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A report on neglected case of systemic lupus erythematosus presenting by Degos' skin disease and diffuse non-scarring alopecia with dramatic response to treatment

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ABSTRACT

In this work a case of female patient (38 years old) suffered from Degos disease, non-scarring alopecia with digital ulcer occurring in the setting of systemic LE was reported. A dramatic hair regrowth was observed with the current treatment protocol. Also, the potential effectiveness of this treatment was discussed.

Keywords: Systemic Lupus Erythematosus, Diffuse non-scaring alopecia, Degos disease.

INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disease with hair loss is a common symptom of (SLE), affecting 17.3%-85.2% of patients. Potentially reflecting SLE disease activity. Alopecia in SLE can be scarring, or nonscarring. It is associated with some devastating psychosocial consequences and may cause depression, anxiety beside poor quality of life (Chen *et al.*, 2003; Lipsker, 2012; Ferraz *et al*., 2006; Gilhar *et al*., 2012; Udompanich *et al*., 2018; Chanprapaph *et al*., 2019)

Degos is described as erythematous papules with a core atrophic lesion that is porcelain white. Coagulopathy, endothelial cell injury, autoimmune disorder, and vasculitis are the causes. Approximately 15% of Degos disease cases are a benign form often limited to the skin without gastrointestinal or CNS involvement (Ramos-Casals *et al*., 2006).

Patients with connective tissue disease like SLE may develop degos-like lesions, due to the extensive overlap in clinical characteristics and histological findings that are indistinguishable from those of cutaneous lupus erythematosus in skin lesions from Degos disease **(**Ball *et al*., 2003).

In this study a female patient was diagnosed to be suffered from Degos disease, non-scarring alopecia with digital ulcer occurring in the setting of systemic LE. She had the required treatment and the results were discussed.

Results

Case report:
No. one patient
Sex: female
Age: 38 years old

Laboratory investigations:

The results of the patient showed the presence of the followings:

- Hypocomplementaemia with complement 3= 65 mg/dL (Normal 90 -

180 mg/dL) and complement 4= 2.5 mg/dL (Normal 9 - 36 mg/dL),
- Elevated titer of anti-nuclear antibodies (ANA 170 EU/ml,
- Speckled immunofluorescent pattern (Normal up to 40.0)
- Positive Anti-nuclear antibody
- Indirect immunofluorescence (ANA-I.F.),
- Positive anti-ds-DNA antibody, anti-Ro antibody, and anti-La antibody.
- Anti-phospholipid syndrome antibodies were normal
- Anti-cardiolipin Ab (IgG) = 7.2 GPL U/ml (Normal Up to 23.0),
- Anti-cardiolipin Ab (IgM) = 3.0 MPL U/ml (Normal Up to 11.0),
- Lupus Anti-coagulant (LA) =37 (Normal 34 – 44).
- Complete Blood Cell Count showed pancytopenia,
- Liver enzymes were within Normal.
- Renal function test showed Hyperalbuminuria: 60.0 mg/dL, Creatinine: 70.0 mg/dL, Albumin/Creatinine ratio: 857.14 mg Alb./gm (Normal <30).
- Urine Analysis: Pus cells 22-25/HPF, RBCs 1-3/HPF.
- Heart echocardiography showed Normal LV systolic function, Grade I diastolic dysfunction with Mild Mitral Regorge I/IV.

Diagnosis:

- Hypertensive
- Non-diabetic,
- Suffered from ischemic heart disease,
- With a diffuse hair loss, recurrent pruritic skin rash, malar rash, scaly Erythematous patches on face and eyebrow and arthritis but lacked signs of CNS vasculitis, such as hemiparesis and impaired vision, or symptoms involving the gastrointestinal tract. Her neurological examination was uneventful. By dermoscopic view of scalp by Dermlite DL4 showed Follicular Plugging with perifollicular brownish scales, Erythematous background (Fig. 1).
- The Patient also had multiple atrophic papules all over the trunk and extremities with black crust which on removal revealed central porcelain white atrophy and a peripheral rim of erythema. By dermoscopic view, white stellate structureless areas surrounded by a rim of short vessels (Fig. 2)

Treatment:

After investigation of the present case the treatment was done, and the diagnosis was confirmed as being active systemic lupus erythematosus. Treatment was started with Hydroxychloroquine 200 mg twice daily, prednisolone 0.5 mg/kg per day, aspirin 75 mg/day and avoidance of sun exposure. After one week the patient came with gangrenous lesion on the index finger's tip then we started therapy with Mycophenolate mofetil 500 mg once daily and we treated the Alopecia with Minoxidil gel 5% twice daily, topical retinoid daily once at night and Caffeine containing shampoo. The patient's lesions showed marked improvement and hair restored after 3 months of treatment (Fig. 3).

