

Generalized Discoid Lupus Erythematosus (DLE) with Systemic Lupus Erythematosus (SLE) in Pregnancy: Case Report

AUTHORS

1) Dr. Gardenia Akhyar

*Head of Division, Dermatovenereologist, Dept of Dermatology Venereology
Medical Faculty of Andalas University, Dr. M. Djamil Hospital, Padang, West Sumatera, Indonesia*

2) Dr. IrdawatyIzrul

*Dermatovenereologist, Dept of Dermatology Venereology
Medical Faculty of Andalas University, Dr. M. Djamil Hospital, Padang, West Sumatera, Indonesia*

3) Dr. SherlyBirawati

*Junior Resident, Dept of Dermatology Venereology
Medical Faculty of Andalas University, Dr. M. Djamil Hospital, Padang, West
Sumatera, Indonesia*

Corresponding Author

Dr. Gardenia Akhyar

*Head of Division, Dermatovenereologist, Dept of Dermatology Venereology
Phone no. +62-822-830244-03, E-mail Id: dr.gardenia94@gmail.com*

ABSTRACT

GENERALIZED DLE: *Patients with generalized discoid lupus erythematosus are at a higher risk of progressing to systemic disease than those with a localized variant. Pregnancy is one of the factors that can trigger lupus erythematosus. There are only one case report published in online search that presented generalized discoid lupus erythematosus in pregnancy. We report a case of 24 year old female patient with 31-32 weeks of gestational age complained black-reddish patches accompanied by fine scales, slightly itchy and a little painful on the forehead, cheeks, bridge of the nose, chin, ears, neck, chest, stomach, back, upper and lower extremities since ± 2 months before. ANA profile with positive result. The results of histopathological and dermoscopy examination suitable with discoid lupus erythematosus. Results of the internal medicine department revealed a diagnosis of systemic lupus erythematosus.*

CONCLUSION: *Management of DLE aims to improve general condition of patient, control the lesions, prevent of the development of further lesions; fetal growth abnormalities; preterm birth; neonatal lupus and fetal death.*

KEYWORDS: *Generalized Discoid Lupus Erythematosus, Pregnancy, Systemic Lupus Erythematosus, Keratotic Plug*

INTRODUCTION

Lupus erythematosus is a chronic autoimmune inflammatory disease that presents with various clinical symptoms. The spectrum of lupus erythematosus varies widely, from confined to the skin (discoid lupus erythematosus or DLE) to involving life-threatening systemic manifestations (systemic lupus erythematosus or SLE).⁽¹⁾ Discoid lupus erythematosus can appear at any age, but more frequently at the age of 20-40 years. It can occur in all ethnicities and occurs more frequently in women than men. The etiology of DLE is still not understood. Some environmental factors associated with exacerbation of DLE are trauma 11%, stress 12%, sun exposure 5%, viral infection 3%, cold exposure 2% and pregnancy 1%. Another risk factor for discoid lupus erythematosus is smoking.⁽²⁾ There are three major forms of cutaneous lupus erythematosus as

follows: chronic cutaneous (discoid) lupus erythematosus (CCDLE), subacute cutaneous lupus erythematosus (SCLE), and acute cutaneous lupus erythematosus (ACLE). Chronic cutaneous DLE affects twice as many women as it does men and usually appears in early-to-mid-adulthood. The lesions tend to heal centrally with atrophy, scars, dyspigmentation, and telangiectases. Patients with CCDLE generally less than 10 percent of them will develop SLE. Discoid lupus erythematosus (DLE) consists of localized and generalized forms. Localized DLE lesions usually only occur in the head or neck area, whereas generalized DLE occurs above and below the neck.⁽³⁾ There are 3 case report published in online search that presented generalized DLE on 1985, 2003 and 2014.^(4,5,6) There are 2 case report published in online search that presented DLE in pregnancy namely on 1984 and 2017.^(7,8) There are only one case report published in online search that presented generalized DLE in pregnancy on 2008.⁽⁹⁾ We herein report the first case of generalized discoid lupus erythematosus in pregnancy in Dermatology and Venereology Outpatient Clinic RSUP Dr. M. Djamil Padang, West Sumatera, Indonesia.

