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Erythrodermic Cutaneous T-Cell Lymphoma: A Great Imitator? Unmasking Its Clinical and Histopathological Clues

Ennesta Asri [1*], Indah Indria Sari [2], Tofrizal [3], Pamelia Mayorita [4], Adianto Jaya Nagara [5]
[1,2,5 *] Department of Dermatology, Venereology and Aesthetic, Medical Faculty, Andalas University / M. Djamil Hospital Padang
[3,4] Department of Pathological Anatomy, Medical Faculty, Andalas University / M. Djamil Hospital Padang
Email: ennestaasri@med.unand.ac.id, indahindria@med.unand.ac.id, tofrizal@gmail.com, mia.pamelia@gmail.com, adianto.jn@gmail.com
*Corresponding Author: ennestaasri@med.unand.ac.id

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KEYWORDS
Erythroderma, Erythrodermic-Cutaneous T-Cell Lymphoma, E-Ctcl, Mycosis Fungoides

Abstract

Background: Both erythrodermic mycosis fungoides (E-MF) and Sézary syndrome are considered forms of erythrodermic cutaneous T-cell lymphoma (CTCL). Erythrodermic CTCL is a term used to describe CTCL subtypes that present with erythroderma as a prominent feature. Diagnosing these conditions can be challenging due to their non-specific clinical and histopathological signs. Several other skin conditions like dermatitis seborhoeic (DS), atopic dermatitis (AD), psoriasis vulgaris, and systemic diseases can present with erythroderma and similar symptoms, leading to a potential misdiagnosis or delayed diagnosis.

Case Report: We reported a case series consisted of four patients of Erythrodermic CTCL in M Djamil Hospital Padang from 2021-2023. Patient no. 1 is 85 years old male with E-MF that initially diagnosed as erythroderma ec. DS. Patient no. 2 is 65 years old male with E-MF that initially diagnosed as erythroderma ec. DS. Patient no. 3 is 70 years old male with Sézary syndrome that misdiagnosed as erythroderma ec AD and psoriasis vulgaris. Patient no. 4 is 33 years old female with Sézary syndrome that initially diagnosed as erythroderma ec. DS. All cases were confirmed with immunohistochemistry using CD3, which showed a positive result of epidermotropism.

Discussion: Establishing the etiology of erythrodermic CTCL poses a challenge. A thorough examination must be conducted to establish the diagnosis, including comprehensive histopathological examination and immunohistochemistry.

1. INTRODUCTION

Erythroderma or exfoliative dermatitis is defined as the emergence of widespread red patches on the skin, covering more than 90% of the body surface area with varying degrees of scaling. This condition is considered a dermatological emergency and can occur in individuals of all age groups. The most commonly affected age group is 45 years or older. Some cases may also be accompanied by erosions, skin thickening, and potential changes in hair and nails.[1-3]

The etiology of erythroderma can be categorized into four categories: pre-existing skin diseases, drug reactions, malignancy-related, and idiopathic. A study from Singapore has concluded that pre-existing dermatological diseases are the most common cause of erythroderma (68.9%), with eczema and psoriasis being the primary causes. Idiopathic erythroderma is the second most common category (14.2%). Drug-related erythroderma is the third most common group (10.7%). Malignancy is a relatively rare cause of erythroderma (4%). A study from Turkey revealed that the most common etiological factor for erythroderma is psoriasis vulgaris (59.6%), followed by drug eruptions (17%), mycosis fungoides (12.8%), atopic dermatitis (4.3%), bullous pemphigoid (2.1%), pityriasis rubra pilaris (2.1%), and erythema multiforme-like eruptions (2.1%).[4-7]

Sézary syndrome is characterized by a triad of diffuse erythroderma, generalized lymphadenopathy, and circulating malignant T cells with cerebriform nuclei,

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known as Sézary cells. Sézary syndrome is a rare form of cutaneous T-cell lymphoma (CTCL), accounting for approximately 3% of all skin lymphomas. Mycosis fungoides (MF) is a form of T-cell lymphoma defined by the epidermotropism of memory/effector T-helper cell subsets. Erythrodermic MF (E-MF) typically presents as a chronic course of MF with generalized erythema or poikiloderma but lacks systemic involvement seen in Sézary syndrome.[8-10]

