Jurnal Saintika Medika
Jurnal Ilmu Kesehatan dan Kedokteran Keluarga
Faculty of Medicine UMM

Vol. 15 No. 2 December 2019
p-ISSN : 0216-759X e-ISSN : 2614-X476X
http://ejournal.umm.ac.id/index.php/sainmed

Case Report

Epidermolysis Bullosa Acquisita Occuring In A Patient With Systemic Lupus Erythematosus

Lita Setyowatie[1*], Yustian Devika Rakhmawati[2], Arif Widiatmoko[3], Diah Prabawati Retnani[4]

[1,2,3]Department of Dermatology and Venereology, Faculty of Medicine Universitas Brawijaya / dr. Saiful Anwar Regional General Hospital, Malang, Indonesia
[4]Department of Patology Anatomy, Faculty of Medicine Universitas Brawijaya / dr. Saiful Anwar Regional General Hospital, Malang, Indonesia
***Telephone : +6281-133-936-79**

Email : lita_centik@yahoo.com

Receive : Sept 25th2019. Revised : Oct 4th2019. Published: Dec 18th2019.

DOI : https://doi.org/10.22219/sm.Vol15.SMUMM2.9884

ABSTRACT

Epidermolysis Bullosa Acquisita (EBA) is a rare, chronic autoimmune subepidermal bullous disease and has been noted to be associated with systemic lupus erythematosus (SLE). The incidence of EBA and SLE in one patient within the period of 1980-1990 found only 7 published case reports. A 23 years old woman with exfoliate skin since 12 years ago. Initially itchy on her buttock then appeared small blister. Blister spread almost the entire body and rupture. This complaint got worsening in a year accompanied with hair loss, weight loss, and oral ulcer. Dermatological examination showed patch eritematosa, hyperpigmentation, hypopigmentation, erotion with erythematous base, yellow brownish crust. Also obtained sclerodactyli toes and nail fingers. Laboratory examination anemia gravis, hypoalbuminemia, Coombs test +2, ANA Test negatif, dsDNA IgG 32,80. Histopathology examination showed blister subepidermal, no vacuolar degeneration, no superficial and deep infiltrat, and minimal lymphocyte. Patient had diagnosed SLE from Internal Department based on MEX-SLEDAI score.The patient was treated with metylprednisolone intravenous pulse dose 500 mg on 3 days then tappering off and wound care. Epidermolysis Bullosa Acquisita immunogenetically related with MHC class II haplotype in particular HLA-DR2. This factor suggest playing role in the development of EBA to express more aggresive SLE.

Keywords : epidermolysis bullosa acquisita, systemic lupus erythematosu.

INTRODUCTION

Epidermolysis Bullosa Acquisita is a chronic, subepidermal blistering disease associated with autoimmunity to the collagen (type VII collagen) and that usually begin in adulthood (Woodley and Chen 2012). Theincidence of approximately 0.2 new cases permillion people. On a case report basis, EBA has been noted to be associated with some of the other autoimmune disease like systemic lupus erythematosus (Ludwig, 2013). Systemic lupus erythematous (SLE) is a chronic autoimmune diseaseaffecting heart, lungs, blood vessels, skin, joints, blood and kidneys (Ganapathy *et al*., 2017). The incidence of 2 types of autoimmune diseases of EBA and SLE in one patient is still very rare. From some case report within the period of 1980-1990 only got about 7 cases have been reported worldwide by Dotson *et al*., 1981, Gammon *et al*., 1985 and Boh Erin *et al*., 1990. Therapy for EBA and SLE patients is unpredictable and still needs challanges. Systemic corticosteroid still be the first choice for both EBA and SLE (Badsha and Edward 2003; Ishii *et al*., 2010). Especially supportive therapy is necessary for all EBA patients to help reduce the risk of complications and improve the quality of life. This includes proper wound care and strategies for avoiding trauma (Gupta *et al*., 2012). An EBA case was reported in a 23 year old woman with severe SLE. Patient got therapy methylprednisolone injection pulse dose and followed with methylprednisolone oral with tappering off and wound care.

