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SEGMENTAL EXTRAGENITAL LICHEN SCLEROSUS ET ATROPHICANS - A RARE ENTITY.

Dermatology

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ABSTRACT

Lichen Sclerosus Et Atrophicans is a disease entity belonging to the group of sclerotic atrophies. Segmental distribution of Lichen Sclerosus Et Atrophicans is a rare presentation. Here we report a case of segmental lichen sclerosus et atrophicans. **Summary** :We report this case in view of its extragenital presentation involving the trunk with unilateral distribution in zosteriform pattern which is a rare entity

KEYWORDS

Lichen Sclerosus Et Atrophicans , Segmental distribution , Extragenital involvement .

INTRODUCTION

Lichen Sclerosus Et Atrophicans is a chronic inflammatory skin condition affecting the perineal and perianal regions in pre-pubertal and perimenopausal women. It can occasionally also appear as extragenital lesions.

The incidence of extragenital Lichen Sclerosus Et Atrophicans is around 15 to 20 %. The etiological factors are usually idiopathic, immunological, hormonal in addition to genetic predisposition. The autoimmune process is usually aided by antibodies against extracellular matrix protein 1, a significant building block in the dermis.

Segmental pattern of lichen sclerosus et atrophicans is due to genetic mosaicism where two distinct cell clones are present. The generation of segmental lesions is primarily due to stimulation of abnormal clone by immunologic or environmental factor .

Case Report:

A 52-year-old female presented to the dermatology OPD with the complaints of asymptomatic yellow colored flat lesions over the lower back on left side for the past 5 yrs. They initially started as single macular lesions and progressed to involve the segment T10 -T11 at the back of the trunk on left side .

The patient had no similar lesions elsewhere in the body. There was no history of similar complaints in the family. There was no history of herpes zoster prior to the onset of lesions . The patient did not have any prior treatment for current clinical presentation. There were no associated co – morbidities.

On examination, multiple atrophic macules were seen over the lower back at the level of T 10 – T11 on left side with few lesions extending to right side. Satellite lesions were present on left side of trunk. Scalp, oral mucosa and nails were normal. Punch biopsy was taken from atrophic macule over the left side of lower back and sent for histopathological examination and it revealed loss of rete ridges in the epidermis , the upper dermis showed pink homogenous eosinophilic collagen with scattered perivascular mononuclear cell inflammation. The patient was started on topical tacrolimus and was lost to follow-up.

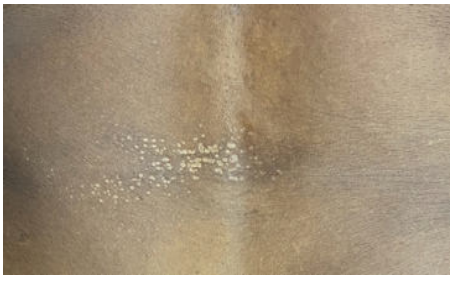

Figure 1 : clinical picture showing yellowish atrophic macules over the back of the trunk on left side with few lesions extending to right side at the level of T10 -T11

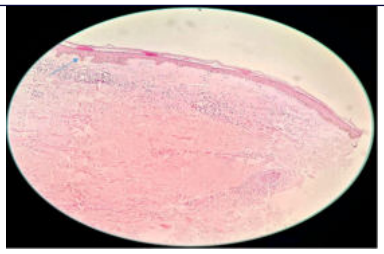

Figure 2a – HPE picture showing features as loss of rete ridges in epidermis

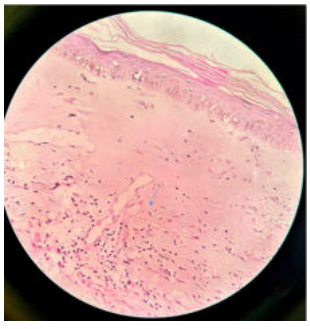

Figure 2b - HPE picture showing pink homogenous eosinophilic collagen with scattered perivascular mononuclear cell inflammation in upper dermis consistent with lichen sclerosus Et atrophicans

DISCUSSION

In case of Extragenital Lichen Sclerosus Et Atrophicans, it is usually asymptomatic and the common sites of involvement are the trunk , proximal extremities and neck. Occasionally pruritus is present. Less common sites include scalp and face.

Diagnosis is primarily clinical but biopsy is needed for atypical extragenital presentation Histologically, LSA has atrophic epidermis , loss of rete ridges with lichenoid infiltrate at the dermo epidermal junction. In early lesions, homogenization of collagen is seen with papillary edema eventually leading to sclerosis LSA can be disfiguring as it is a progressive disease. Early treatment usually includes high potent topical corticosteroids. Calcineurin inhibitors can be used to avoid the long term side effects of topical corticosteroids. Other treatment options include phototherapy and oral retinoids. Carbon di oxide laser, cryotherapy and focused ultrasound can be used with varying rates of success.

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Conflict Of Interest: Nil

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