DISCUSSION

Degos disease in the skin is initially manifested with erythematous, pink or red papules that heal to leave scars with pathognomonic, central, porcelain white atrophic centers and usually have a peripheral telangiectatic rim(1)(Udompanich et al., 2018). Ball et al (2003) proposed that Degos disease is just a variant of lupus, while Whitney et al [4] 2004 declared that Degos disease may not be a specific entity due to the great overlap in its clinical and histological findings but, rather, it may represent a common end point to a variety of vascular

insults, where many of which have not been fully elucidated.

Dermoscopically, Papules are characterised by a reddish-purple backdrop and purpuric spots in the early stages. At the intermediate stage, papules have a targetoid pattern with necrotic centers and erythematous halos. Finally, healed lesions have a rim of short, curved vessels surrounding a pale, structureless center [(3)](Anker et al., 2014). Vasculopathy can appear as Degos-like lesions and be etiologically implicated in the pathophysiology of Degos disease when it occurs as an acute or subacute manifestation of lupus [(2)].(Ball et al, 2003).

In the current patient, neither blood fibrinolytic activity nor aberrant platelet aggregation were found. She had positive anti-La antibody and hypocomplementaemia with complement 3 and 4 at the time of diagnosis. Assuming that anti-La antibody is related to the prevalence of vasculitis in SLE patients [(5)](Ramos-Casals et al., 2006), according to clinical, laboratory and dermoscopic examination, we suspected vasculitis in our patient, which can cause Degos-like skin lesions which were associated with SLE as a reaction pattern that resulted from peripheral artery blockage brought on by vasculitis related to SLE or our patient had hidden vasculopathy with seronegative. Based on our experience with this case, when pathognomonic Degos-like lesions appear as porcelain white scars with an erythematous rim, after carefully reviewing the clinical history and undergoing physical and laboratory tests for SLE, a clear diagnosis should be determined and detect the associated internal organs diseases early as possible.

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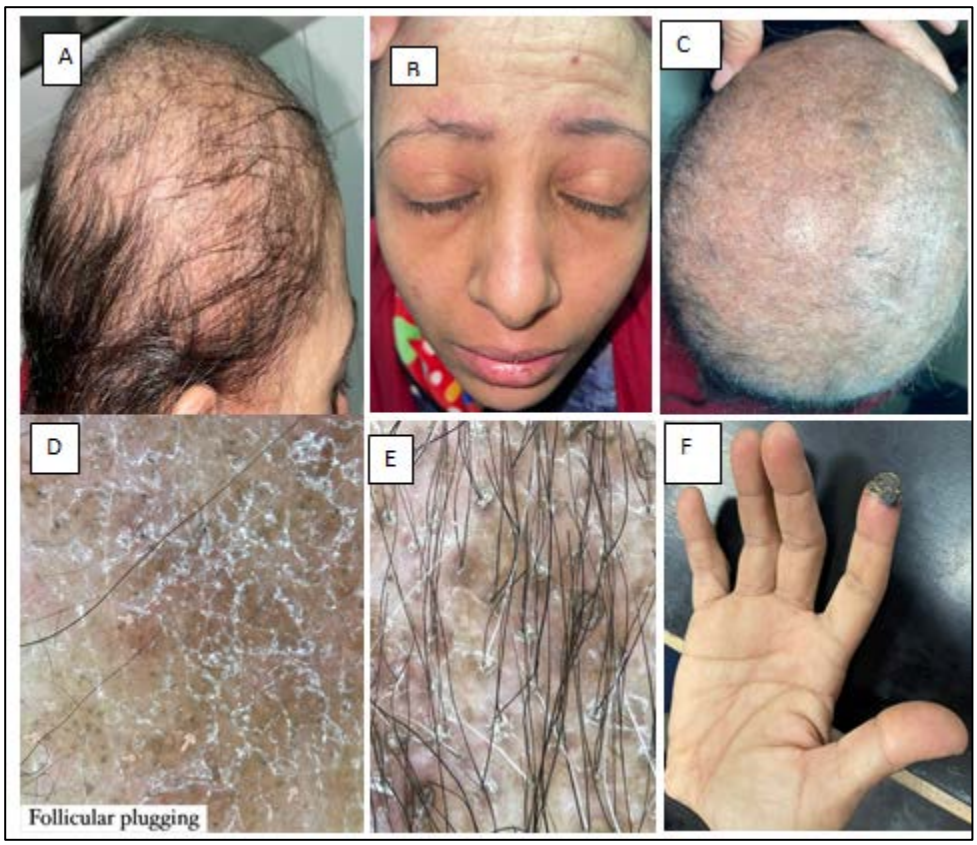