CASE

A 23 year old woman referral from Ngudi Waluyo Hospital to the emergency room dr.Saiful Anwar general hospital with chief complaint exfoliate skin since 12 years ago. Initially there were itchy on her buttock then appeared small blister. That blister and formed pink area and wet. Then many other blister appeared and spread throught the thigh, chest, stomach, back, arm and leg. Almost the area of the body are involved except face area. Patient also complaint in the past 12 years fingers and toes become stiff and gradually tapers fingertips. About 1 year ago, patient complaint itchy and appear blister in her face. Initially in her forehead and spread to the cheek and chin. The blister and then rupture and leaving a wet and reddish wound. This complaint of wound getting worse in a year.

Patient complaint about hair loss within a year. Ulcer in mouth and tongue waxing and waning affect the patient's appetite. During this time, patient only simply check up at the general practitioner and got some medicine there are dexamethasone tablet, desoximethasone ointment and gentamycine ointment.

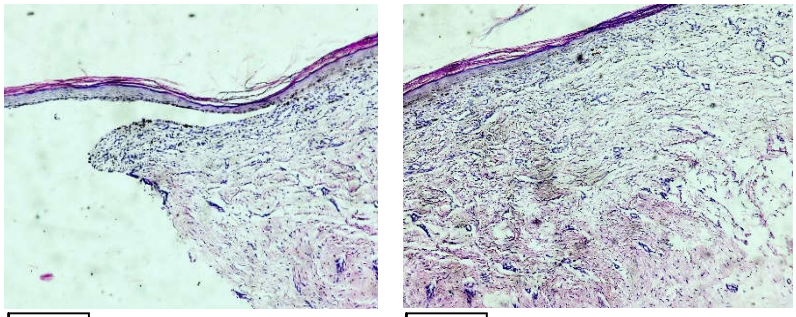
History of the same complaint, history of malar rash, history of skin rash or scald when exposed to sunlight was denied. Patient got menarche at 20 years old and only lasts for a year. No family history of the same complaint.

General examination patient looks severaly ill, composmentis. Vital sign blood pressure 100/60 mmHg, pulse 80x/minute, respiratory rate 20x/minute, and temperature axillae 36,3°C.

Eyes examination of conjunctival anemis. Thorax and abdomen within normal limit. Examination of the extremities of the palms and soles appear pale and there is sclerodactili on the fingers and toes. No enlargement of lymphonodi colli and inguinal, while lymphonodi axillae is hard to evaluate because there are some wound. The patient current weight is 25 kg.

Dermatological examination on facialis obtained hyperpigmented and hypopigmented macules and patches, multiple, irregular shaped, varied in size. Scalp area with alopecia areata and thin whittish scales. Almost the entire body obtained patch erythematousa, patch hyperpigmented, well defined, irreguler, varied in size and erosion with erithematous base (+), brownish crust (+). There is no appear pubic hair in the patient's genital area.

Laboratory examination was found Hb 2,90 g/dL, albumine 1,09 mg/dL, Coombs test +2, ANA Test 0,80 (negative), Anti dsDNA IgM 3,60 (negative), and anti dsDNA IgG 32,80 (positive), rapid test non reactive. Renal function test with ureum 7,40 mg/dL. Liver function test, urinalysis examination and chest X-ray within normal limit. Histopathology examination showed blister subepidermal, no vacuolar degeneration, no superficial and deep infiltrat, and minimal lymphocyte in conjunction to epidermolysis bullosa.

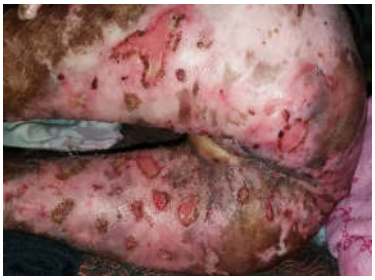


1a 1b

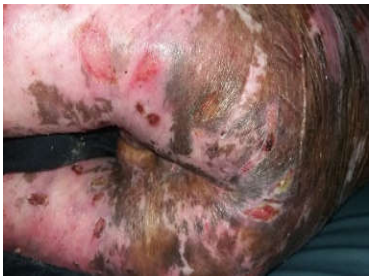
Figure 1a,1b. Histopathology examination showed blister subepidermal, no vacuolar degeneration, no superficial and deep infiltrat, and minimal lymphocyte. (HE, 1000x).

