Lupus panniculitis: a rare bilateral and symmetrical manifestation of cutaneous lupus erythematosus in an adolescent girl

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Abstract

Lupus panniculitis is included in the chronic cutaneous lupus erythematosus group. An 18-year-old female patient came with the complaint of lumps on her face. When she was 16 years old, the patient started to complain about lumps on her right lower arm. Lumps were observed on her left cheek and right chin during the ongoing treatment. Histopathology results showed lymphocyte infiltration in between lobular adipocyte with fibrotic and fat necrosis in the subcutis. Lupus panniculitis lesions in this patient were found both on her face and on her lower arms, which are not considered common predilection sites of lupus panniculitis. The skin lesion observed in this patient was also bilateral and symmetrical, which was a rare finding.

Introduction

Lupus panniculitis is a rare variant of chronic cutaneous lupus erythematosus, associated with systemic lupus erythematosus (SLE) in 10-15% of cases, and discoid lupus erythematosus (DLE) in 30-40% of cases.[1,2] It is an inflammatory change that mainly affects the deep dermis and subcutaneous fat and appears as tender subcutaneous nodules and plaques on the face, scalp, lateral side of upper arms, shoulders, breasts, hips, and buttocks, which frequently resolve in depressed lipoatrophic areas.[1,2] At Dr. Sardjito General Hospital, Yogyakarta, Indonesia, between 2016-2020, there were 98 visits for 28 patients diagnosed with lupus panniculitis.

Case Report

An 18-year-old female patient was referred by the autoimmune clinic with the complaint of two lumps on her face: one on her left cheek, and one on the right side of her chin. Two years before admission, the patient noticed a lump on her right lower arm, without tenderness, and without fever. While the lump became larger, other lumps were observed in several areas on the right and left lower arms. After a few weeks, all of the lumps became smaller and sunken. Eight months before admission, the patient was diagnosed with SLE with the involvement of mucocutaneus, hematologic and immunologic systems. The patient did routine medical check-ups and was given methyl prednisolone 4 mg and cyclosporine 2×50 mg as the treatment. Six months before admission, the patient complained that the lumps observed on her left cheek and right chin became larger. The patient recalled no family history for multiple lumps. She also denied any history of previous or current photosensitivity, arthritis, oral ulcers, and traumas. At physical examination, erythematous patches, annular shaped, size of the nodules 2×3×3 cm were found on the left cheek and on the right side of the chin. Skin atrophies with the size of 2×3×1.5 cm was found on the right and left lower arms (Figure 1A-C).

Blood results showed the anti-ds DNA was slightly increased (32.10 U/mL), with normal value of electrolytes (Na 139 mmol/L, K 3.91 mmol/L, chloride 105 mmol/L), liver (SGOT/AST 14 U/L and SGPT/ALT 11 U/L) and renal function test (BUN 10.30 mg/dL and Creatinine 0.62 mg/dL). On the contrary, the abnormal results were erythrocytopenia (3.65×10^6/μL), monocytosis (11.5%), eosinopenia (0.2%) prolonged activated partial thromboplastin time (aPTT = 43.8 seconds) and shorter time of partial thromboplastin time (PTT = 13.3 seconds).

Microscopic examination of skin biopsy from the right side of the chin and the lower left arm showed orthokeratotic epidermis with lamellar and basket weave type, elongation of irregular rete ridges, and basal cell vacuolization. Superficial and deep perivascular lymphocytic infiltrations and lymphocyte infiltrations in between lobular adipocyte (lobular panniculitis) with fibrotic and fat necrosis were found in the dermis and subcutis, respectively (Figure 1D-G). The patient was then diagnosed with lupus panniculitis and treated with oral antimalarial agent. Unfortunately, the patient was lost to follow up.