Fig. 1. Photographs of 38-years-old female patient presented to us by :
A,B,C. Clinical views;
A, Diffuse hair loss. B, scaly Erythematous patches involving the Eyebrows. C, Diffuse Non-Cicatricial Alopecia

Dermoscopic views of Scalp lesions before treatment
D, Follicular Plugging , E, perifollicular brownish scales, Erythematous background
F, Clinical views of digital gangrene during treatment

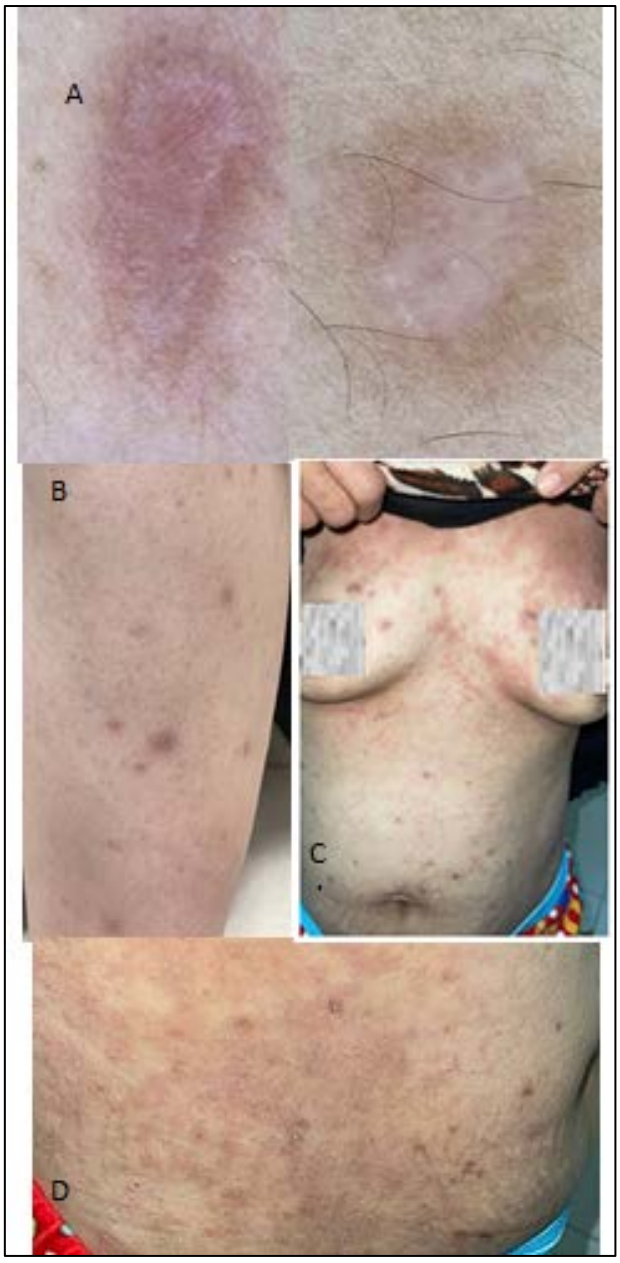

Fig. 2 Phographs showing the Patient also had atrophic papules all over the trunk and extremities (A, B,C,D).

Dermoscopic view by Dermlite DL4 showing atrophic papular lesions , white stellate structureless areas surrounded by a rim of short vessels suspected Degos disease.