2. CASE REPORT

2.1 CASE 1

Mr. M, an 85 years old man, married, retired soldier, lived in Rengat, was reffered from dermatologist with suspected erythroderma ec susp seborhoeic dermatitis, to emergency installation of Dr. M. Djamil Padang Hospital on April 9th 2023, with complaint reddish patch with white scale on almost the whole body that felt itchy which gradually developed since 2 weeks ago. This complaint appeared initially 2 months ago as reddish patch without scale that not feel itchy on almost the whole body. Around 1 month ago, white scale begins to appeared and the patient went to dermatologist in Rengat, got hospitalized for 10 days, received metilprednisolone and hydrocortisone, then the symptoms subsided for a moment. Patient request for early discharged from hospital. Within 1 week later the symptoms increased again. The patient went to dermatologist in Rengat again and then reffered to Dr. M. Djamil Padang Hospital. There was no history of new medications within the last 2 months before and no history of blister in genital or oral. History of drug allergy before was denied. There was history of head dandruff when the patient was young. No history of frequent sneezing in the morning and asthma before. No history of reddish patch with micaceous white scale on the elbow and knee before. No history of using topical drugs before. Patient was a heavy smoker which consume 2 pack of cigarettes a day. He stopped smoking since 5 years ago. Patient denied history of weight loss within the last 1 month. There was no history of diabetes mellitus, hypertention, renal disease and liver disease.

On physical examination, the blood pressure was 150/70 mmHg. From head and neck examination, chest, abdomen, upper and lower extremities were within normal limit. There was no enlargement of the cervical, axillar, and inguinal lymph nodes. The body mass index was 22.65 (normoweight). Dermatological state revealed erythematous macules, hyperpigmented macules, whitish scales, and erosion on almost the whole body with universal distribution. Laboratory examination found anemia (11,8 g/dl) and hypoalbuminemia (2,4 g/dl). Other laboratory examination including serum electrolyte, liver and renal function are within normal limit. There was no sezary cell found. From dermoscopy examination showed whitish scale. Skin sample for histopathology and immunohistochemistry examination was taken.

Initially patient was diagnosed as erythroderma ec susp seborrheic dermatitis DDx cutaneous T-cell lymphoma. Patient then treated with methylprednisolone 32mg/day PO, lansoprazole 1x30mg PO, cetirizine 1x10mg PO, lanolin oint 10% 30 minutes before taking a bath on upper part of body in the morning, and on lower part of body in the evening, hydrocortisone 2,5% on upper part of body in the morning, and on lower part of body in the evening. Patient consulted to internal department for the newly diagnosed hypertention stage I and received amlodipine 1x5mg. There was improvement after 1 week of therapy. Patient then discharged from hospital and advised to control to Dermatology and Venereology department outpatient clinic. But the patient didn't come again and loss to follow up.

From histopathological examination found lichenoid reaction with hyperplasia, parakeratoses, and irregular acanthosis on epidermis. There was also focus of lymphocyte infiltrate at basal epidermis (epidermotropism). On dermis found lymphocyte perivascular and perifollicular infiltrate with follicular athropy. These features could be found in cutaneous T-cell lymphoma (CTCL). Immunohistochemistry examination is needed to confirm the diagnosis. From CD3 immunohistochemistry examination found dense clusters of CD3 (+) in the upper dermis, dermal papillae and some infiltrative cells into the epidermis (a feature of epidermotropism). These microscopic findings suitable for CTCL.

