MYCOSIS FUNGOIDES: A CASE REPORT

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ABSTRACT Mycosis Fungoides is a type of Cutaneous T-Cell Lymphoma (CTCL), Non-Hodgins Lymphoma (NHL) manifestations in other organs are known as extranodal NHL and on the skin known as Primary Cutaneous Lymphoma which is the second most common after NHL proliferation in gastrointestinal organs. It has been reported a 42-year-old male, a Balinese. Based on clinical symptoms as well as histologic and immunostaining examinations, these patients are then diagnosed with Mycosis Fungoides. Early diagnosis in patients with Fungoides Mycosis is often difficult to do because it has similar clinical features with other diseases and depends on what stage patient came. Regular observation and repeat biopsy is necessary in patients with a history of chronic and progressive dermatosis.

KEYWORDS Cutaneous T-Cell Lymphoma, Mycosis Fungoides, Non-Hodgins Lymphoma

Introduction

Mycosis Fungoides is a type of Cutaneous T-Cell Lymphoma (CTCL), the term first used by Alibert, a French dermatologist to describe a disease in patients with clinical features of tumour necrosis shaped like a broad fungus on the skin surface of the patient. [1] Mycosis Fungoides is characterized by the presence of a collection of T cell neoplasms in skin tissue. CTCL itself is one of the manifestations of Extranodal Non-Hodgins Lymphoma (NHL). [2,3]

Non-Hodgins Lymphoma manifestations in other organs are known as extranodal NHL and on the skin known as Primary Cutaneous Lymphoma which is the second most common after NHL proliferation in gastrointestinal organs in the United States. In Primary Cutaneous Lymphoma, 75% is a T cell derivative and is known as CTCL in which the most common forms are Mycosis Fungoides, Primary cutaneous Peripheral T-Cell Lymphoma (pcPTL), CD30+ T-cell lymphoproliferative disorders of the skin (CD30+LPD), and Sezary syndrome. [3,4] CTCL estimated as 0.29 cases per 100,000 population in Western Europe and North America, its about 4 cases per 100,000 population/year.[4] In this case, the report will be reported patients diagnosed with Mycosis Fungoides and their therapeutic problems. As these cases are rare in Indonesia and there are often delays in diagnosis, it is expected to add insight to early diagnosis and provide optimal therapy to prolong patient life expectancy.

Figure 1: Clinical features of the patient’s skin.
Case Report

A 42-year-old male, the Balinese, came to the surgery department in our Hospital in January 2013, the patient was complaints of lumps in the left buttocock that felt increased in the last month. The lump was initially small but gradually felt enlarged. The lump increases in size and secretes fluid, and the patient complains of more pain. Also, skinned patients also appear brownish spots that long thickened and become blackish. The spots were initially itchy but gradually itchy disappeared. Patients had to go to a general practitioner and were given a skin ointment, the patient forgot the name of the medicine, but the spots did not disappear but even worsened until one of them became a large bump.

No history like this before. No family member suffers like this illness before. Patients have worked as bus drivers and pickups, no smoking and no consuming alcohol.

From physical examination, vital signs patient were stable with the Visual Analog Scale (VAS) score 2/10. Conjunctiva patient is slightly anaemic. No hepato or splenomegaly. In left gluteal regio patient there was an ulcer, 15 × 10 cm in size, positive necrotic tissue and pus. There was also hyperpigmentation on the skin surface around the ulcer (Figure 1). From the laboratory examination we found Hb 7.7 g / dL (12-16 g / dL), MCV 83.7 fL (80-100 fL), MCH 23.9 pg (26-34 pg), MCHC 28.5 g / dL (31-36 g / dL), WBC 42.07 x103 / µL (4.1-11 x103 / µL), with Neutrophil 41.04 x103 / µL (2.5-7.5 x103 / µL), lymphocytes 0.31 x103 / µL (1-4 x103 / µL), monocytes 0.49 x103 / µL (0.1-1.2 x103 / µL), eosinophil 0.03 x103 / µL (0-0.5 x103 / µL), basophil 0.01 x103 / µL (0-0.1 x103 / µL). Blood chemistry results increase in BUN 90.6 mg / dL (8-23 mg / dL), creatinine serum 2.2 mg / dL (0.5-0.9 mg / dL), but the other were in normal limit.

Patient chest x-ray show normal form both lung and heart (Figure 2). We did not do a PET scan in this patient because we did not have this facility in our hospital. A fourth month ago, patients planned to do Fine Needle Aspiration Biopsy (FNAB), and the result was epithelioid hemangioendothelioma with an abscess (Figure 3).

