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Case Report

Discoid lupus erythematosus in an 8-year-old girl: A rare case

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Abstract

Background: The occurrence of discoid lupus erythematosus in children is uncommon. The global prevalence of childhood-onset SLE (cSLE) varies between 3.3 and 8.8 cases per 100,000 children. The objective of this article was to present a case of DLE in a child, aiming to establish a diagnosis, provide suitable management, and consider the potential risk of developing SLE.
Case Illustration: An 8-year-old girl visited the polyclinic at Dr. Moewardi General Hospital complaining of reddish spots, scales with a black core, and a burning sensation on her cheeks and nose for three months. Initially, small pimples and reddish spots appeared on her face, which grew in size. A dermatological examination of the face showed partly hyperpigmented erythematous plaques, multiple well-defined scales, and partly merged plaques were observed.
Discussion: In pediatric cases of DLE, the primary treatment approach involves minimizing exposure to UV radiation using sunscreen. Low-potency topical corticosteroids were administered on active lesions on the facial region. Systemic therapy may be considered, which may involve using immunomodulatory medications such as systemic corticosteroids and antimalarials. However, in this patient's case, antimalarials were not given as clinical improvement was observed with topical corticosteroids.
Conclusion: This case's DLE diagnosis is based on the patient's history, physical, and supporting examination. Education on risk factors and drug selection according to complaints is the key to successful therapy for DLE patients.

Keywords: *antinuclear antibody, corticosteroid, pediatric discoid lupus erythematosus*

Background

Discoid lupus erythematosus (DLE) is the most common form of chronic cutaneous lupus erythematosus (CCLE), which is part of the spectrum of cutaneous lupus erythematosus (CLE).[1,2] The cause of DLE is yet unknown.[2] Numerous variables, including genetics and environmental ones such as trauma, stress, sun exposure, viral infections, exposure to cold, pregnancy, and smoking, are suspected of inducing DLE.[3,4]

Discoid lupus erythematosus in pediatric patients is rare. The prevalence of childhood-onset SLE (cSLE) in the world ranges from 3.3–8.8 cases per 100,000 children, with involvement of skin lesions as much as 60–85%.[5] Risk factors for the progression of DLE to SLE are difficult to ascertain because the symptoms of SLE are more severe in children than in adults,with an aggressive clinical course characterized by multiorgan involvement, particularly of the kidney, skin, cardiovascular, musculoskeletal, and central nervous systems.[6,7] Diagnosis and treatment delays might increase morbidity and mortality.[6] Diagnosis is required to provide proper therapy to pediatric DLE patients; dermoscopy and ANA testing can be performed. Sunscreen and topical corticosteroids are the most frequently used treatments for DLE in children. This paper aims to describe a case of DLE in a child to establish a diagnosis, provide appropriate management, and take into account the risk of developing SLE.

Case Illustration

An 8-year-old girl presented with the main

complaint of reddish spots accompanied by scales with a black core and a burning sensation in the cheeks and nose, which she had been experiencing for about three months. Before the first spots emerged, the patient stated that she had not taken any medicine or herbal medicine, and she had already been to a dermatologist for treatment of her skin concerns and had been given an ointment, but that she had forgotten the name of the medicine. Small pimples and reddish spots appeared on the face at first, then they grew. Reddish areas thickened and became scaly, and the thicker skin became redder, which then blackened and became uncomfortable, especially if the patientis exposed to sunlight. The patient had no joint discomfort but was weak, coughing, and had a diminished appetite. The patient had never had an illness like this before, and a history of food and drug allergies was ruled out. The patient's family signed an informed consent form for the publication of the patient's data.

On physical examination, the patient appeared compos mentis with normal vital signs. Routine blood tests and clinical chemistry were unremarkable. However, the antinuclear antibody (ANA) profile revealed a positive value for the ribonucleoprotein/antigen Smith's (RNP/Sm), indicating the presence of an autoimmune process, one of which caused SLE. Borderline values for the Sm antigen were also found. PM-Scl100 (polymyositis/scleromyositis antibody 100) is a polymyositis/scleromyositis antibody associated with connective tissue disease. The positive titer value is not explained in detail because the examination was conducted in a private lab, and the data provided is limited.

A dermatological examination of the facial region revealed partially hyperpigmented erythematous plaques, multiple sharply defined scales, and partially confluent plaques (Figure 1). On dermoscopic examination, the initial lesion was erythematous with white scales, follicular plug, and whitish perifollicular halo on the sides of the lesion. Other findings suggest a lesion with more prominent scaly and pigmented structures in the shape of a honeycomb consistent with features of chronic DLE lesions (Figure 2). On histopathological examination, skin tissue have atrophic epidermis, with follicular plug in the dermis lymphocyte and histocyte infiltration especially on perivascular and periadnexal without any signs of malignancy. The results of these examinations support the diagnosis of DLE (Figure 3).