CASE REPORT

A 24-year-old female patient with a gestational age of 31-32 weeks with a round, pin-sized reddish patches appeared on the right cheek which gradually widened to the bridge of the nose, left cheek, forehead and chin about 7 months before. The reddish patches gradually become thickened, scaly, and painful when exposed to sunlight. Then reddish patches appeared on both ears which gradually expanded to the neck, arms, hands, back, chest and abdomen. Reddish patches spread to both legs and feet, which over time became thickened and scaly and whitish patches appeared on the lips about 2 months before (figure 1). Patient works as a farmer. *Cutaneous LE Disease Area and Severity Index* score is 49 interpreting severe disease. *Dermatology Life Quality Index* score is 12 interpreting very large effects on patient life. RNP/Sm, Sm, SSA, Ro-52 test was positive. Dermoscopy examination (figure 2) and histopathological examination (figure 3) suitable for discoid lupus erythematosus. Treatment was methylprednisolone tablet 32 mg, osteocal tablet 3x1000 mg, omeprazole tablet, folic acid tablet, mometasonefuroat 0.1% cream on the face, hydrocortisone 2.5% cream all over the body, urea 10% cream and sunscreen lotion SPF 45.

DISCUSSION

There are 3 case report published in online search that presented generalized DLE.^(4,5,6) George *et al*, on 1985 reported a 20-year-old patient had SLE and generalized DLE that failed respond to conventional treatment with topical steroids and high doses of hydroxychloroquinesulphate. The skin lesions responded dramatically to 100 mg of azathioprine sodium daily, flared when the drug treatment was discontinued, and again responded on reinstatement of the same dosage of azathioprine. The case report suggests that generalized discoid skin lesions can be successfully treated with oral azathioprine.⁽⁴⁾ Abdullah *et al*, on 2003 describe 2 patients with SLE whose generalized DLE was unresponsive to systemic steroids and antimalarial agents. They showed dramatic improvement to thalidomide at a dose of 300 mg/d, with maximum benefit achieved within 15 weeks of therapy. Dosages of 50 to 100 mg/d were effective in maintaining remission for 1 year.⁽⁵⁾ Enver *et al*, on 2014 reported a 63-year-old man presented generalized DLE and given imiquimod cream 5% was applied three times a week, every other week. After 24 applications over a period of two months, an almost complete recovery was achieved.⁽⁶⁾

There are 2 case report published in online search that presented DLE in pregnancy namely on 1984 and 2017.^(7,8) Gawkrödger and Beveridge, on 1984 reported a young woman with inactive DLE gave birth in three successive pregnancies to four male infants who showed cutaneous, and in one case cardiac, signs of neonatal LE. Maternal discoid LE may give rise to neonatal LE, and successive siblings can be affected. The explanation of this female preponderance in neonatal LE is unknown, but suggesting that other factors, either genetic or environmental, are required for the manifestation of the condition.⁽⁷⁾ Hideto *et al*, on 2017 reported a 35-year-old woman with DLE was admitted at 11 weeks gestation with a persistent fever. Laboratory studies revealed pancytopenia, elevated liver enzymes, and hyperferritinemia. Bone marrow aspiration confirmed the diagnosis of hemophagocytic syndrome (HPS). She had no findings of infection or active SLE. The administration of high-dose corticosteroids resolved the clinical and

laboratory findings. She delivered a healthy baby at 35 weeks' gestation. The explanation of DLE and pregnancy is unknown.⁽⁸⁾

There are only one case report published in online search that presented generalized DLE in pregnancy on 2008.10 Dadlaniet *al*, on 2008 reported a 24-year-old pregnant African-American woman had a 3-4 year history of chronic, scarring, hyperpigmented plaques on her scalp, face, trunk, and extremities. She complained of joint pain and fatigue. Clinical presentation, laboratory data, and histopathologic features were consistent with SLE in a patient with generalized DLE.⁽⁹⁾

CLINICAL PRESENTATION:



Figure 1. erythematous plaques (butterfly rash) accompanied by fine scales in the malar region, erythematous plaques with an atypical round shape on most of the body, hypopigmented centered erythematous plaques, erythematous macules, and atrophic scarring.