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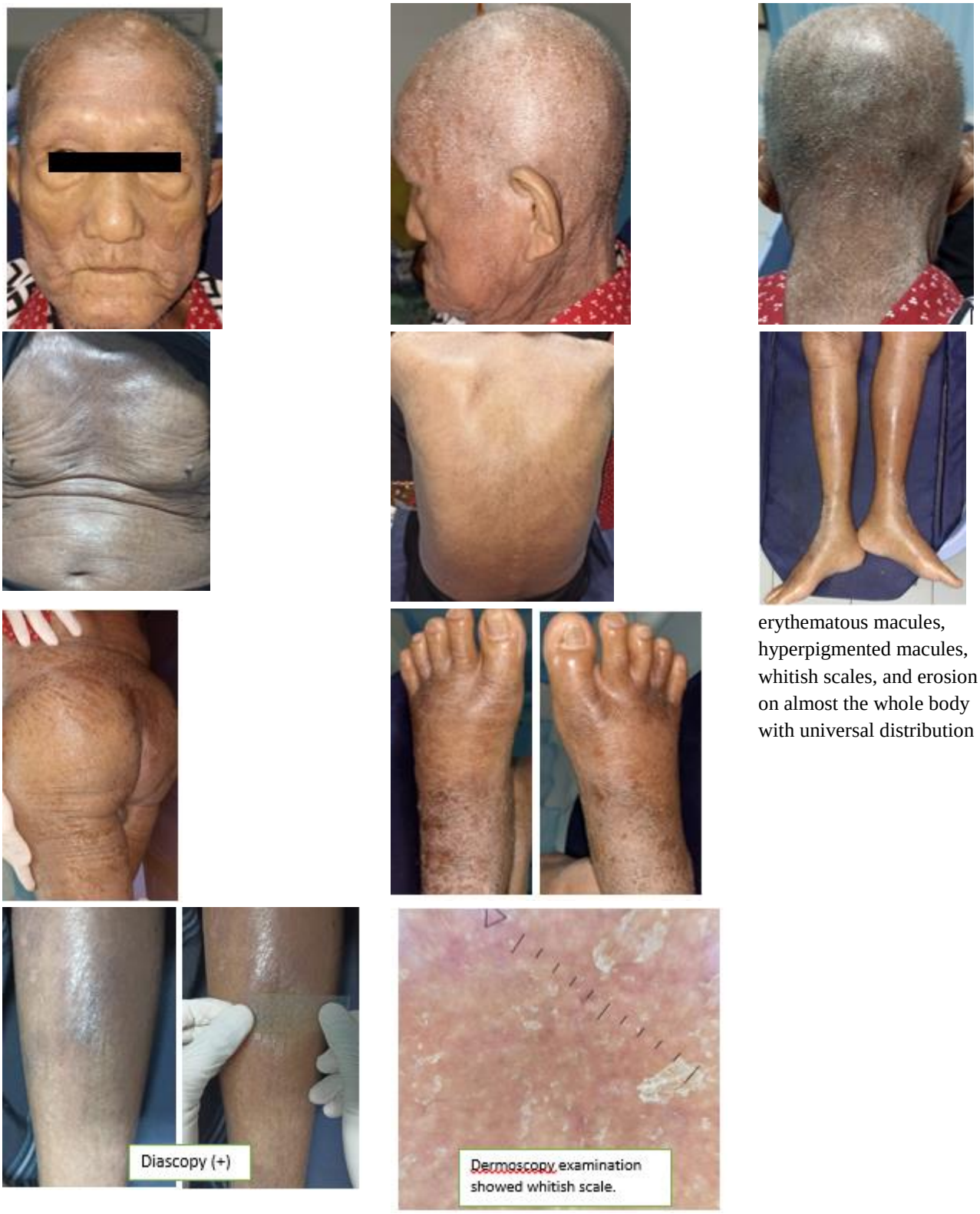

Figure 1A. Clinical presentation of the patient.

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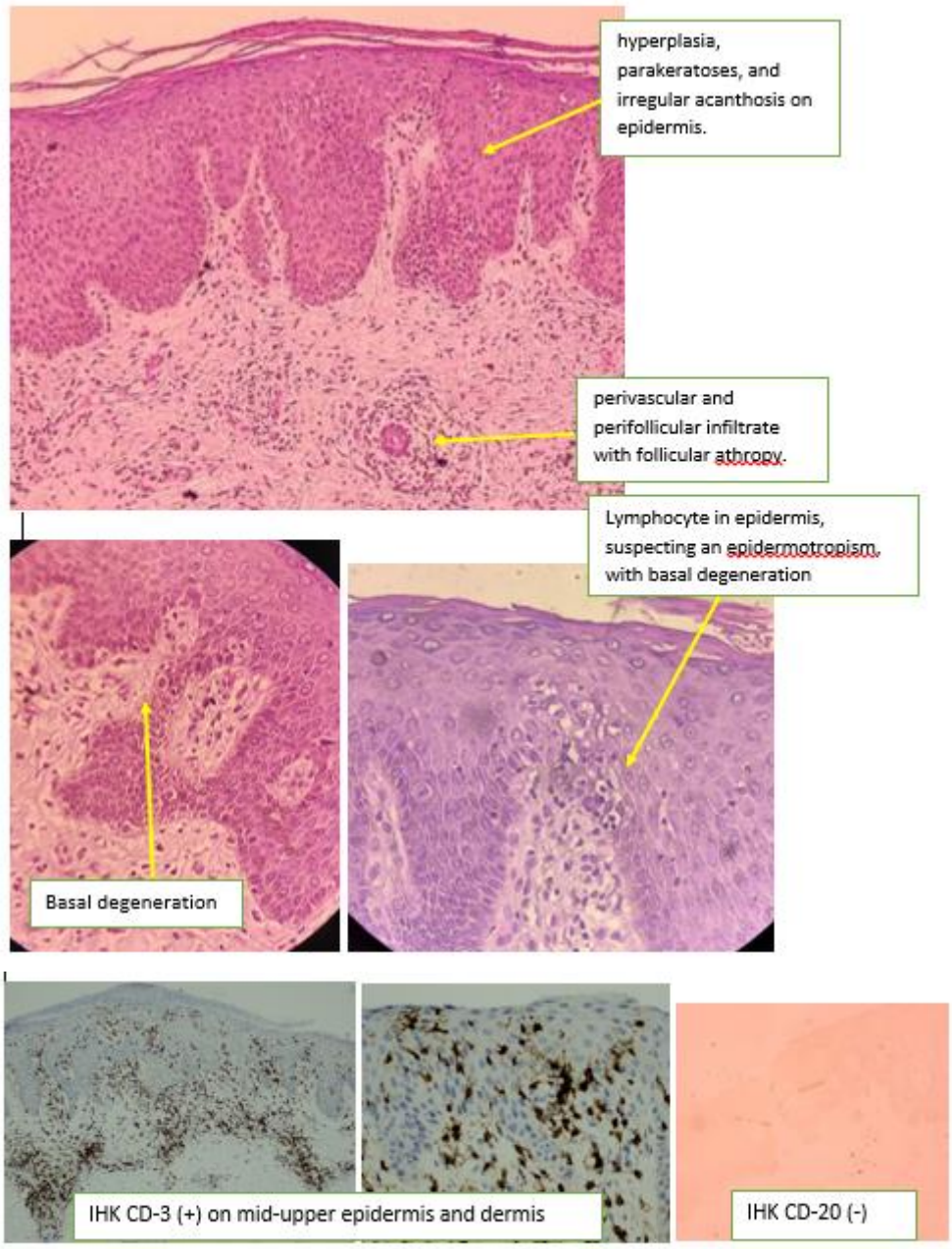