The differential diagnoses in this patient are DLE, SLE, and tuberous sclerosis complex (TSC) based on the history, physical examination, and investigations. The diagnosis of DLE in this patient was made based on his anamnesis, physical examination, and histopathology. This patient was treated with sunscreen, mometasone furoate 0.1% cream twice daily, and moisturizer containing glycyrrhetic acid (Atopiclair® lotion, A. Menarini, Philippines) twice daily for two weeks. Glycyrrhetic acid has pharmacological activities, such as anti-inflammatory, anti-ulcerative, antiallergic, and immunomodulatory effects.

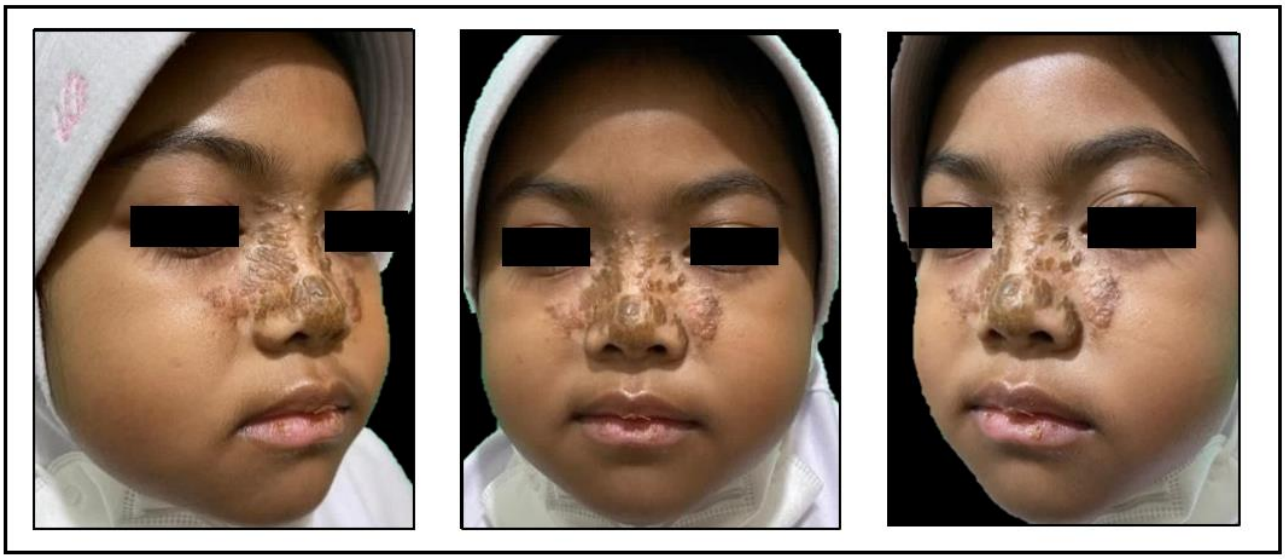

Figure 1. Clinical Appearance of the Lesion on the Facial Region

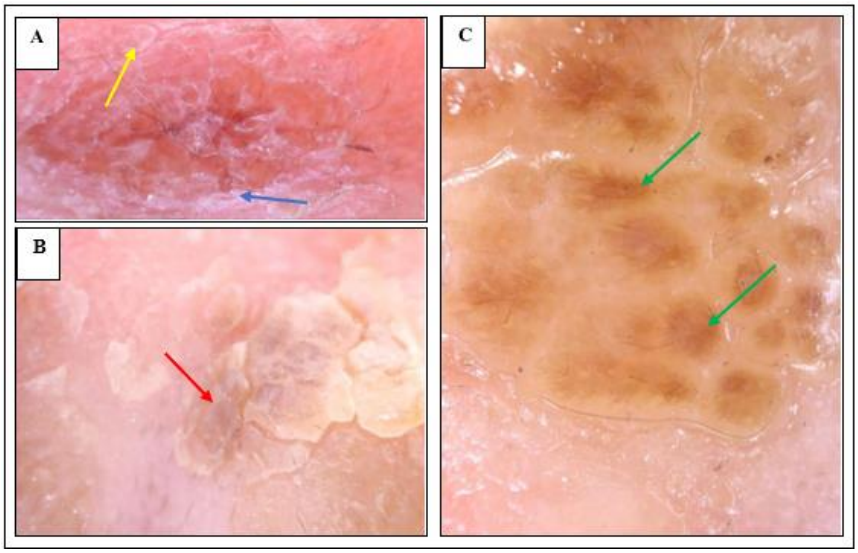

Figure 2. Dermoscopic Findings (**A**). Early discoid plaques are seen as erythematous with white scales (**yellow arrows**) and a characteristic whitish perifollicular halo (**blue arrows**).
(**B**)Thickening of the scales and the start of hyperpigmentation in older lesions (**red arrows**). (**C**) Later stages are characterized by more prominent pigmentation structures, either as honeycomb networks (**green arrows**) or irregular pattern networks.

