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Oleh:

Satya Wydya Yenny

Isramiharti

Gardenia Akhyar

Puridelko Kampar

Yenita

Najirman

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Satya Wydya Yenny, Isramiharti, Gardenia Akhyar, Puridelko Kampar, Yenita*, Najirman**
Dermato-Venereology Department, Anatomical Pathology Department*, Internal Medicine Department** Dr. M. Djamil Hospital/Medical Faculty of Andalas University, Padang West Sumatera, Indonesia

ABSTRACT

Background: Systemic lupus erythematosus (SLE) is a multisystem disease caused by production and deposition of immune-complex resulting in tissue damage. Subacute cutaneous lupus erythematosus (SCLE) is part of cutaneus lupus erythematosus. Morphea (localized scleroderma) is a fibrosing disorder of the skin and subcutaneous tissues. Systemic lupus erythematosus with SCLE type and morphea in the same patient is a rare case and the incidence remains unknown, but there were six cases have been reported. Defect of apoptosis regulation suggested play a role in both of these autoimmune diseases.

Case report: Reported a case of SLE with SCLE type and morphea in a 17-year-old female. There was history of hard palpable red-brownish patches on both of cheeks, right nasolabial fold, under both eyes, both legs, both arms, back, and scalp since 7months ago. Patient easily feels tired, complaining of joint pain, hair easy to fall, fever, decreased of body weight. Physical examination: hair pull test (+). There were hard palpable atrophic scars, hyperpigmentation plaques, atrophy lesions with various colors (white, brown, black,and violaceus), hyperpigmentation and erythematous macules, erythematous plaques with atrophic scars in the centre with erythematous edge, yellowish-brown scales. Mild anemia, leukopenia, and anti-dsDNA (+) found in this patient. Initially patient was diagnosed by morphea, but new lesions still appeared. Histopathological examination was performed and the result was SCLE. Patient matched 4 of the 11 criterias of the American Colleague of Rheumatology (ACR) and diagnosed by SLE.

Discussion: Morphea and SCLE are skin diseases with different clinical and pathological appearance. The relationship between the two diseases has been widely debated for decades, and has conducted research to find the relationship between the two diseases.

Conclusion: Working diagnosis in this patient was SLE with SCLEtype and morphea. These three cases can occur at the same time. This case is still under follow up.

Keywords:*systemic lupus erythematosus, subacute cutaneous lupus erythematosus, morphea*

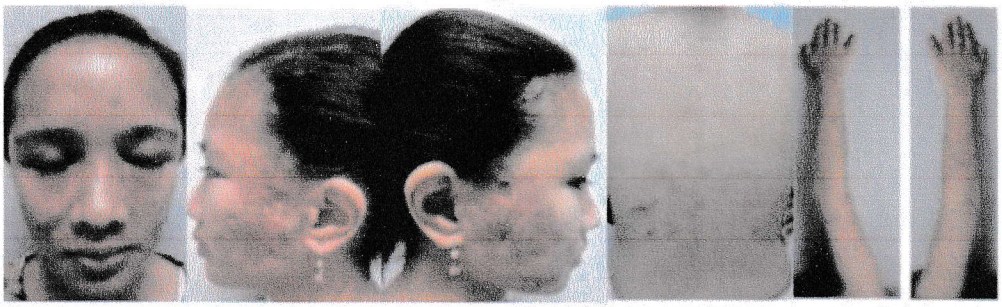
INTRODUCTION

Systemic lupus erythematosus (SLE) is a multisystem disease caused by the production of antibodies and immune complex deposition resulting in tissue damage. Lupus erythematosus (LE) is a term for various diseases associated with autoimmunity in nucleosome molecules and ribonucleoprotein. Cutaneous LE (CLE) is the name for the three main categories of LE-cutaneous: acute cutaneous LE (ACLE), subacute cutaneus LE (SCLE), and chronic cutaneus LE (CCLE). SCLE lesions is formed as erythema and hyperpigmentation with its center formed of scar atrophy and hypopigmentation.[1,2] Morphea known as local scleroderma fibrosis is a disorder of the skin and subcutaneous tissue. Morphea distinguished from systemic sclerosis (scleroderma) by their sclerodactyly, Raynaud's phenomenon, and the change-nail fold capillaries. Etiopathogenesis still unknown, but suspected many factors, such as trauma, toxins, some

medications. Laboratorium commonly found ANA test (+) in 46% of cases.3Systemic lupus erythematosus with SCLE type and morphea in the same patient is a rare case and the incidence remains unknown, this is the seventh cases that have been reported since last 10 years.