Figure 1B. Histopathology and Immunohistochemistry of the patient.

2.2 Case 2

Mr. S, a 65 years old man, married, worked as a farmer, lived in Pesisir Selatan, came Dermatology and Venereology Department outpatient clinic of Dr. M. Djamil Padang Hospital on July 13th 2022, with complaint reddish patch with fine white scale on almost the whole body that felt itchy which gradually developed since 1 month ago. This complaint appeared initially 4 months ago as itchy reddish patch with fine white scale on the scalp, chest, and back. Around 2 weeks later, the patient complained of intermittent fever, which was felt especially at night. Around 1 month ago, reddish patches and white scales are felt worsen on the scalp, chest and back, then the itchy reddish patch with fine white scale started to appeared on arm and leg. There was no history of new medications within the last 2 months before and

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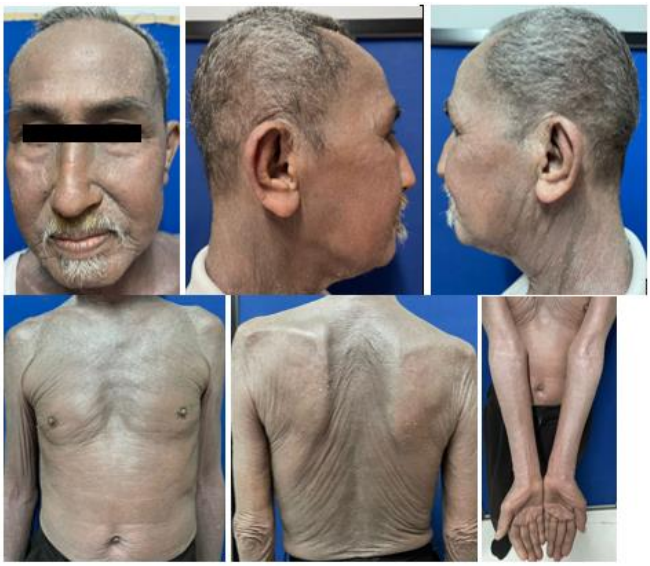
no history of blister in genital or oral. There is a history of intermittent reddish patch with white scales around the eyebrows and head dandruff since 5 years ago but the patient didn't treat this complaint. No history of frequent sneezing in the morning and asthma before. No history of reddish patch with micaceous white scale on the elbow and knee before. No history of using topical drugs before. Patient said he had weight loss within the last 3 months, without any history of chronic cough before. There was no history of diabetes mellitus, hypertention, renal disease and liver disease.

On physical examination, the vital sign was within normal limit. From head and neck examination, chest, abdomen, upper and lower extremities were within normal limit. There was no enlargement of the cervical, axillar, and inguinal lymph nodes. The body mass index was 16.5 (severely underweight).

Dermatological state revealed erythematous macules, hyperpigmented macules, whitish scales, and erosion on almost the whole body with universal distribution. Laboratory examination found increase of creatinine (1,6 g/dl). Other laboratory examination including routine blood examination, liver function, and blood sugar are within normal limit. There was no sezary cell found. From dermoscopy examination showed whitish scale and from trichoscopy found whitish-yellowish scale with erosion. Skin sample for histopathology and immunohistochemistry examination was taken.