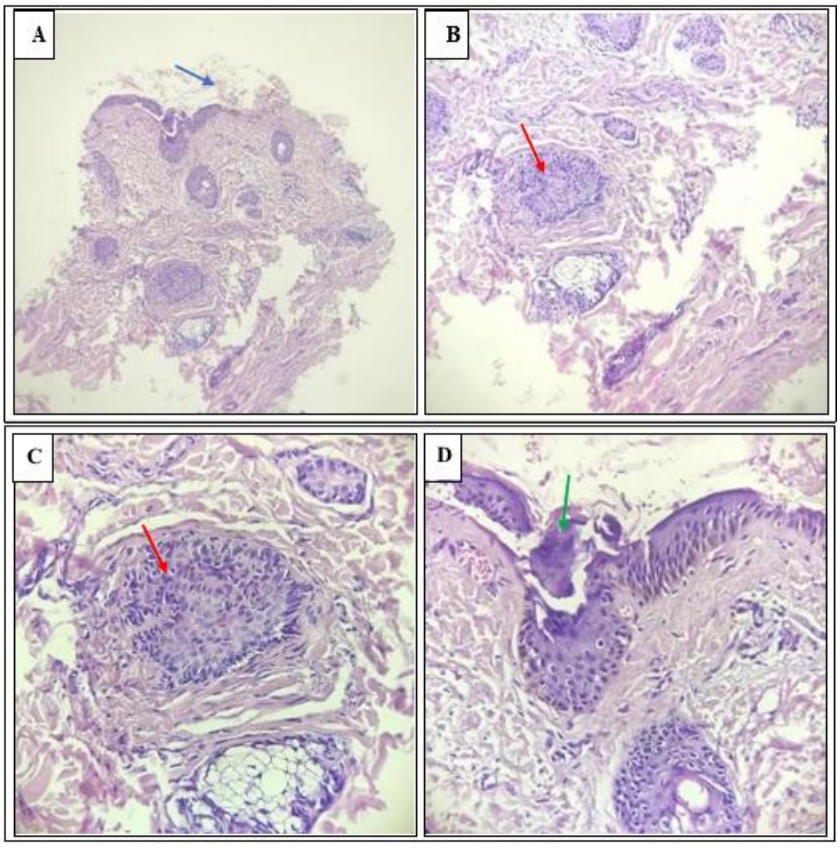

Figure 3. Histopathological Examination with Hematoxylin and Eosin (HE) Staining. (**A**). The epidermal layer is degenerated and shows hyperkeratosis (**blue arrows**) (HE, 10x). (**B**). The dermis layer indicates the presence of inflammatory cell infiltration (**red arrows**) in the subepidermal area (HE, 20x). (**C**). The dermis layer shows the inflammatory cell infiltration (**red arrow**) consist of lymphocytes (HE, 40x). (**D**). In the epidermal layer, a follicular plug (**green arrow**) appears at the top of the epidermis (HE, 100x).