After approximately four months of treatment from oncologic surgery, the patient had did not improve, and the lump became bigger than the baseline. At that time, they consult to the internal medicine department, and we suggest performing
The term Cutaneous T-Cell Lymphoma (CTCL) was first expressed in 1979 in an International Workshop organized by the National Cancer Institute (NCI) describing the presence of a population of patients with Mikosis Fungoides namely the population of T-cell clone, decreased antitumor response and epidermis as a microenvironment. In Mycosis Fungoides a malignant process occurs from the activity of skin-homing T cells, i.e. T cells that have cell surface receptors known as cutaneous lymphocyte-associated antigen (CLA). [1,7]

Three important components occur in the pathophysiology of patients with Mikosis Fungoides namely the population of T-cell clone, decreased antitumor response and epidermis as a microenvironment. In Mycosis Fungoides a malignant process occurs from the activity of skin-homing T cells, i.e. T cells that have cell surface receptors known as cutaneous lymphocyte-associated antigen (CLA). [1,7]

Figure 5: Patient Timeline.

an open biopsy for better diagnosis, and checking the immunohistochemical result of atypical lymphoid infiltrate that invades into the epidermis (epidermotropism), dermis and cutis without any description of suspected spongiosis and conclude as Mycosis Fungoides (Figure 4). The diagnosis becomes Cutaneous T-Cell Lymphoma (Mycosis Fungoides) stage IV because the disorder may spread throughout the body. Immunohistochemical results were positive diffuse CD3+ and CD4+, with the conclusion of Immunohistochemical examination is to support the diagnosis of Non-Hodgkin’s T cell lymphoma. Patients then planned to undergo chemotherapy with regimen given was Cyclophosphamide 1200 mg in NaCl 0.9% 100 cc spent in 30 minutes, Doxorubicin 70 mg in NaCl 0.9% 100 cc was spent in 30 minutes, Vincristine 2 mg in NaCl 0.9 % 5 cc was given with a slow bolus and Methylprednisolone 3 x 16 mg administered orally (CHOP regimen). After the third series of CHOP regimen, tumour size didn’t reduce, and we conclude there is no response of therapeutic.

We change the regimen become Ifosfamide, Carboplatin and Etoposide (ICE regimen). However, four days after regimen administer, the patient had febrile neutropenia, and pass away in 6 days of treatment due to sepsis condition.

Discussion

The term Cutaneous T-Cell Lymphoma (CTLC) was first expressed in 1979 in an International Workshop organized by the National Cancer Institute (NCI) describing the presence of a collection of lymphoproliferative disorders characterized by the localization of T cell neoplasms in the skin. [1,5,6] A European case-control study report that CTCL is a rare disease and its risk factors have not been studied in any great detail. CTCL was the most common Cutaneous Lymphoma subtype accounting for 71% of cases/years. From that, 38% was Mycosis Fungoides, followed by pcPTL 29%, CD30+LPD 14%, and Sezary syndrome 0.8%, [7,8]

Mycosis Fungoides is very rare. In the United State, there are between 16.000 and 20.000 cases of the illness. Each year, there about 1000 newly diagnosed cases. The incidence of Mycosis Fungoides were 0.41/100.000 person-years in 2001-2005. [8]

The aetiology of Mycosis Fungoides is still not known clearly, but some retrospective studies suggest exposure from the environment, viral infections such as HTLV I / II, HIV, HSV, etc. are thought to trigger the onset of the disease. Another study suggests that the cause may be related to chronic antigen stimulation that is secondary to exposure to chemicals or pesticides. [1,7]

Sepsis is a frequent terminal complication. [15,16] In our case, first FNAB show epithelioid hemangiendothelioma with abscess maybe he came in the mycotic stage. So in our FNAB didn’t conclude mycosis fungoides, but after four months, he becomes in tumour stage. However, there were no atypical cells in the peripheral blood, ruling out the possibility of Sezary Syndrome. Transformation to large highly atypical lymphocytic infiltration in the dermal component is associated with the development of an aggressive biological course. [4]

Although Mycosis Fungoides is a malignant lymphoma of low-grade malignancy with prolonged survival, important prognostic parameters are the stage at diagnosis, the absence of complete remission after first treatment. Average life expectancy is 7 to 10 years after diagnosis, if with proper treatment. However, it depends on staging the patient and the remission status after the therapy. [14] If the absence of complete remission the prognostic parameters do not influence survival and prognosis is bad. Sepsis is a frequent terminal complication. [15,16]

Summary

It has been reported a 42-year-old male, a Balinese. Based on clinical symptoms as well as histologic and immunostaining examinations, these patients are then diagnosed with Mycosis Fungoides stage IV. The management given is chemotherapy with a 3rd series of CHOP regimen and change to ICE regimen because there is no complete remission. Early diagnosis in patients with Mycosis Fungoides is often difficult to do because it has similar clinical features with other diseases and depends on what stage the patient came. Regular observation and a repeat biopsy are necessary for patients with a history of chronic and progressive dermatosis. Early detection and treatment will provide a better prognosis and prolong the patient’s life expectancy.

Competing Interests

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