Patient then hospitalized with diagnosis erythroderma ec susp. Seborrheic dermatitis. Patient treated with methylprednisolone 32mg/day PO, lansoprazole 1x30mg PO, cetirizine 1x10mg PO, lanolin oint 10% 30 minutes before taking a bath on upper part of body in the morning, and on lower part of body in the evening, hydrocortisone 2,5% on upper part of body in the morning, and on lower part of body in the evening. There was a bit improvement after 2 weeks of therapy. Patient then discharged from hospital and advised to control to Dermatology and Venereology department outpatient clinic. During control at the outpatient clinic, complaints of reddish spots and white scales in patients felt no significant improvement.

From histopathological examination found spongiotic reaction and interface dermatitis with basal cell degeneration. Epidermis acanthosis, irregular, spongiotic areas with degenerating keratocytes. The papillary dermis contains infiltrate of lymphocytes. lymphocyte invasion seen into the basal epidermis with some of them have irregular nuclei. In upper dermis found lymphocytes infiltrate and atrophied hair follicles with remnants of the arrector pili muscles. No perifollicular lymphocytic infiltrate and shoulder parakeratosis were found. These features could be found in seborrheic dermatitis but the possibility of cutaneous lymphoma could't be excluded. Immunohistochemistry examination is needed to confirm the diagnosis. From CD3 immunohistochemistry examination found there was dense clusters of CD3 (+) in the upper dermis, dermal papillae and some infiltrative cells into the epidermis (a feature of epidermotropism). The CD20 immunohistochemistry was negative. These microscopic findings suitable for CTCL. Patient then consulted to internal department for chemotherapy.



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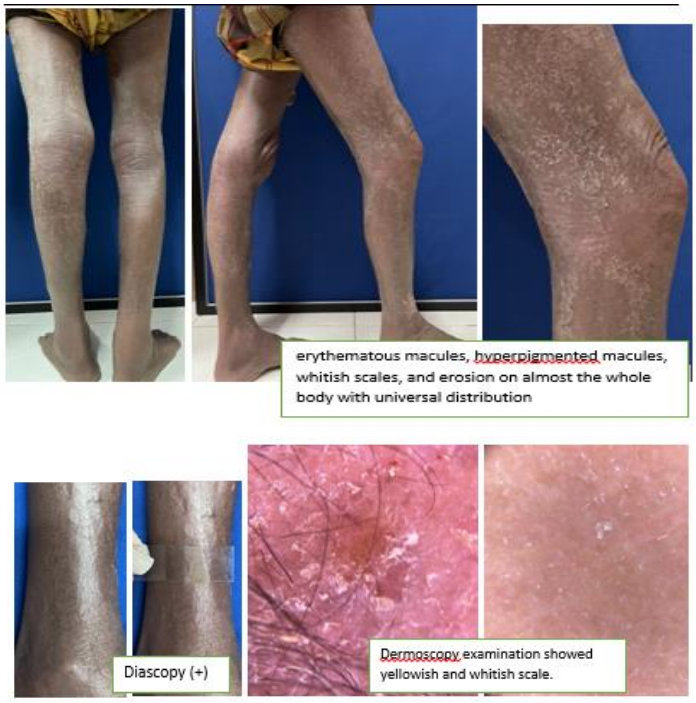

Figure 2A. Clinical presentation of the patient.

