Peripheral blood abnormality in Sezary Syndrome with bacteremia

Wizar Putri Mellaratna¹*, Zubir², Juwita Sahputri³, Irwandi⁴

INTRODUCTION

Sezary syndrome is defined as the presence of three clinical symptoms: erythroderma, generalized lymphadenopathy, and Sezary cells in the peripheral blood, skin and lymph nodes. However, erythroderma and Sezary syndrome are difficult to be differentiated. The criteria diagnosis of laboratory includes clonal T cells in the peripheral blood with a combined absolute Sezary cell count 1000 cell/mcl or immunopathological abnormalities (increased of CD4 T cell population resulting in an increased of CD4/CD8 ratio > 10.1).¹ The middle adulthood and old age are more susceptible to get Sezary syndrome, although occasionally seen in second decade of life. The certain risk factors of Sezary syndrome are still unknown, but there are reports of HTLV-1 associated with Sezary syndrome.² Acute T-cell leukemia closely related to HTLV-1 infection with clinical characteristics consisting of generalized lymphadenopathy, visceral involvement, hypercalcemia, skin involvement, lytic bone lesion, and lymphocyte involvement. Pleiotropic cell (flower cells) in some cases.³

Clinical sign of Sezary syndrome is erythroderma accompanied by scaling and fissures on the palms and soles, alopecia, onychodystrophy, and associated with exfoliation, edema, lichenification and severe itching. Rarely hyperpigmentation may be seen. The histologic result of Sezary syndrome resembles mycosis fungoides and requires repeated biopsies because of the frequent non-diagnostic results.⁴ There is a proliferation of clonal t cells in skin lesions of Sezary syndrome. The T cell population cannot be detected from a subset of skin biopsies because the T cell population is commonly found in peripheral blood compared to the skin, so that detection of a clonal T-cell population in blood compared to a skin biopsy can confirm the diagnosis of Sezary syndrome.⁵

The presence of Sezary cells in peripheral blood is not specific for diagnosis of Cutaneous T cell lymphoma (CTCL) because Sezary cells can also be found in benign conditions. The involvement of lymph nodes characterized by the presence of a Sezary cell infiltrate.⁶ Differential diagnosis of Sezary syndrome includes erythroderma and non-Sezary syndrome leukemia accompanied by skin involvement. Because the clinical signs of erythroderma are not specific, the history of patients and examination of peripheral blood are required to distinguish with other differential diagnoses.⁷ The risk factor of bacteremia with gram-positive bacteria are erythroderma (especially Sezary syndrome), central venous...

CASE REPORT

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Case: Patients with chief complaints red and scaly patches cover almost part of the body accompanied by itching since 1 week ago. The previous history of taking herbal medicines 3 weeks earlier. There is an erosion and crusting at inferior lips. There is also fissure at left side ventral hand region. There is no generalized lymphadenopathy. The patient has fever, hypertension and hyperglycemia. The laboratory result showed leukocytosis with Sezary cells > 1000 cells/mm³, and bacteremia (coccus). Differential diagnosis of patients was Sezary syndrome, drug-induced erythroderma and mycosis fungoides. The diagnosis was Sezary syndrome because erythroderma and Sezary cells > 1000 cells/ mm³ were found. The patient was treated with injection of corticosteroid, antibiotic, barrier repair moisturizer and potent topical corticosteroid.

Conclusion: Sezary syndrome is often accompanied by bacteremia due to impaired skin barrier function and decreased cellular immunity. Examination of biopsy, bacterial culture and flow cytometer is required for diagnosis.

Keywords: sezary, bacteremia, cerebriform, erythroderma.


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ABSTRACT

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kidney function examinations revealed no abnormalities. The peripheral blood morphology examination results showed Sezary cell > 1000/mcl and bacteremia. The patient's peripheral blood morphology can be seen in figure 2.

The differential diagnosis of patient was Sezary’s syndrome + bacteremia + hypertension + diabetes type 2, generalized exfoliative dermatitis due to drugs + bacteremia + hypertension + diabetes type 2, mycosis fungoides + bacteremia + hypertension + diabetes type 2. Diagnosis of Sezary syndrome is based on the results of laboratory tests.

The patient was consulted to the internist on the first day of admission. Patients are given supportive treatment to prevent hypothermia, maintain fluid balance, and adequate nutrition. The patient was given parenteral methylprednisolone 125 mg/ day, meropenem 1 gr/12 hours, injection of insulin, antihypertensive drugs, potent topical corticosteroid applied 2 times a day, 15-30 minutes after moisturizers, and mild potency steroid applied 2 times daily to face and skin fold area. Moisturizer content ceramide applied 2 times a day all over the body.