Patients treated together with internal medicine department with a diagnosis severe degree of Systemic Lupus Erythematosus based on MEX-SLEDAI score. From dermatovenereology department based on anamnesis, dermatological and histopathology examination, patient was diagnosed with Epidermolysis Bullosa Acquisita. Patient treated with methylprednisolone injection intravena pulse dose 500 mg for 3 days followed by tappering off methylprednisolone oral 3x16 mg at the 4th day, lansoprazole injection intravenous 1x30 mg, metoclopramid injection 3x10 mg, PRC tranfusion 500cc/day, albumin tranfusion 20% 100 cc, kalk oral 1x500 mg, wound care with wet dressing NS 0,9% 2x15 minute/day, gentamicine cream for the erosion, and olium olivarum for xerosis cutis.

Follow up at the 18th day in the outpatient dermatovenereology obtained clinical improvement with minimal erosion. Treatment followed with methylprednisolone oral 3x16 mg, lansoprazole oral 0-0-1, chloroquine oral 1x250 mg, and gentamicine cream.



2a



2b

Figure 2a. Gluteus and leg posterior D/S obtained patch erythematousa, patch hyperpigmented, well defined, irreguler, varied in size and multiple erosion with erithematous base (+), brownish crust (+). **Figure 2b**. Obtained clinical improvement with minimal lession of erosion.

DISCUSSION

Epidermolysis Bullosa Acquisita (EBA) occurs in bothgenders, at all ages, in all ethnic groups but from several studies many reported onset at an adulthood and genetically associated with the major histocompatibility complex (MHC) class II haplotype, in particular human leucocyte antigen (HLA)-DR2 (Kim and Kim 2013). Theincidence of approximately 0.2 new cases permillion people (Ludwig, 2013). On the other hand, the annual incidence of SLE which is also a kind of autoimune disease is estimated to be 6.4-7.6 cases per 100,000, whereas, approximately one case per 2,000 Caucasians are reported. Systemic Lupus Erythematosus occurs more commonly in females (15-45 years) than males in the ratio of 9-10:1. The etiopathogenesis of SLE is associated with several genetic factors (HLA-DR2 and HLA-DR3; C4a and C4b, C1q, C1r/s and C2 (complement consumption and immune complex deposition) (Ganapathy *et al.*,2017). The same immunogenetic background related with particular HLA-DR2 between EBA and SLE that will contribute to the develpoment of EBA to express a more aggressive SLE and vice versa.

Several criteria have been established for EBA diagnosis : (1) onset in adulthood, (2)absence of a family history of the disease, (3) clinicallesions consisting of bullae produced by trauma, milia,atrophie scars, and nail dystrophy, and (4) exclusion oflichen planus, erythema multiforme, lupus erythematosus (LE), and bullous drug eruption (Dotson et al.,1981). Dotson *et al.*,1981 have been reported a SLE case in patient with EBA in 19 year old woman, where is initially appear lession of blister accompanied by itching starting at the age of 13 years.Boh Ering *et al.*,1990 also report three patients with EBA in whom SLE subsequently developed.

Histopathology examination from lession in the right thigh with punch biopsy methode obtained sub epidermal blister, no vacuolar degeneration, no superficial and deep infiltrat, and only minimal lymphocytes. Subepidermal blister is appropriate with one of the criteria from Dotson *et al.,1981* in support EBA. Another criterion that has been met is the onset of of disease beginning at the age of 12 years, no history of the same disease in family, blister that ruptured leaving trauma, milia, and there is nail distrophic. The lession that present in patient start from the trauma area of the gluteus/sacrum and are not limited to sun exposed areas only.

Patient has been diagnosed severe SLE from internal medicine department based on MEX-SLEDAI score with organ involvement : renal disorder, haemolysis, mucocutan disorder, and fatigue. Laboratory examination obtained Hb 2,90 g/dL with Coomb Test +2, hypoalbumine, and dsDNA IgG positive. Clinical feature EBA and SLE in this patient which may be modified by genetic susceptibility on HLA-DR2. This patient demonstrated both the clinical of classical EBA in association with SLE. It is possible that this represents the coexistence of two distinct entities.