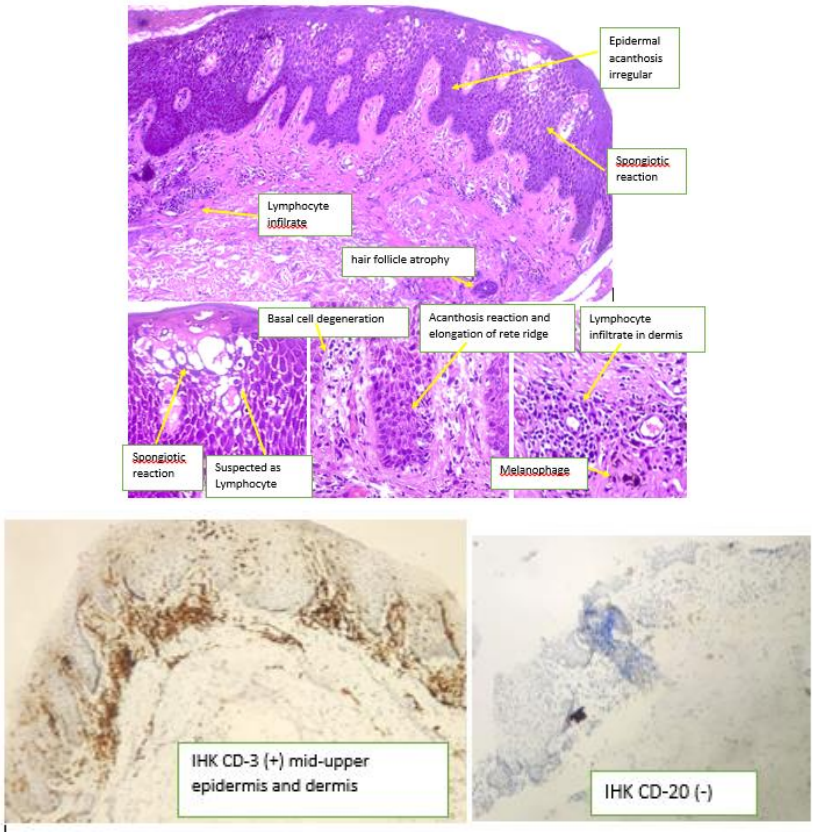

Figure 2B. Histopathology and Immunohistochemistry of the patient.

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2.3 Case 3

Mr. M, a 70 years old man, married, a retired civil worker, lived in Padang, came Dermatology and Venereology Department outpatient clinic of Dr. M. Djamil Padang Hospital on May 7th 2022, with complaint itchy reddish patch with white scale on almost the whole body that felt itchy which gradually developed since 2 weeks ago. This complaint appeared initially 2 years ago as itchy reddish patch with white scale on the arm, leg, both of palm and sole. Patient routinely controlled to Dermatology and Venereology Department outpatient clinic of Dr. M. Djamil Padang Hospital since 1 year ago with the diagnosis eritrhroderma ec atopic dermatitis. There was no history of new medications before reddish patch appeared and no history of blister in genital or oral. No history of intermittent reddish patch with yellowish scales around the eyebrows, nasolabial fold, chest or upper back. No history of head dandruff before. Patient have history of frequent sneezing in the morning and intermittent conjunctivitis without history of asthma. Patient sometimes felt dryness on his skin particularly on his hand. No history of reddish patch with micaceous white scale on the elbow and knee before. No history of using topical drugs before. Patient denied history of weight loss within the last 1 month. Patient had history of hypertention and routinely consuming amlodipine 1x10mg. There was no history of diabetes mellitus, renal disease and liver disease. Patient had received various oral medication to treat this condition including methotrexate, cyclosporine, topical treatment urea 10%, and hydrocortisone 2,5%, but these medications didn't give significant improvement.

On physical examination, the vital sign was within normal limit except the blood pressure was 150/80 mmHg. From head and neck examination, chest, abdomen, upper and lower extremities were within normal limit. There was no enlargement of the cervical, axillar, and inguinal lymph nodes. The body mass index was 24,6 (normoweight).

Dermatological state revealed erythematous macules, hyperpigmented macules, whitish scales, erosion, excoriation, and lichenification on almost the whole body with universal distribution. Laboratory examination found anemia (8.5 g/dl) and leucocytosis (14.900/mm^3). Histopathology examination was performed and revealed a psoriasiform reaction, hence the patient diagnosis changed to erythroderma ec psoriasis vulgaris. Patient then hospitalized and received secukinumab injection 150mg/week subcutaneously, cetirizine 1x10mg PO, lanolin oint 10% 30 minutes before taking a bath on upper part of body in the morning, and on lower part of body in the evening, and hydrocortisone 2,5% on upper part of body in the morning, and on lower part of body in the evening. After 2nd session of secukinumab injection, the patient began to have a fever, and the symptoms got progressively worse. The histopathology examination performed again with peripheral blood examination, immunohistochemistry CD3 and CD20 examination.

From histopathological examination found psoriasiform reaction and interface dermatitis with basal cell degeneration. Epidermis acanthosis, irregular, spongiotic areas. The papillary dermis contains infiltrate of lymphocytes. There was also focus of lymphocyte infiltrate at basal epidermis (epidermotropism) and upper dermis. No finding of microabcess munro nor spongiform pustules of kogoj. These features could be found in cutaneous lymphoma. From CD3 immunohistochemistry examination found there was dense clusters of CD3 (+) in the upper dermis, dermal papillae and some infiltrative cells into the epidermis (a feature of epidermotropism). The CD20 immunohistochemistry was negative. These microscopic findings suitable for CTCL. The peripheral blood examination confirmed the presence of Sézary cells, leading to the establishment of a diagnosis of Sézary syndrome. The patient was subsequently referred to the internal medicine department for chemotherapy.

