ORIGINAL RESEARCH PAPER

Dermatology

A CLINICAL STUDY OF GENITAL AND EXTRAGENITAL LICHEN SCLEROSUS ET ATROPHICUS

KEY WORDS: Extragenital, Koebnerization, Lichen sclerosus et atrophicus(LSA).

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ABSTRACT

Lichen sclerosus et atrophicus (LSA) is an uncommon entity that commonly affects the genital region but cases of extra genital LSA have been reported. We report eight cases of extragenital LSA and twelve cases of genital LSA. All cases of extragenital LSA presented with porcelain white macules and follicular papules. Koebnerization was observed in all cases of extragenital LSA. All cases were diagnosed clinically and supplemented with histopathology where there was follicular plugging, interface dermatitis and homogenization of dermal collagen. Cases are reported for their rarity and their varied presentation.

INTRODUCTION

Lichen sclerosus et atrophicus (LSA) is an uncommon chronic inflammatory disease of unknown aetiology with genital and/or extragenital involvement.[1] It predominantly affects the anogenital area with white porcelain-like sclerotic lesions.[2] Extragenital lichen sclerosus is rare, mostly affects the neck and shoulders, axilla, upper arms, wrists and around umblicus[3]. This present study was conducted to know the various manifestations of genital and extragenital LSA.

METHODS

The study was conducted for a period of 2 years from January 2015 to December 2017. 20 patients who presented with clinical features of LSA attending OPD of our hospital were included in the study and the diagnosis of LSA in all cases was made based on the clinical features and few cases were supplemented with histopathological findings. All the patients were interrogated for a detailed history and a meticulous examination of each case carried out and recorded giving special emphasis to the duration of the disease, age of onset, precipitating factors and any other cutaneous or systemic illness. Apart from routine blood and urine examination, dermascopy and biopsy was done in 6 out of 8 extragenital LSA cases.

RESULTS

A total of 20 patients who presented with LSA were included .The age group of patients ranged from 8 years to 55 years . Males (60%) were affected more commonly than females (40%), thereby giving a male: female ratio of 1.5:1. 8 out of 20 cases had exclusive extragenital LSA while 12 cases had genital LSA.

In the patients with genital LSA, the lesions were pruritc/burning, white or red with ill defined borders and a patchy appearance predominantly affecting the prepuce and the areas of the glans covered by the prepuce in males (figure 1). On the other hand in females, only the opposed surfaces of the vulva were seen to be predominantly affected (figure 2) in such a manner that no lesions were visible unless the labia were separated manually.

The extragenital LS lesions were asymptomatic, pink to ivory white, coalescing macules or patches with well-defined borders and follicular oriented papules (figure 3).5 out of 8 cases of extragenital LSA showed koebnerization at trauma prone sites (figure 4).

There was no loss of sensation, scalp or nail involvement, and no systemic co-morbidities. Family history was insignificant. Complete hemogram, liver and renal function tests and blood sugar levels were seen to be within normal limits. Dermoscopy of extragenital LSA showed multiple whitish structureless areas, some surrounded by an erythematous halo, with the presence of telangiectasia and comedo like openings (figure 5). Skin biopsy was done in 6 out of 8 extragenital LSA cases with a differential diagnosis of Vitiligo and Atrophic lichen planus. Histopathology revealed hyperkeratosis, follicular plugging and irregular acanthosis in the epidermis, dense collagenisation in the upper dermis. Basal layer showed vacuolization with deeper dermis showing dense aggregates of chronic inflammatory cells in perivascular region and between collagen bundles (Figure 6 & 7). On the basis of clinical and histopathological grounds the diagnosis of genital and extragenital lichen sclerosus was established in all the above cases.

DISCUSSION

Lichen sclerosis is a chronic uncommon inflammatory dermatoses of unknown ethiology first described clinically by Hallopeau in 1887 and histopathologically described by Darier in 1892. [4] It has an association with specific HLA types and various autoimmune diseases which suggests that LSA is an autoimmune process. [4]Role of extracellular matrix protein 1 has been implicated in recent years. [5]

It is predominantly seen in women with female to male ratio of 6:1. It is most commonly seen in the genital area, but can also occur over the extragenital areas. Approximately 15-20% of cases of anogenital LSA also involve the extragenital areas, while exclusive extragenital area involvement is seen in 2.5% of all LSA cases. [6] Extragenital LSA are usually asymptomatic hence often underestimated. [5]

Koebner phenomenon is a known association with Lichen Sclerosus. Surgical operation, infections, burn scar, injection sites, sexual abuse, tight clothing, and radiotheraphy are all causes of koebnerization in LSA.[7]Extragenital lesions can commonly occur in damaged areas and preexisting scars. Boyd and Neldner have classified Lichen Sclerosus et Atrophicus as occasionally occurring Koebner phenomenon.[8] The presence of Koebner phenomenon in a patient with LSA is important as it indicates the presence of active disease. It is an indication that various irritants, surgeries have to be avoided.[9]

Diagnosis of LSA is usually clinical and may be supplemented with histopathology examination and dermoscopy. Dermoscopy can serve as a non invasive, reliable tool for diagnosis as well as help in predicting the activity of the disease process.[10] Dermoscopy of recent LSA lesions characteristically shows homogeneous whitish structure-less areas surrounded by erythematous halo and presence of yellow circles (comedo like openings) that correlate with follicular plugging and orthohyperkeratosis on histopathology. While older lesions shows no halo with poorly-differentiated diffuse margins.[11]

Histopathology is a reliable way to confirm the diagnosis. All our cases had common findings of epidermal atrophy, follicular plugging and interface dermatitis. Histopathology in early and old cases may considerably vary and late cases may not show all these characteristic findings. Selection of biopsy site is crucial when extensive lesions are seen and multiple biopsies may be taken.