Treatment for EBA still needs challenges until now. Systemic corticosteroid dose 1-1,5 mg/day still the main choice for both EBA and severe SLE (Badsha and Edward 2003; Ishii *et al.*, 2010). Methylprednisolone injection intravenous pulse dose 500 mg for 3 days are said to have significant anti-inflammatory and immunosuppressive effects which will be effective in generating shorter remission periods in both EBA and severe SLE patient (Badsha and Edward 2003; Ishii *et al.*, 2010; Kim and Kim 2011).

Other therapies that can support patient's clinical improvement are extensive erosion treatment in patients. This wound care can prevent other more serious infections (Gupta *et al.*,2012). To date, there are no specific wound care guidelines or any evidence that address the wound care challenges of the EBA population, but in a report by Pope *et al.,* 2012 mentioned some recommendations of wound care for EBA patient there are nutrition management for patient with low intake or got hypoalbumine, management for maintain haemoglobin level above 8 mg/dL, gentle cleansing wound with a saline solution, topical antibiotics short term and rotated every 6 weeks to prevent resistance (Pope *et al.*,2012).

CONCLUSION

There have been reported cases of EBA in 23 year old woman with severe SLE. Enforcement of EBA diagnosis based on onset that occurs from adulthood, no family history of the same disease, clinicallesions consisting of bullae produced by trauma, milia, atrophie scars, and nail dystrophy. Subepidermal blister from histopathology examination is appropriate with one of the criteria. While the diagnose of severe SLE based on MEX-SLEDAI criteria. Patient got therapy methylprednisolone intravenous pulse dose for 3 days with tappering off and wound care. Clinical improvement was obtained on the 18th day of treatment. There are few reports in the literature describing classical EBA in patient with SLE. Finding in this patient add further support to the suggestion that EBA occuring in association with SLE.

REFERENCES

Badsha H, Edward CJ. (2003) Intravenous pulses of metylprednisolone for systemic lupus erythematosus. Semin Arthritis Rheum, 32(6), 370-77.

Boh E, Roberts LJ, Lieu TS, Gammon WR, Sontheimer RD. (1990). Epidermolysis bullosa acquisita preceeding the development of systemic lupus erythematosus. J Am Acad Dermatol, 22: 587-93.

Dotson AD, Raimer SS, Purslye TV, Tschen J. (1981). Systemic lupus erythematosus occuring in a patient with epidermolysis bullosa acquisita. Arch Dermatol, 117, 422-26.

Gammon WR, Woodley DT, Dole KC, Briggaman RA. (1985). Evidence that anti-basement membrane zone antibodies in bullous systemic lupus erythematosus recognize epidermolysis bullosa acquisita autoantigen. The Journal Of Investigate Dermatology, 84:472-76.

Ganapathy S, Vedam V, Rajeev V, Arunachalam R. (2017). Autoimmune disorder-immunopathogenesis and potential therapies. J Young Pharm, 9(1) , 14-22.

Gupta RBS, Woodley DT, Chen Mei. (2012). Epidermolysis bullosa acquisita. Clin Dermatol, 30(1), 60-69.

Ishii N, Hamada T, Dainichi T, Karashima T, Nakama T, Yasumo S et al. (2010). Epidermolysis bullosa acquisita : what's new?. Journal of Dermatology Japanese Dermatological Association, 37 , 220-30.

Kim JH, Kim YH, Kim SC. (2011). Epidermolysis bullosa acquisita : a retrospective clinical analysis of 30 cases. Acta Derm Venereol, 91, 307-12.

Kim JH, Kim SC. (2013). Epidermolysis bullosa acquisita. JEADV, 1-10.

Ludwig RJ. (2013). Clinical presentation, pathogenesis, diagnosis, and treatment of epidermolysis bulosa acquisita. ISRN Dermatology, 1-26.

Pope E, Corrales IL, Mellerio J, Martinez A, Schultz G, Burrell R, et al. (2012). Consensus approach to wound care in epidermolysis bullosa. J Am Acad Dermatol, 67(5) , 904-17.

Woodley DT, Chen M. Epidermolysis Bullosa Acquisita. In Goldsmith LA, Katz SI, GilchrestBA, Paller AS, Leffel DJ, Wolff K, editor. (2012). Fitzpatrick's dermatology in general medicine. 8th edition. New York : McGraw Hill Companies ; .p.634-41.