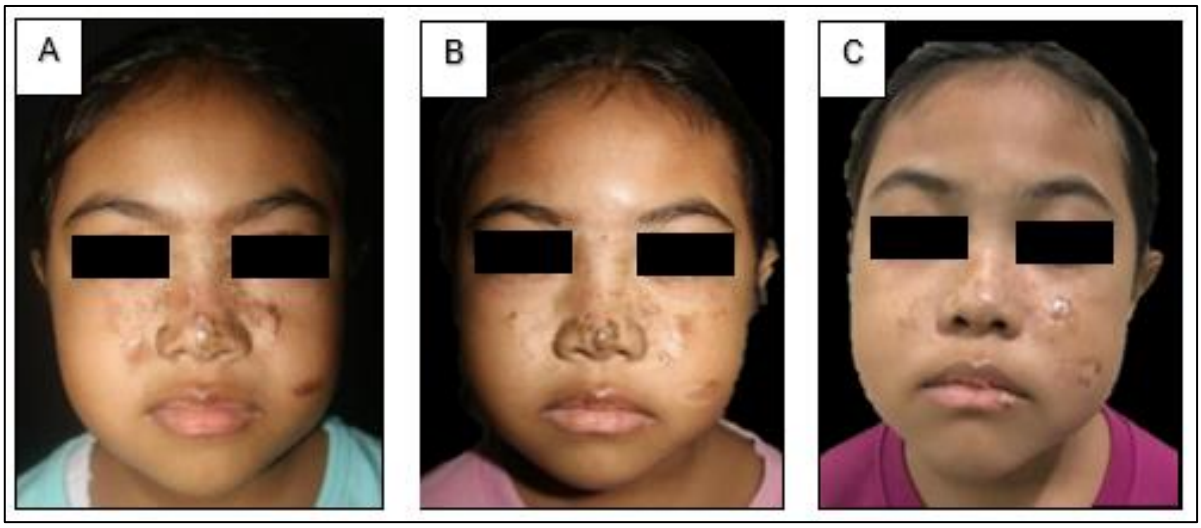

Figure 4. Follow-up and Evaluation of Therapy. (**A**). After two weeks; (**B)** After four weeks;(**C**) After eight weeks

Clinical improvement in the lesions was shown in the form of diminished hyperpigmented lesions in the facial region during the second week of therapy, but new lesions were found in the left buccal region (Figure 4A). We replaced mometasone furoate 0.1 % cream with desonide 0.05 % cream after the second week of therapy and continued with other medications. After four weeks of therapy, old lesions in the facial region appear to increase again, indicating inadequate topical treatment. The lesion in the left buccal region had improved (Figure 4B). The patient was then given the systemic corticosteroid methylprednisolone 12 mg/day and mometasone furoate 0,1% cream applied to the lesion area. The patient's family was educated to examine the patient for re-examination and therapy evaluation within four weeks to adjust the oral corticosteroid dose (tapper off) to 8 mg/day. Evaluation of therapy after eight weeks of treatment with oral corticosteroid dose (tapper off) to 8 mg/day, old lesions in the facial region were significantly reduced, and lesions in the left buccal regions also improved (Figure 4C).

Discussion

Systemic lupus erythematosus can appear at any age, but more often at the age of 20-40years with women more often than men.[3] A study done in United State of America at 2021 reported that the prevalence of SLE was 4.3 cases per 100,000 population with the proportion of women 2-3 times more than men. Discoid lupus erythematosus is responsible for 50-85% of SLE cases.[7] There are no studies that explain the prevalence of DLE, specifically in children. In this case, the patient is an 8-year-old girl, which is rare.

DLE is diagnosed based on the patient's history, clinical examination, laboratory findings, and histology. The earliest clinical sign of DLE is coin-shaped (discoid) erythematous patches of different sizes, followed by adherent follicular hyperkeratosis.[1] DLE lesions are most frequently found in locations exposed to UV light. However, they can also develop in the palmoplantar region and, in rare circumstances, the inguinal folds.[8] In this case, clinical signs of DLE in the facial region were discovered by erythematous plaques accompanied by fine scales with hyperpigmentation and several distinct, partially crusted borders.

The ANA profile examination revealed positive values for the RNP/Sm antigen and borderline values for the Sm and PM-Scl100 antigens. These findings suggest that there is a possibility of systemic involvement in this case. Although the Sm antigen is detected inapproximately 15%–30% of patients with SLE, it is rarely detected in patients with other disorders or healthy individuals. Increases in the Sm antigen titer can indicate the disease's recurrence rate in people with SLE. Patients with a positive ANA test who are also symptomatic should have their ANA profile re-examined every 1 to 3 months. Meanwhile, asymptomatic patients with a positive ANA test should have their ANA profile re-examined every 6 to 12 months.[9] The ANA test result was positive in this case, and dermoscopy revealed erythema with white scales, a follicular plug and a whitish perifollicular halo on the lesion's side. Additionally, the presence of more prominent scaly and pigmented structures in the shape of a honeycomb is consistent with the appearance of chronic onset

DLE lesions.[10]

Histopathological examination of DLE revealed abnormalities in all layers of the epidermis in the form of atrophy and diffuse follicular hyperkeratosis (hyperkeratosis plugging), hydropic degeneration in the stratum basalis, and some disorganization of the basal cells in the form of weak cohesion with irregularly sized cavities, resulting in thickening of the basement membrane epidermis, and adnexal basement membrane.[8,10] Histopathological examination in this patient showed the presence of skin tissue with atrophic epidermis, follicular plug appearance, dermis with lymphocyte and histocyte infiltration, and perivascular and periadnexal skin without any signs of malignancy, in accordance with DLE.