CASE REPORT

Reported a rare case morphea with SLE, SCLE type in a same woman, 17 yearsold. Anamnesis of these patients appear brownish red spots on both cheeks, nasolabial fold, under the eyes, both forearms, back, scalp expanded since 7 months ago. The history of exposure to the sun without protection, the patient easily tired since eight months ago, joint pain, while in the physical examination found Body Mass Index 18,36 (underweight) with hair pull test (+). Dermatological status: hard palpable atrophic scars, hyperpigmentation plaques, atrophy lesions with various colors (white, brown, black, and violaceus), hyperpigmentation and erythematous macules, erythematous plaques with atrophic scars in the centre with erythematous edge, yellowish-brown scales. Normal mucous membrane. Laboratorium Hb 10,6 mg/dl, leucocyte 4.700/mm3. Patient had 4 of the 11 criteria of ACR, photosensitivity, arthritis, hematological disorder (mild anemia and leucocytopenia), and positive anti nuclear-antibodies (ANA). We found squamous stratified epithelium, partly atrophy, most hyperplasia (acanthosis) with hyperkeratosis and flat 'rete ridges', vacuol epithelial in epidermis and in dermis we found mild-moderate lymphocytes and PMN infiltration. Histopathologic changes in SCLE especially dermatitis lesions with poor-cell interface. Working diagnosis in this patient was SLE with SCLE and morphea.



DISCUSSION

Morphea has different lesions with cutaneous lesions of lupus in some reported cases. In two cases, patients suffering from SLE and also linear morphea face 4.5 years after the diagnosis of SLE.Whilethe two other cases, patients with morphea who later complained of joint pain, malar rash and laboratory tests lead to SLE The fifth case in patients with morphea 8 years, without any other complaint is found coexistence with SLE. Whereas in the case of sixth obtained coexistence DLE and SLEin the same lesion.6 Laboratory findings in SLE is a high titer ANA and anti-

dsDNA. ANA titer can occur with low/normal in 30-40 percent of patients with DLE; However, less than 5 percent had a high value of ANAwhich are characteristic of SLE patients (>1:320) but this is not spesific for SLE and can be found in other autoimune disease.1,4 In this patient, we found high ANA titer. American College of Rheumatology criteria for SLE is malar rash, discoid lesions, photosensitivity, oral ulcers, arthritis, serositis, kidney disorders, neurological disorders, hematological disorders, immunological disorders, and anti-nuclear antibodies (ANA).1,5 In this patient we found 4 from 11 criterias.

Patients with systemic lupus erythematosus who also morphea in the same patient is seldom reported in the incident are still unknown. Lesions develop within a few months of widening the upper right arm with histopathologic examination support to morphea overlap DLE, laboratory tests support towards SLE. Second histopathological examination was performed and the result was SCLE. Limphohistiocytic cellular infiltrate relatively rare. Rarely seen thickening of the basement membrane area, follicular plugging, or changes in epidermal thickness, and sometimes epidermal atrophy. Infiltrating cells often looked quite solid and typically extends into the deeper dermis (reticular) and/ or subcutaneous, which may help to distinguish it from ACLE or SCLE.1,5 In this patient we found histopathologic epidermal atrophy and mild-moderate lymphocytes and PMN infiltration.

CONCLUSION

Morphea and SCLE are skin diseases with different clinical and pathological appearance. These cases can occur at the same time. The relationship between the two diseases has been widely debated for decades and has conducted research to find the relationship between the two disease.

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