Discussion

We reported a case of lupus panniculitis in an 18-year-old female patient, who was diagnosed with SLE. There were several notable findings in this case, namely the age of the patient, the location and distribution of the lesion. Lupus panniculitis was predominant in females with age 30-60,[1,2] however in this case, the patient who initially

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experienced lupus panniculitis lesions was 16 years old. The locations of lupus panniculitis in this case were both on the lower arms and on the face. The predilection areas for lupus panniculitis are face, proximal extremities, and trunk.[1,2] Several lesions on the patient's lower arms were not on the areas where lupus panniculitis commonly developed. Lastly, the symmetrical and bilateral fashion of this case is rare; indeed, only two cases were reported.[3,4]

During the initial 2 years of the disease course, the patient also complained about other lumps. From anamnesis, the course of the disease was similar to the course of lupus panniculitis,[1,2] where she had lumps under her skin which eventually became smaller and sunken (Figure 1C). The patient story was concordant with lupus panniculitis lesions from subcutaneous nodules to atrophy. All lumps have this similar characteristic. The patient stated that there was no history of trauma prior to the lump on the right and left lower arms.

The lesions on the lower arm had started 2 years before admission, then, she was diagnosed with systemic lupus panniculitis only 8 months before admission. Thus, this case is similar to other patients with lupus panniculitis where the lesions occur prior to the onset of SLE.[1,2] The pathological results from the biopsy taken from the right side of her chin and her lower arm also showed histological similarities with lupus panniculitis (Figure 1D-G).

Differential diagnoses for this case were morphea and subcutaneous panniculitis-like T-cell lymphoma (SPLTC). Morphea has a similar disease course as lupus panniculitis.[1,2] Based on microscopic features found in this patient, the differential diagnosis of morphea was excluded.

Another differential diagnosis for this case was SPLTC. The histological characteristics of SPTCL can mimic lupus panniculitis' since it also showed lobular infiltration of atypical lymphocytes. However, in SPLTC, rimming adipocytes by lymphocytes expressing CD8 should be observed (CD8 data was not included).[1,2,5]

The course of the lupus panniculitis is correlated to innate immune system. The expression of TLR1, TLR2, TLR3, TLR4, and TLR6 was observed in adipocytes and the expression of TLR5, TLR7, TLR8, TLR9, and TLR10 was found in the stromovascular fraction of adipose tissue. In this autoimmune disease, recognition of self-DNA and self-RNA occurred since TLR7 and TLR9 have the ability to recognize RNA and DNA patterns.[6,7] Subsequently, the adaptive immune system was activated, then, autoantibodies were produced for the nucleic acids and proteins