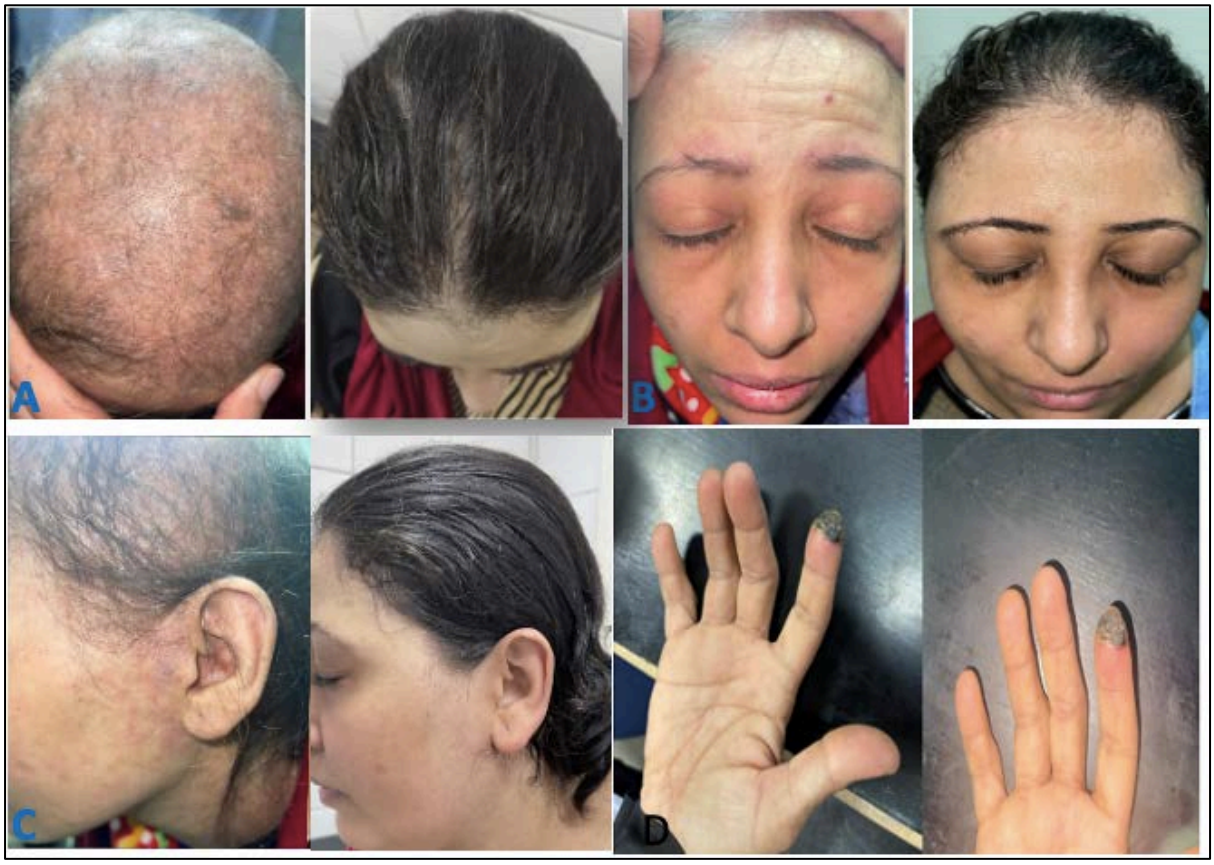

Fig. 3. Photographs shown the Clinical views of the patient after treatment. A, restoring of hair. B,C, disappearance of scaly erythematous patches , D, the gangrenous digit is controled.

تقرير عن حالة مهملة من الذئبة الحمامية الجهازية تظهر بسبب مرض ديغوس الجلدي وثعلبة منتشرة غير تندبية مع استجابة مثيرة للعلاج

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المستخلص

في هذه الدراسة تم الإبلاغ عن حالة مريضة (٣٨ عامًا) تعاني من مرض ديجوس، وثعلبة غير تندبية مع قرحة رقمية تحدث في حالة الذئبة الحمامية الجهازية. وقد لوحظ نمو الشعر بشكل كبير مع بروتوكول العلاج الحالي. كما تمت مناقشة الفعالية المحتملة لهذا العلاج.