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A FEMALE WITH UNILATERAL FACIAL DISCOID LUPUS ERYTHEMATOSUS-AN UNUSUAL PRESENTATION: A CASE REPORT IN BANGLADESH

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ABSTRACT

Lupus erythematosus (LE) is a chronic autoimmune disease with a diverse array of clinical illness among which Discoid Lupus Erythematosus (DLE) is one of the commonest forms involving the integumentary system mostly.Usually, classic DLE lesions exhibit some common features like discoid shape, dyspigmentation, adherent scale and atrophy, but sometimes theseclassic scenario may be absent. In case of atypical presentation, clinical assessment of the disease may aid in early diagnosis and prompt treatment of the condition. This would help prevent disfigurements, progression to systemic disease and poor quality of life. In our case, the patient presented with unilateral, asymptomatic plaque on her left malar region for last 15 years. The diagnosis was delayed due to unusual presentation and final conclusion was drawn by means of histopathological examination.

Keywords: Atypical DLE, Unilateral, Asymptomatic, Histopathology.

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INTRODUCTION

Lupus erythematosus (LE) is defined as a chronic autoimmune disease with a broad range of cutaneous and systemic manifestation which predominantly affects the skin and joints[1]. Chronic cutaneous LE has different variants among which discoid lupus erythematosus (DLE) is more common with typical presentation but some other morphologies are also found such as periorbital DLE, comedogenic DLE and hypertrophic DLE[2]. This condition most frequently affects the sun exposed areas of the body, specially face and scalp and characterized by well circumscribed, erythematous, scaly patches or plaques with atrophy, scarring and pigmentary changes[3]. In our case, the patient presented with unilateral, asymptomatic, erythematous plaque on her left malar region which differs from classic presentation by lacking pronounced atrophy, scarring and pigmentary changes. This less classical presentation of the case delayed the diagnosis.

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CASE PRESENTATION

A 37-year-old female presented with an asymptomatic, well-defined, unilateral erythematous plaque on her left malar region since 15 years. The plaque was about 5×1 cm in diameter. On examination, the plaque is elevated with slight scaling and atrophy was present at one border (Figure 1 A). She had no history of photosensitivity, pruritus and no other exacerbating factors. Rest of the integumentary examination including hair and nail revealed no abnormality. She did not have any other systemic complaints. With above mentioned complaints, she visited different dermatologists and applied various creams. However, there was no improvement and previous physicians could not reach a firm diagnosis. When she visited our Outpatient Department with above mentioned scenario, we decided to perform biopsy for histopathology considering the differentials of atypical presentation of DLE, Lupus Vulgaris and Sarcoidosis.

Her routine hematological and other biochemical investigations revealed normal findings and anti-nuclear antibody was negative. An incisional biopsy from lesional skin showed epidermal atrophy, hyperkeratosis and follicular keratotic plugging. Histopathology also revealed vacuolar degeneration in the basal layer. It also showed superficial, as well as, deep perivascular and periadnexal lymphocytic infiltrate. There was also pigmentary incontinence in the dermis (Figure 2).