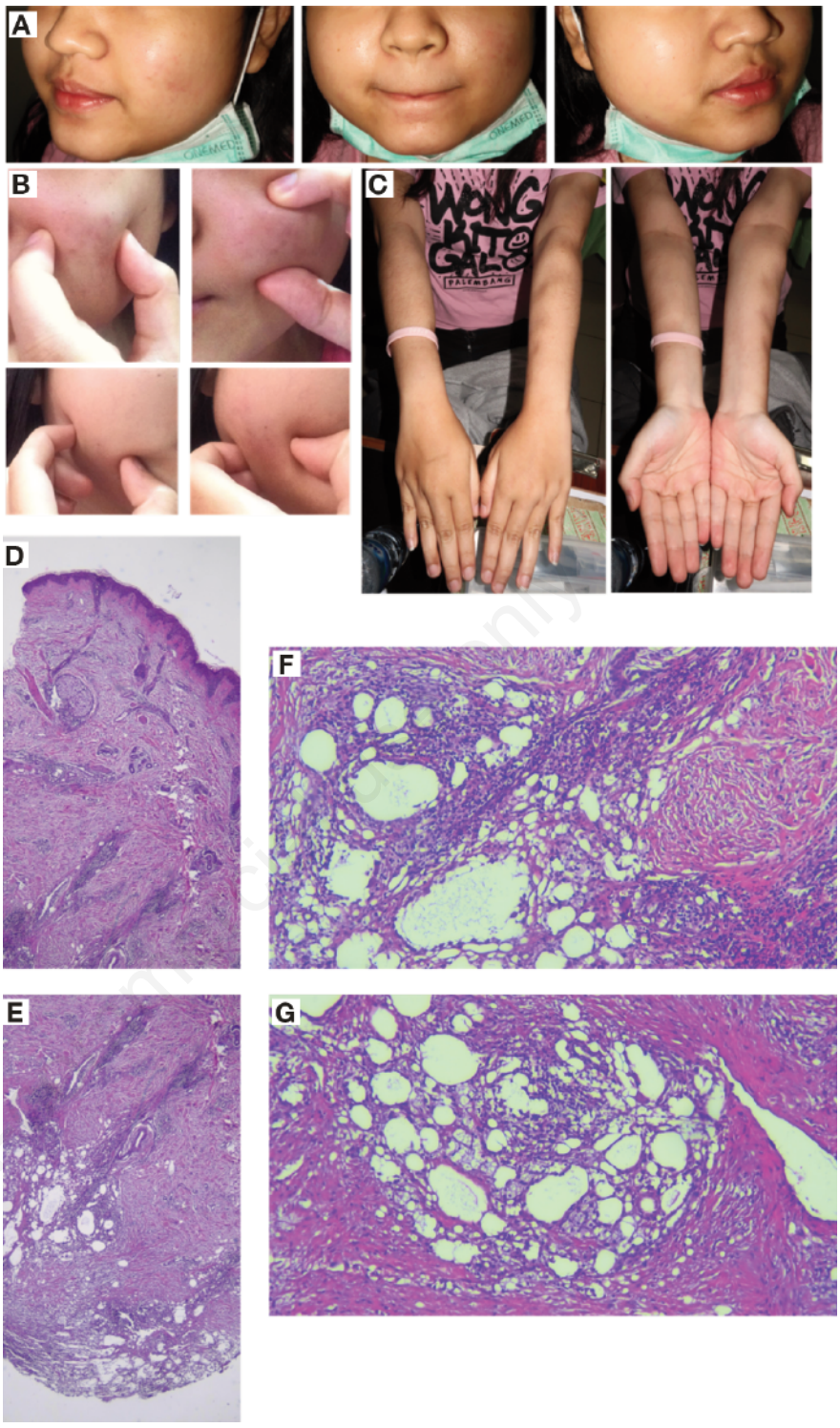

Figure 1. Lupus panniculitis lesions of an 18-year-old female patient who suffered from subcutaneous nodules on her face and atrophy on her right and left lower arms; A) Physical examination of the skin revealed erythematous patch on her left cheek and right chin, with some telangiectasia. Single hard nodule in each side of the face was able to be observed during palpation; B) Skin atrophy was found in several areas in both of the lower arms; C) Pathological results showed in the epidermis: lamellar and basket wave type of orthokeratosis, elongation of irregular rete ridge, basal cell vacuolization; D-G) In the dermis: perivascular lymphocytic infiltrate; in the subcutis: lymphocyte infiltrate in between lobular adipocyte with fibrotic and fat necrosis.

bound to nucleic acid. A previous study showed that hydroxychloroquine was used as treatment for lupus panniculitis because the drug was able to block intracellular TLR *in vitro*.[8,9] It was possible that the treatment given was adequate to decrease the autoimmunity in this patient since the patient still developed new lumps during the last visit. This showed that the current immunomodulator and immunosuppressant drugs were inadequate to suppress the autoantibodies. The addition of other immunomodulator drugs or the increase of the current immunosuppressant dosage was needed to control the condition. Since it was almost certain with the histopathologic results that the patient had lupus panniculitis, the antimalarial agent may be useful to prevent further worsening conditions of lupus panniculitis.

This patient was worried that the lesions on her face would eventually become sunken, and have cosmetic consequences on her appearance. This atrophy, which can be viewed as aesthetic defects, affects patient's psychological health, especially when the lesions were on the face. There are a few available therapies to overcome this major concern for most of the patients.