The differential diagnosis for this patient includes DLE, SLE, and TSC. The differential diagnosis of TSC was chosen because the lesion centered on the middle face. Tuberous sclerosis complex is a multisystem neurocutaneous genetic illness with diverse clinical involvement, including the brain, skin, kidneys, heart, eyes, and lungs. The conclusive diagnosis of TSC is made when two main clinical criteria are met, or one major criterion with two minor criteria is met. The major criteria are hypomelanotic macules (≥3 lesions, 5 mm diameters), angiofibromas, ungual fibromas, Shagreen patch, multiple renal hamartomas, cortical dysplasia, subependymomal nodules, subependymal giant cell astrocytoma, cardiac rhabdomyoma, lymphangioleiomyomatosis and angiomyolipomas. The minor criteria are “confetti” skin lesions, dental enamel pits, intraoral fibromas, retinal achromic patch, multiple renal cysts, nonrenal hamartomas.[11] In this patient, the prominent symptom is red patches that gradually thicken and become scaly. The thicker skin turns redder and then blackens, especially if the patient is exposed to sunlight and does not fulfill the TSC criteria, so the differential diagnosis of TSC can be ruled out.

The differential diagnosis of SLE is in accordance with the guidelines of the European League Against Rheumatism and the American College of Rheumatology (EULAR/ACR). To diagnose SLE, after a positive ANA test, the patient must meet at least ten points of the additive EULAR/ACR SLE criteria.[12] The overall score in this patient was two criteria with the Anti-sm test and ACLE (acute cutaneous lupus erythematosus), hence the patient could not be classified into SLE.

In this case, the treatment aims to improve the patient’s overall health, control the lesion, inhibit the development of atrophic scars, and prevent further lesion development. The primary treatment for DLE in children is limiting UV exposure, particularly between 10 a.m. and 4 p.m., and applying sunscreen. Every day, regardless of the weather, sunscreen with a minimum SPF (Sun Protective Factor) of 30 should be applied and reapplied every 2 hours, especially after swimming or severe sweating.[9] Topical corticosteroids of high potency, such as mometasone, are recommended to applied to active lesions.[13] The reason for initiating therapy with a high-potency topical corticosteroid in this patient is that the lesions are still limited to the facial skin. In this case, sunscreen is prescribed, and the patient is instructed to limit sun exposure, and the lesions are treated with mometasone furoate 0,1% cream used twice daily and Atopiclair® lotion applied twice daily for two weeks. After two weeks of therapy, the topical corticosteroid mometasone furoate 0.1% cream was replaced with a lower potency, desonide 0.05% cream.

Systemic therapy with immunomodulatory or immunosuppressive drugs, such as systemic corticosteroids and antimalarials, may be provided if topical therapy is inadequate.[14] Oral corticosteroids are used to treat inflammatory disorders at doses ranging from 0.5–1 mg/kg/day for 2-4 weeks.[15] Antimalarials and oral corticosteroids help regulate and suppress the patient’s inflammation.[15] In this patient, we did not use antimalarials because we assessed sufficient clinical improvement shown by the therapy given. Clinical evaluations should be performed every 4-6 months in pediatric patients with DLE receiving systemic therapy, and laboratory evaluations should be performed every six months.[15] Methylprednisolone was chosen because it inhibits the immune system’s function, making it a safer treatment with fewer adverse effects. The patient’s family was advised to re-examine the patient to evaluate therapy within two weeks to reduce the dose of oral corticosteroids (tapper off) to 8 mg/day if clinical improvement was shown.

The prognosis for this patient is good, but relapses may occur. This is due to children’s low compliance with repeated sunscreen treatments, which results in the formation of new lesions. Additionally, a member of the patient’s family is a smoker. Secondhand smoke exposure has been shown to exacerbate DLE.[9] Patients are recommended to perform follow up in 4 weeks, consistent with a study published in 2020 by Elman et al., who concluded that individuals with DLE had a better prognosis than those with SLE. Lesions may reoccur if the patient fails to avoid risk factors for

DLE.[16] This patient is still on observation thus we cannot conclude the remission of the disease. This case report limited to one case; it may be difficult to generalized the condition of the disease broadly and globally.

Conclusion

We present an 8-year-old child with discoid lupus erythematosus. The diagnosis is based on the patient's medical history, physical examination, dermoscopy, histology, and ANA profile. The crucial aspect of achieving effective treatment for individuals with DLE lies in educating them about the factors that increase the risk and selecting appropriate medications based on their specific symptoms.

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Author Contribution

All authors contribute equally for this project in the study preparation, data collection, case analysis and the writing of the manuscript.

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