CONCLUSION

We report this study because of the varied manifestations of genital and extragenital LSA and rarity of exclusive extra-genital LSA. 12 out of 20 cases were males which is also uncommon. Koebnerization was seen in all cases of extragenital LSA which is a rare occurrence. LSA should be considered as a differential diagnosis when a patient presents with atrophic depigmented macules with koebnerization. The use of dermoscopy to aid in the diagnosis is essential.

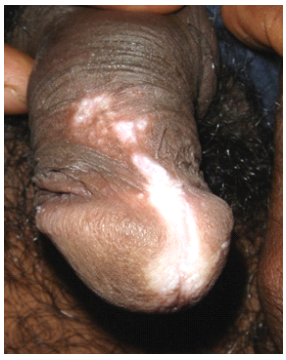

Figure 1 Depigmented macules over the prepuce and glans penis

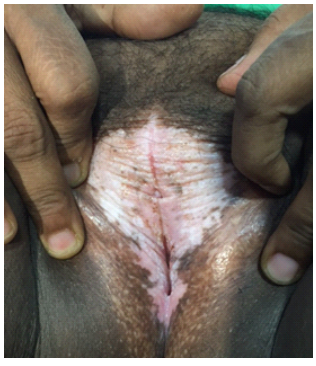

Figure 2 Depigmentation and sclerosis over the vulva

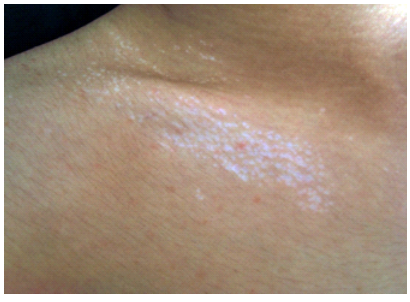

Figure 3 Depigmented macules and follicular oriented papules were seen over the right shoulder with koebnerization

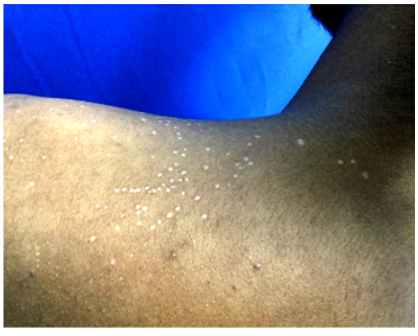

Figure 4 Linear lesions of LSA suggesting koebnerization

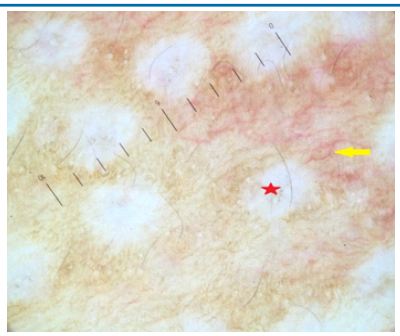

Figure 5 Dermoscopy showing whitish structureless areas (red star) with telangiectasia (yellow circle) of varied lengths and comedo like openings

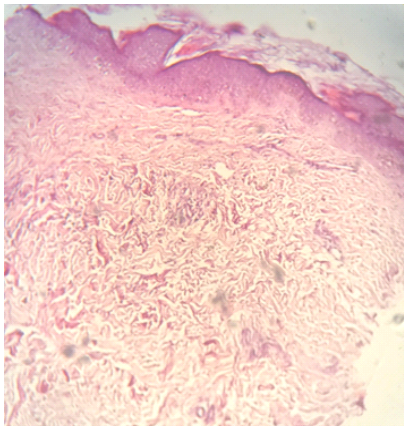

Figure 6 Epidermal hyperkeratosis, follicular plugging with dense collagenisation in the upper dermis. Basal layer showed vacuolization with inflammatory infiltrate in dermis [H and E, x10]

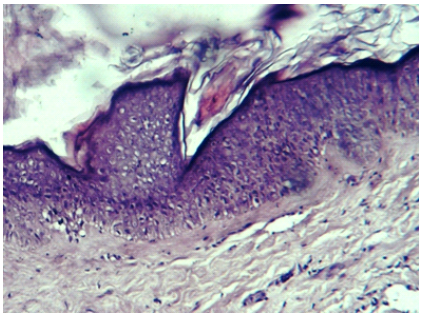

Figure 7 Histopathology showing follicular plugging and vacuolar degeneration of the basal layer [H and E, ×40]

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