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LICHEN SCLEROSIS ET ATROPHICUS MASCQUERADING AS VITILIGO- A CASE SERIES

Dermatology

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ABSTRACT

Lichen sclerosis et atrophicus (LSEA) is a chronic inflammatory disease which predominantly affects the anogenital region which impairs the quality of life due to intense itching. The disease can also affect other areas of the body which maybe asymptomatic unlike the genital lesions. Here we report a case series of 3 patients with LSEA whose clinical presentation mimicked vitiligo, but confirmed upon histopathology and detailed history. The patients were treated with potent topical steroids and tacrolimus and there was improvement in the symptoms and started resolving.

KEYWORDS

CASE SERIES

CASE 1:
A 28-year-old female presented to the OPD with complaints of multiple hypopigmented patches over the back and left arm for the past 10 years. The patches slowly progressed to increase in size over the past 5 years. There were no constitutional symptoms associated with the lesions. Patient had taken treatment in the form of topical and oral antifungals with no response.

The general and systemic examination was normal. Cutaneous examination revealed multiple hypopigmented patches on the upper back and extensor surface of the left arm. There were few pigmented areas admixed within the lesions. No scaling was present. Oral and genital mucosa was normal.

Biopsy from the lesion in the back revealed epidermal atrophy with flattening of the rete ridges, basal cell degeneration, sub-epidermal clear zone and homogenization of collagen in the papillary dermis. These histopathological findings were suggestive of LSEA.

CASE 2:
A 10-year-old female child presented to the OPD with complaints of hypopigmented patches over the lateral aspect of the right chest and the other patch over the flexor aspect of the right forearm for the past 2 years. The lesions were asymptomatic except for occasional mild itching.

The general and systemic examination was normal. Cutaneous examination revealed a well-defined depigmented patch with few areas of pigmentation present within the patches. Another hypopigmented patch was present over the flexor aspect of the right forearm. No scaling was present in both the lesions. Biopsy was taken from the chest lesion and features were suggestive of LSEA.

CASE 3:
A 45-year-old female came with the chief complaints of depigmented patches over the vulva for the past 8 years. The patient had consulted physicians and was given topical treatment, but there was no improvement. There was minimal itching in the lesion. Oral mucosa was normal.

Cutaneous examination showed depigmented patch over the labia majora and minora. There were few pigmented areas within the patch. Biopsy was performed and features were suggestive of LSEA.

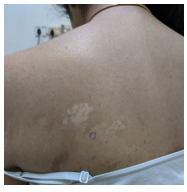

FIGURE 1: CASE 1

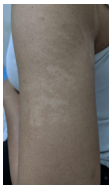

FIGURE 2: CASE 1

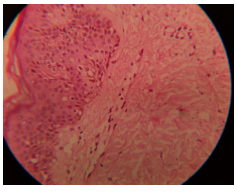

FIGURE 3: CASE 1

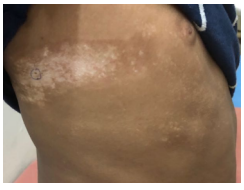

FIGURE 4: CASE 2

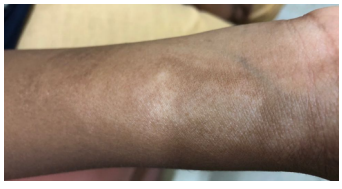

FIGURE 5: CASE 2

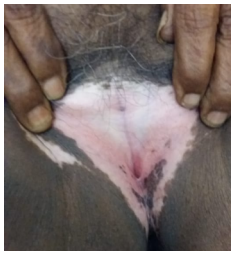

FIGURE 6: CASE 3

DISCUSSION:

LSEA occurs at all ages and in both sexes with female to male ratio being 5:1.[1] It usually affects females in the post-menopausal age group, though it can also affect children in the age under 10 years.[2]

The exact cause of LSEA is unknown. A genetic predisposition can be attributed as there are high rates of familial LSEA cases being reported. Association with HLA class II antigen DQ-7 was observed. Auto-antibodies against extracellular matrix protein 1(ECM 1) can be found in 80% of the patients. Evidence linking infectious agents like acid-fast rods, spirochetes or Borrelia to the development of lesions is lacking.[3]

The disease predominantly affects the anogenital region presenting as polygonal papules and porcelain-white patches with atrophic skin, telangiectasias, erythema, scaling and varied amount of sclerosis in the anogenital region often in the figure-8 pattern in females. Genital lesions can lead to intractable pruritus, soreness, dyspareunia, dysuria and if the involvement extends to the anal orifice can lead to difficulty in defecation. With progression, obliteration or synechiae of the labia minora and clitoris can occur and finally leading to obliteration of the introitus.[4]

In males, LSEA usually affects prepuce and glans penis, leading to phimosis most commonly later giving rise to priapism.

Extragenital sites are neck, trunk and proximal extremities are common affected.

The differential diagnosis to be considered are vitiligo, lichen planus, contact dermatitis and child abuse for genital lesions. For extragenital

lesions, the differentials are morphea, pityriasis versicolor, vitiligo, anetoderma and other causes.

The classical histopathological findings are hyperkeratosis with follicular plugging, epidermal atrophy with flattening of the rete ridges, basal cell degeneration, sub-epidermal clear zone and homogenization of collagen in the papillary dermis

Treatment consists application of super-potent topical steroids limited for 4-6 weeks and continued with mid potent steroids and calcineurin inhibitors like tacrolimus.[5] Systemic therapy with retinoids, penicillin, chloroquine and methotrexate with pulsed steroids can be tried.

CONCLUSION:

LSEA can easily be confused with vitiligo and other pigmentary disorders. Proper examination with particular attention given to palpate the lesions to appreciate the atrophy and histopathological examination usually leads to the diagnosis. Early diagnosis is warranted as the condition significantly impairs the quality of life and in the long term, malignancies may develop in it.

LEGENDS TO THE FIGURE:

FIGURE 1: Hypopigmented patch over the back
FIGURE 2: Hypopigmented patch over the left arm
FIGURE 3: HPE showing revealed epidermal atrophy with flattening of the rete ridges, basal cell degeneration, sub-epidermal clear zone and homogenization of collagen in the papillary dermis
FIGURE 4: Hypopigmented patch over the chest
FIGURE 5: hypopigmented patch over the forearm
FIGURE 6: Depigmented patch over the vulva

CONFLICTS OF INTEREST: NONE

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