Figure 1 A: The patient with unilateral facial DLE before treatment

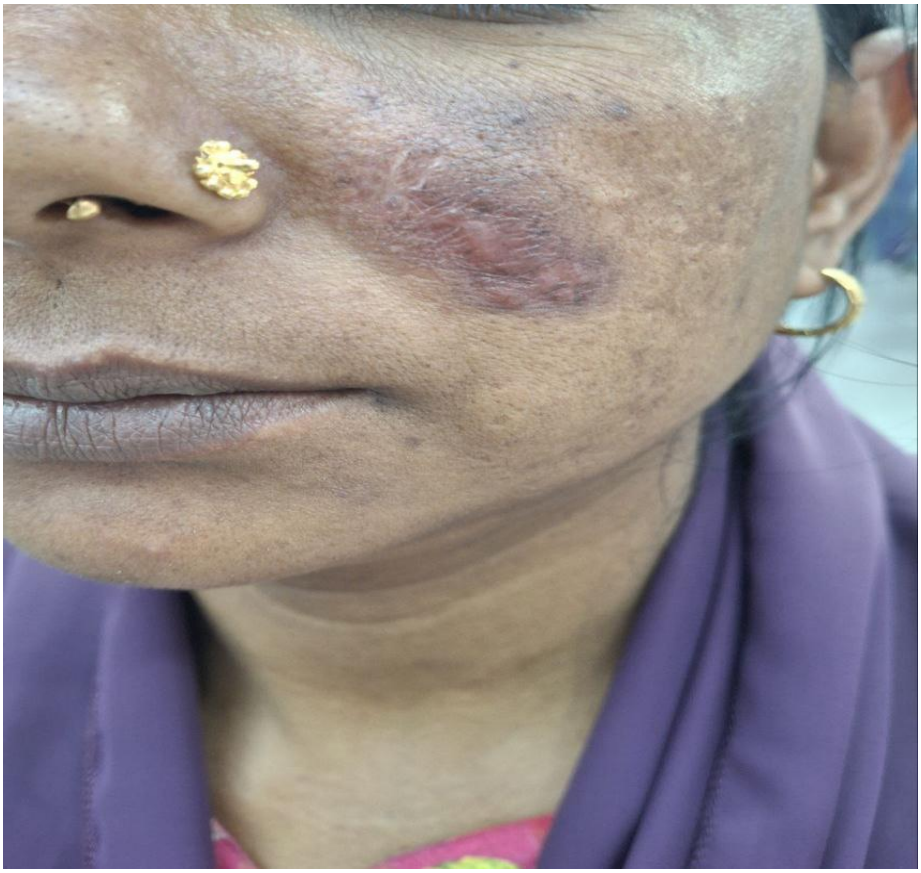

Figure 1 B: The patient with unilateral facial DLE before and after treatment

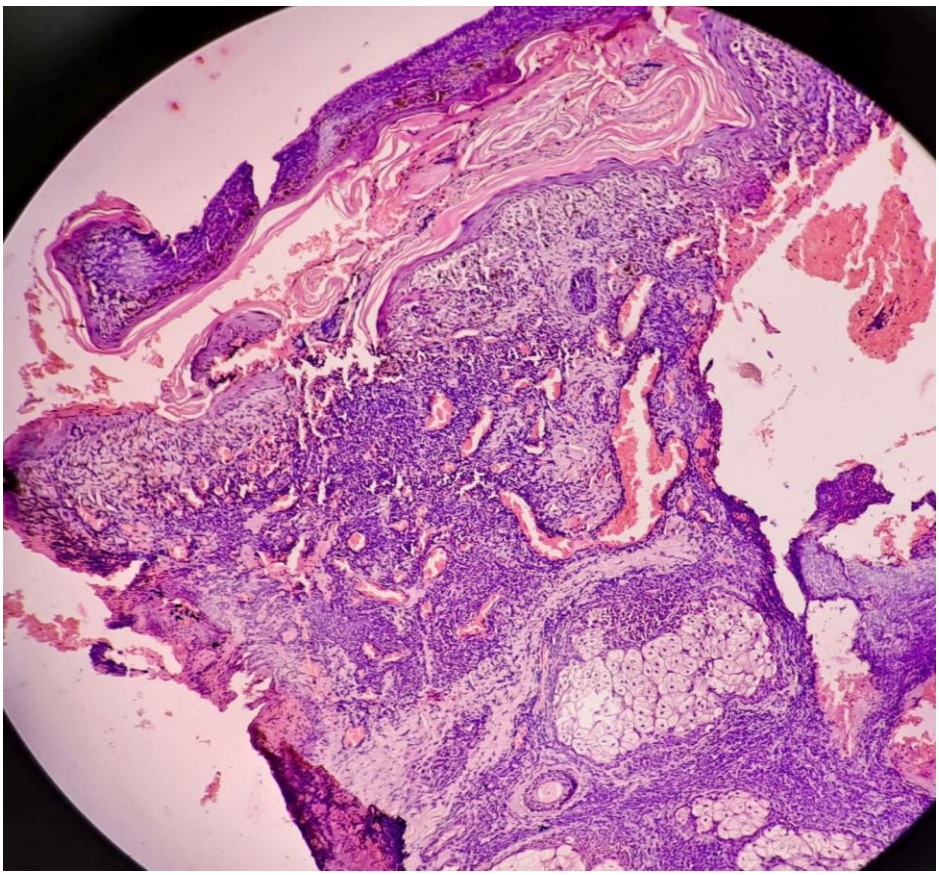

Figure 2: Microscopic section of the lesional skin of the patient

DISCUSSION

DLE is a chronic variant of cutaneous LE, commonly develops on sun exposed areas with different morphological pattern, causing diagnosis to be perplexing at times[4]. It usually affects young adults, with female outnumbering male 2:1. DLE more commonly occurs in African Americans, but are also found in Asian and white population. Genetic predisposition, sunlight exposure, cigarette smoking may play role in the development of DLE, though the pathophysiology of the disease is not elucidated properly[5]. Lesions are localized above the neck usually with favored sites of scalp, bridge of the nose, malar areas, lower lip and ears; hence is termed as localized DLE[6]. Generalized DLE is more commonly associated with underlying Systemic Lupus Erythematosus and affects commonly thorax and upper extremities, althoughthey can occur at any site of the body. Lesions of palms and soles can be painful[7]. Typical lesions begin as dull red macules or indurated plaques that develop an adherent scale, then evolve with atrophy, scarring and change in pigmentation. In darker complexion, lesions typically demonstrate areas of both hyperpigmentation and depigmentation; whereas in lighter skin patients the plaques may appear gray or have minimal pigment alteration[6].

In about 24% of DLE patients, mucosal surface is affected, involving the mouth, nose, eyes or vulva[5], rarely aggressive squamous cell carcinoma may arise; specially in longstanding cases[6].

If treatment is not started earlier, scarring, atrophy, hypopigmentation may lead to permanent disfigurement[8]. Its chronic nature impacts quality of life of the patients and increases the psychological burden and high morbidity[4]. Sometimes lesion with uncommon presentation mimicking other dermatoses may lead to difficulty in clinical diagnosis; in such cases, lesional biopsy for histopathology and Direct Immunofluorescence are conclusive. In our case, patient was suffering for last 15 years, as definitive features like discoid shape, scarring, hypopigmentation were absent. So, we decided to perform histopathology keeping DLE as one of our differentials. Ultimately histopathology report revealed epidermal atrophy, hyperkeratosis and follicular keratotic plugging. There was also vacuolar degeneration in the basal layer, superficial and deep perivascular and periadnexal lymphocytic infiltrate and pigmentary incontinence in dermis-all these features are consistent with discoid lupus erythematosus. Treatment was given with sun protection, tablet hydroxychloroquine and topical corticosteroid application. Her condition has been gradually improving (Figure 1 B shows the comparison of her condition before and after treatment. Marked improvement was observed following treatment).

The purpose of reporting this case is to establish definitive diagnosis of such unusual presentation of DLE with proper investigative evaluation rather than applying any medication.

CONCLUSION

DLE is a chronic disease with varied morphological appearance. Sometimes it may progress to SLE specially in disseminated DLE patient. Localized DLE may be complicated by pigmentary changes and scarring; so effective sun protection as well as early diagnosis and management is mandatory to prevent such consequences.

CONFLICT OF INTEREST

There is no conflict of interest.

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