Soft tissue fillers were reported to be useful in a previous study. Both permanent and nonpermanent fillers had been used as a treatment for permanent atrophy in lupus panniculitis. It was mentioned that the medical therapy for autoimmunity was continued to prevent disease reactivation or reaction to the filler. Poly-L-lactic acid was injected in the subdermal and the treatment was repeated 4 weeks later with subsequent hyaluronic acid after 5 months, for further volume enhancement. There was no disease reactivation observed over the 11 months of follow-up,[10] confirming purified, inert fillers have never been reported to cause reactivation or stimulation of connective tissue disease, given the theoretical probability as a result of antigenic stimulation.[11] Another previous study reported that polyacrylamide hydrogel injectable filler was used to treat atrophy in lupus panniculitis.[12]

Another therapy for facial depression due to lupus panniculitis is autologous fat grafting. The fat grafts are suctioned from the abdomen and centrifuged at 500× g for 2 minutes. The upper oil and bottom liquid are then discarded before the middle fat is injected to the atrophy areas.[13] However, in our clinical setting in Dr. Sardjito Hospital, Yogyakarta, injection of soft tissue filler is more suitable as it is readily available.

Conclusions

A case of lupus panniculitis in an 18-year-old female patient was reported. Lupus panniculitis lesions can develop in an area which is not a predilection site, such as lower arms. Adequate treatment is needed to control the ongoing lupus panniculitis.

References

1. Lake EP, Worobec SM, Aronson IK. Panniculitis. Fitzpatrick's Dermatology in General Medicine. 9. New York: McGrawHill Education; 2019.
2. Burns T, Breathnach S, Cox N, Griffiths C, eds. Rook's Textbook of Dermatology. Vol 1. Chichester: Wiley-Blackwell; 2010.
3. Thiruveedhula H, Kurapati A, Samanthula H, Bora MKR. Bilaterally symmetrical lupus profundus with livedo reticularis as a sole cutaneous manifestation in a case of systemic lupus erythematosus. Int J Res Dermatol 2020;6:795-7.
4. McCarty M, Jeffries M, Hansen R. Lupus erythematosus profundus presenting as morphea-like lesions involving bilateral malar areas of a 12-year-old Philippine male with a negative ANA. J Am Acad Dermatol 2011;64.
5. Massone C, Kodama K, Salmhofer W, et al. Lupus erythematosus panniculitis (lupus profundus): clinical, histopathological, and molecular analysis of nine cases. J Cutan Pathol 2005;32:396-404.
6. Kopp A, Buechler C, Neumeier M, et al. Innate immunity and adipocyte function: ligand-specific activation of multiple toll-like receptors modulates cytokine, adipokine, and chemokine secretion in adipocytes. Obesity 2009;17:648-56.
7. Kono DH, Haraldsson MK, Lawson BR, et al. Endosomal TLR signaling is required for anti-nucleic acid and rheumatoid factor autoantibodies in lupus. Proce Natl Acad Sci USA 2009; 106:12061-6.
8. Lafyatis R, York M, Marshak-Rothstein A. Antimalarial agents: closing the gate on toll-like receptors? Arthritis Rheum 2006;54:3068-70.
9. Barrat FJ, Coffman RL. Development of TLR inhibitors for the treatment of autoimmune diseases. Immunol Rev 2008;223:271-83.
10. Eastham AB, Liang CA, Femia AN, et al. Lupus erythematosus panniculitis–induced facial atrophy: effective treatment with poly-L-lactic acid and hyaluronic acid dermal fillers. J Am Acad Dermatol 2013;69:e260-2.
11. Vera-Lastra O, Medina G, Cruz-Dominguez MdP, et al. Human adjuvant disease induced by foreign substances: a new model of ASIA (Shoenfeld's syndrome). Lupus 2012;21:128-35.
12. Gupta K, Bhari N, Verma KK, Gupta S. Permanent injectable polyacrylamide hydrogel dermal filler for a large subcutaneous defect secondary to lupus panniculitis. Dermatol Surg 2017;43:152-4.
13. Lei H, Ma GE, Liu Z. Evaluation of repairing facial depression deformities secondary to lupus erythematosus panniculitis with autologous fat grafting. J Craniofac Surg 2016;27:1765-9.

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