The treatment is continued for several
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days until there is a clinical response. On August 28, there was an improvement in the lesions, so methylprednisolone was tapered off to 62.5 mg/24 hours and other drugs were continued. The following are the results of patient evaluation after one-week treatment shown in figure 3.

Two weeks after treatment, the scales on trunks, face, right and left superior and inferior extremities have disappeared. The patient was discharged and prescribed oral methylprednisolone 48 mg/day, moisturizer twice daily and mid potency steroid twice daily. When patients were discharged, the blood pressure was controlled. The antibiotic was stopped.

DISCUSSION

Erythroderma is one of clinical symptoms of Sezary syndrome. The erythroderma characterized by exfoliation, edema and lichenification and severe itching. Hyperkeratosis of palmar and plantar is commonly found. There is a report of six cases of Sezary syndrome without accompanied by erythroderma. Sezary syndrome without skin lesions was confirmed by histopathological examination of normal skin lesions. Patient with Sezary syndrome without erythroderma skin disorder only complain of severe itching without any skin lesions. This case showed Sezary syndrome with erythroderma manifestation and atypical lymphocytes found in peripheral blood.

The patient had history consuming herbal medicines. Food supplement commonly contains ALA substances which can cause adverse drug reaction. Some side effects of drugs due to ALA include skin disorders (rash, angioedema, itching and urticaria) and autoimmune insulin syndrome with symptoms of hypoglycemia that can be life-threatening. ALA intoxication can also cause anaphylactic shock. Erythroderma due to drugs manifests as generalized exfoliative dermatitis and classified as severe drug reaction. Drugs that can trigger erythroderma include antiviral, antituberculosis, sulfamethoxazole, carbamazepine, traditional Chinese medicines, antibiotics, immune checkpoint inhibitors and thalidomide.

The patient complained of hypertension. The incidence of hypertension is high in CTCL because angiotensin can be a trigger of T cell proliferation and stimulates cytokine release. The incidence of hypertension is higher in CTCL patients compared to non CTCL patients (58.5% vs 48%).

There was no generalized lymphadenopathy found in patient. Henn stated that Sezary syndrome is a CTCL variant with clinical features of erythroderma, Sezary cells more than 1000 cells/mm³ in peripheral blood examination with or without lymphadenopathy: fine needle aspiration can be conducted in case lymphadenopathy is found as an additional diagnostic and morphological study.

The patient complained of generalized scale and fissure in palmar of the hand. The clinical features of Sezary syndrome are generalized scales with prominent itching, the disease is expanding rapidly, the violaceous to red erythema of the skin can be found, leonine facies, ectropion, nail dystrophy and palmoplantar keratoderma can also be found.

The etiopathogenesis of Sezary syndrome includes viral, environment, genetic, occupational, and chromosome instability (involvement of gene in cell cycle, epigenetic regulation and JAK/STAT pathway).

The patient had a high blood glucose. There is still no report on the association between diabetes type 2 and Sezary syndrome. Several comorbid factors that are related to diabetes include old age and obesity. The patient suspected to have a comorbid risk factor for suffering diabetes. The peripheral blood examination showed bacteremia. The risk of infection with staphylococci, streptococci and herpetic viral increases in erythrodermic skin lesions. Antibiotic administration can improve the infection and clinical manifestation of the disease.

Sezary cells is not pathognomonic for...
Sezary syndrome because it can be found in other benign skin disease (Sezary cell < 1000 cell/mm³). The diagnostic criteria of Sezary syndrome based on ISCL/WHO/ EORTC are Sezary cell > 1000 cell/mm, the ratio of CD4/CD8 > 10, expression of pan T cell (CD2, CD3, CD4 and CD5), increased lymphocyte count, and chromosomal abnormalities of clonal T cells. CD4+CD27, CD4+CD26 (with or without CD27), CD158 can also be found.

Sin-directed therapy for Sezary syndrome consists of emollients, potent topical corticosteroids and topical chemotherapeutic agents. Emollient can reduce the itching and repair barrier function. Topical potent corticosteroid decrease mitosis activities and destroys malignant cell. Systemic antihistamines can be administered to reduce itching symptoms.

Bacteremia can cause mortality and morbidity. The risk factor for bacteremia include ages, female sex, and chemotherapy. Patients with stage 3 of Sezary syndrome had higher risk of bacteremia. The parenteral antibiotic is required to prevent systemic infection. Patient in this case refused to do a biopsy, so we can not determine the staging of the disease, so the determination of the certain therapeutic agents is also difficult.

CONCLUSION
Sezary syndrome is associated with bacteremia because of the skin barrier disorder and cellular immunity decrease. Biopsy examination, bacterial culture and flow cytometer are needed for the enforcement of diagnosis.

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ETHICAL STATEMENT
Patients had received signed written informed consent regarding publication of their respective medical data in medical journals with confidentiality of patient personal information/identity.

ABBREVIATIONS

REFERENCES