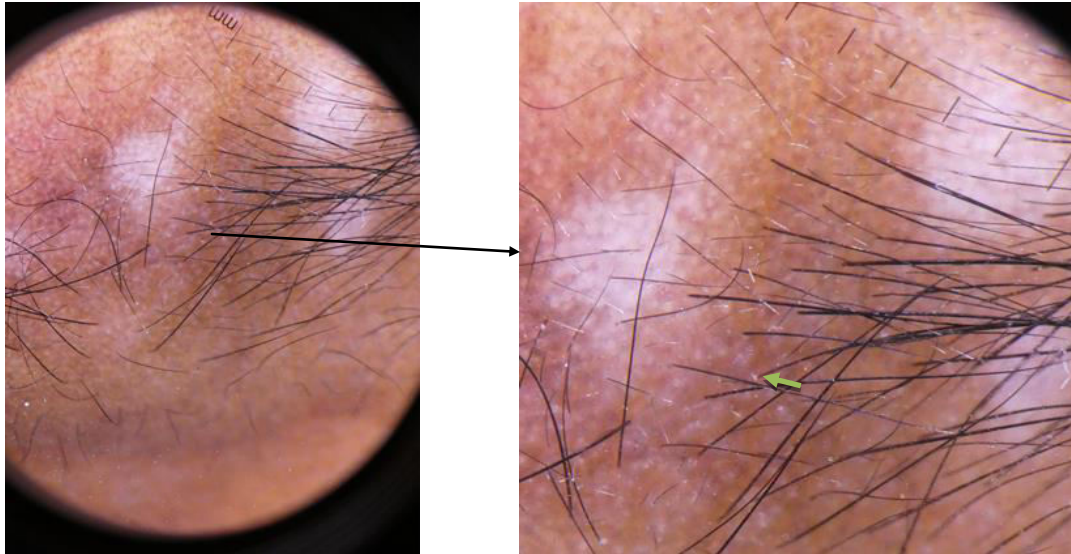
DERMOSCOPY FINDINGS OF PATIENTS:

Figure 2.Whitish scale and follicular plugs (green arrow).

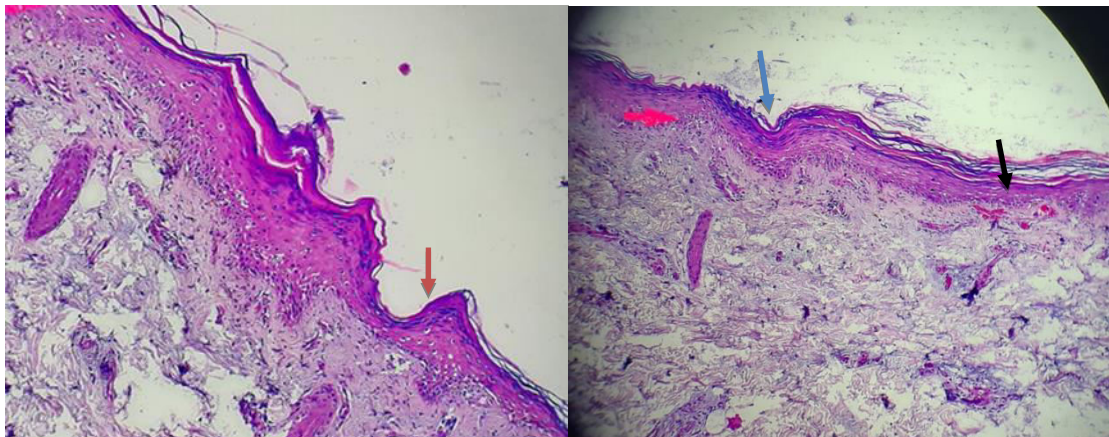
HISTOPATHOLOGICAL FINDINGS OF PATIENTS:

Figure 3.Atrophic epidermis and dermatitis interface (black arrow), hyperkeratotic epidermis (red arrow), parakeratosis, keratotic plug (blue arrow), areas with hypergranulosis, basal degeneration of epidermal cells, flat basal epidermis, lots of melanin incontinence (pink arrow).

Clinical presentation, laboratory data, and histopathologic features in this case were consistent with generalized DLE. In this case, patient was diagnosed with SLE. This is according to the literature if the rash occurs in the malar area of the face, the lesion can resemble a malar rash in SLE so that it can be diagnosed with SLE. To diagnose SLE must contain at least 4 of the 11 ARA criteria.⁽³⁾ In this patient met these criteria namely malar rash, discoid rash, photosensitivity and the ANA result in SLE was positive.

CHOICE OF DRUG- During pregnancy, use of corticosteroids with adequate doses should be given immediately until 6 months postpartum to suppress disease activity. It is estimated that only 10% of dose received by mother will cross the placenta and reach the fetus. Steroid administration will also stimulate fetal lung maturation in preterm fetuses.⁽²⁾

CONCLUSION

Long-term use of corticosteroids in mothers during pregnancy is generally relatively safe for generalized DLE with SLE.

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