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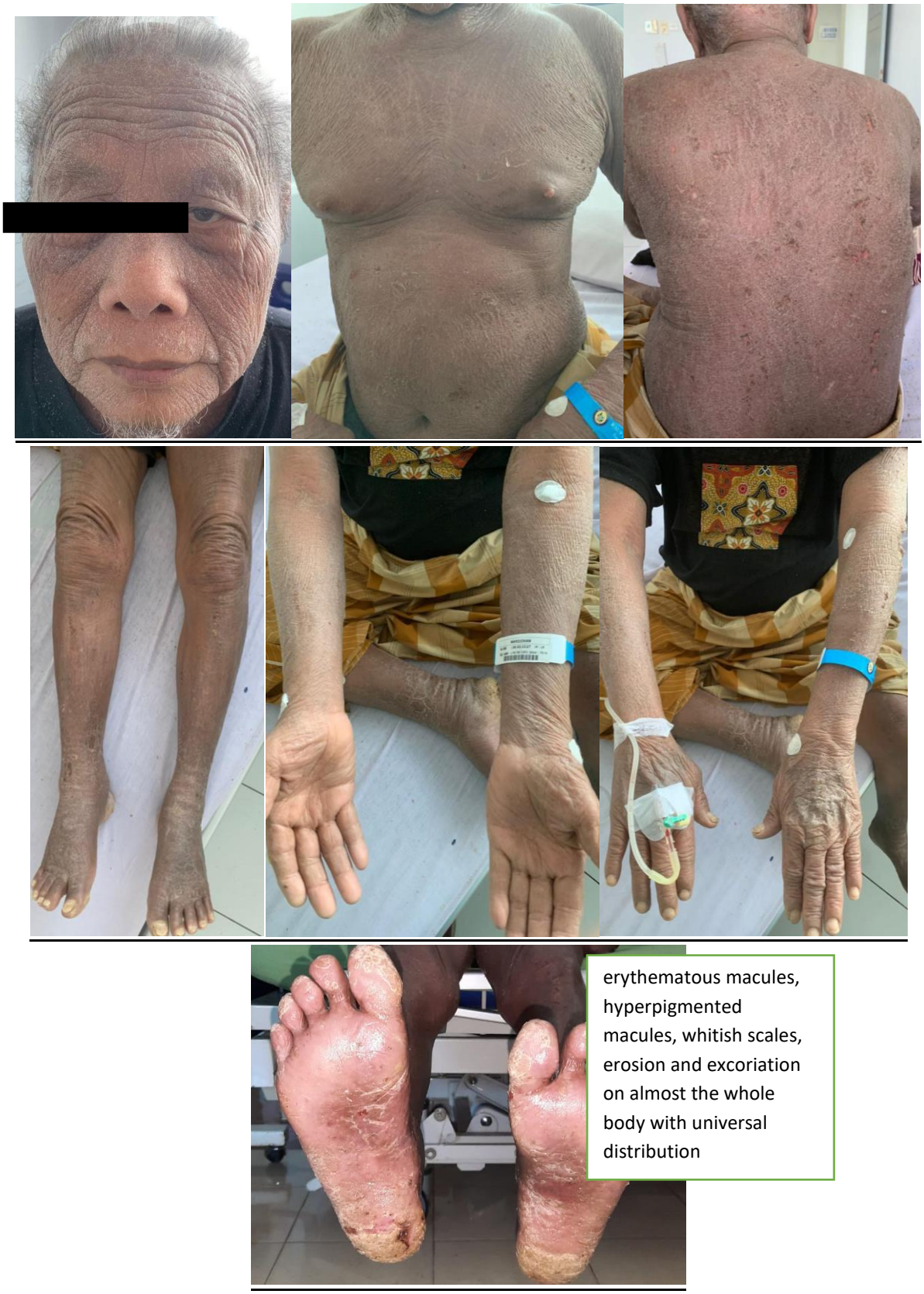

Figure 3A. Clinical presentation of the patient.

