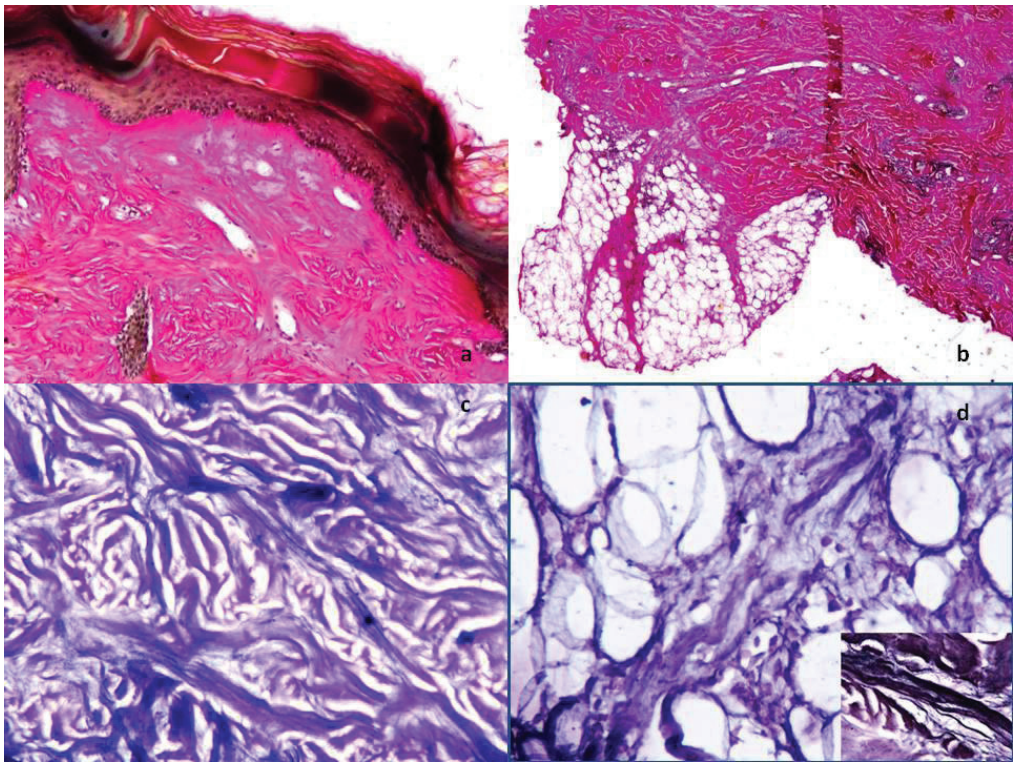

Figure 2 a) Shows pallor in the papillary dermis (MT stain ;40x) .b) shows thickened collagen bands in the reticular dermis with mild inflammation(Morphea) (H & E ; 400x). c) shows thick elastic fibers lying parallel to the thickened collagen bundles (Morphea) (VGE; 400x). d) Shows elastic tissue stain in the subcutaneous tissue (VGE; 400x), inset shows prominent elastic fiber (VGE; 1000x oil)(Morphea)

elastin stain revealed thick elastic fibres parallel to collagen bundles, features consistent with morphea (Fig 2c, 2d).

Discussion:

Till date there have been only a very few cases reported in the literature describing coexistent features of LSA and morphea[5], because the simultaneous presentation of different skin diseases in a single patient is uncommon. Sometimes it may represent a diagnostic challenge for the clinicians[5]. Generalized morphea and LSA are now classified under localized scleroderma by Peterson et al [6]. Scleroderma is a group of chronic autoimmune diseases with thickening of the skin as a hallmark of the disease. And it encompasses two groups-localized and systemic scleroderma. Generalized scleroderma is characterized by widespread sclerosis of the skin where plaques become confluent affecting more than two body sites. Sometimes it may develop as an extension of localized morphea or can be associated with LSA[7].

Our patient had a linear non pruritic hypopigmented to depigmented sclerotic patch over left shoulder, gradually extending towards index finger over 3 years. There was no systemic involvement which supports the benign nature of the disease. Extension of the lesion over the elbow joint and dorsal surface of the hand was associated with formation of elbow contracture. This could be due to an acute inflammatory process causing fibrosis of the affected site. Various radiological changes like periarticular erosion, osteopenia and flexion deformities have been reported with localized and generalized morphea[8].Flexion deformity was noted in our patient. There are very few cases reported in the literature describing simultaneous presentation of both LSA and morphea in the same patient, however coexistence of these lesions in a single lesion involving extragenital site is extremely rare [9]. Morphea and LSA may have similar clinical presentations. Overlapping clinical and histopathological features of both conditions have been described in the literature, which have led to speculate that they represent different features along the same disease spectrum [10]. It has been postulated that there is a common etiologic agent involving autoimmune pathways for both the lesions[10]. LSA is now categorized as subepidermal morphea [8].

Our patient had few unusual presentations which are difficult to explain in spite of good history and clinical examination and seems worth reporting. Therefore to conclude, morphea and LSA may coexist in a same patient and in spite of different clinical and histomorphological presentations, simultaneous occurrence of both conditions in the single lesion indicates overlapping similar possible etiopathogenesis.

Authors declare no conflict of interest

References:

1. Nishioka S. Histological comparison of morphea and lichen sclerosus et atrophicus. *Kurume Med J*. 1997;**44**(2):83-90.
2. Meffert JJ, Davis BM, Grimwood RE. Lichen sclerosus. *J Am Acad Dermatol*. 1995;**32**:393–416.
3. Tremaine R, Adam JE, and Orizawa M. Morphea coexisting with lichen sclerosus et atrophicus. *Int J Dermatol* 1990; **29**:486-489.
4. Rowell NR, and Goodfield MJD. The connective tissue disease. In: Textbook of Dermatology, 5th ed. Champion RH, Burton JL and Ebling FLG, Blackwe Scientific Publications, London, pp 2163-2294, 1992.
5. Connelly MG, Winkelmann RK. Coexistence of lichen sclerosus, morphea and lichen planus. *J Am Acad Dermatol* 1985; **12**: 844-51.
6. Paterson LS, Nelson AM, Su WP. Classification of morphoea (localized scleroderma). *Mayo Clin Proc* 1995;**70**:1068-76.
7. Patterson JA, Ackerman AB. Lichen sclerosus et atrophicus is not related to morphoea: A clnical and histologic study of twenty-four patients in whom botn conditions were reputed simultaneously. *Am J Dermatopathol* 1984;**6:**323-35.
8. Virdi SK, Kanwar AS. Generalized morphea, lichen sclerosis et atrophicus associated with oral submucosal fibrosis in an adult male. *Indian J Dermatol Venereol Leprol* 2009;**75:**56-9
9. Uitto J, Santa Cruz DJ, Bauer EA, Eisen AZ. Morphea and lichen sclerosus et atrophicus: Clinical and histopathologic studies in patients with combined features. *J Am Acad Dermatol* 1980;**3:**271-9.
10. Rose AE, Boyd KP, Meehan SA, Latkowski JA. Lichen sclerosus et atrophicus. *Dermatol Online J*. 2013 Dec 16;**19(**12):20714.