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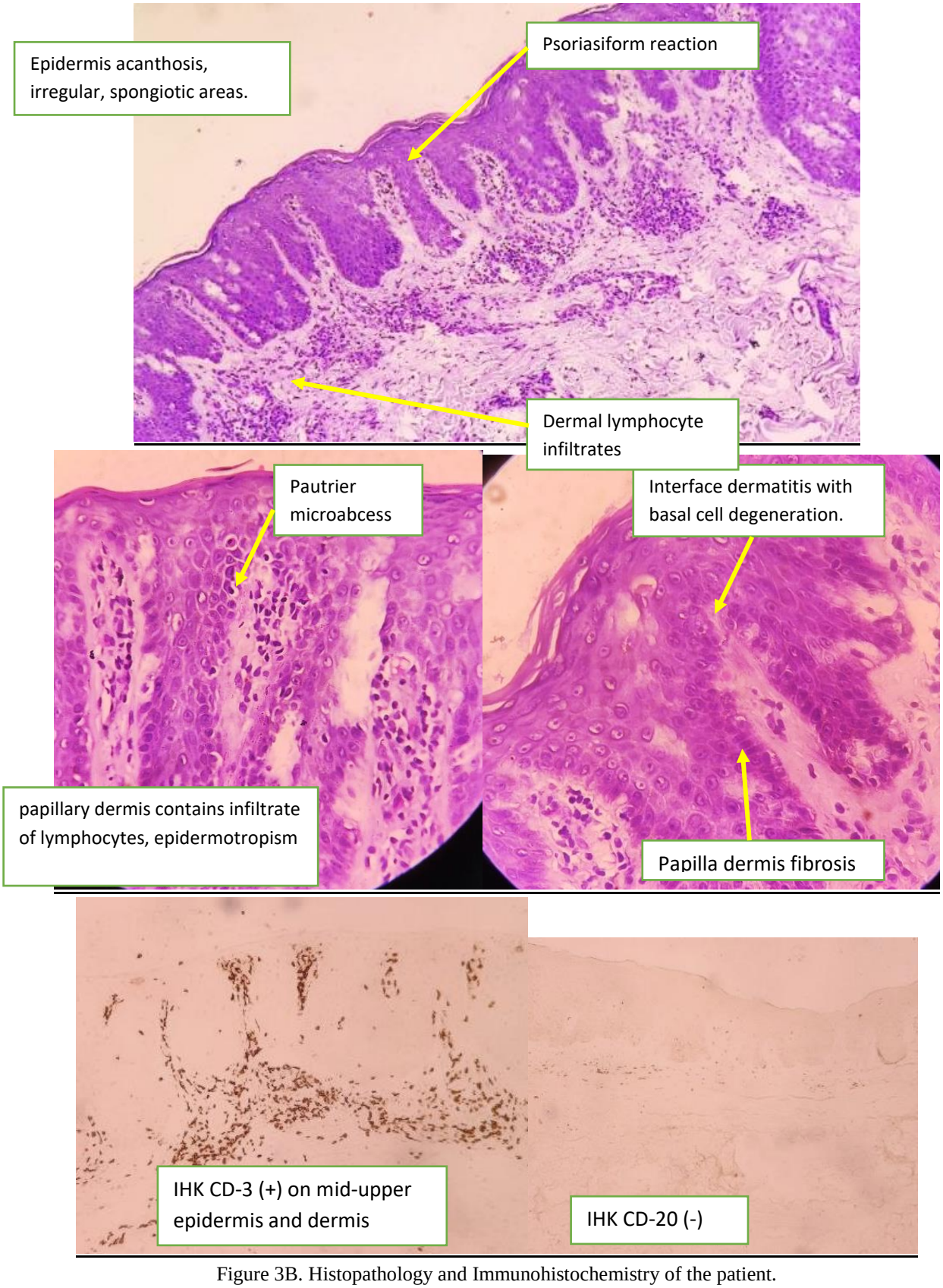

Figure 3B. Histopathology and Immunohistochemistry of the patient.

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2.4 Case 4

Mrs. R, a 33 years old woman, married, worked as a housewife, lived in Batusangkar, came Dermatology and Venereology Department outpatient clinic of Dr. M. Djamil Padang Hospital on October 14th 2021, with complaint reddish patch with fine white scale on almost the whole body that felt itchy which gradually developed since 2 weeks ago. This complaint appeared initially 2 months ago as itchy reddish patch with fine white scale on the scalp, behind the ear, upper back and both of the arm. Around 2 weeks later, the symptoms start to appeared on almost the whole body. There was no history of new medications within the last 2 months before and no history of blister in genital or oral. There was history of head dandruff when the patient was young but history of intermittent reddish patch with white scales around the eyebrows was denied. No history of frequent sneezing in the morning and asthma before. No history of reddish patch with micaceous white scale on the elbow and knee before. No history of using topical drugs before. Patient denied history of weight loss within the last 1 month. There was no history of diabetes mellitus, hypertention, renal disease and liver disease.

On physical examination, the vital sign was within normal limit. From head and neck examination, chest, abdomen, upper and lower extremities were within normal limit. There was enlargement of the left axillar, and left inguinal lymph nodes. The body mass index was 22,7 (normoweight).

Dermatological state revealed erythematous macules, whitish scales, and erosion on almost the whole body with universal distribution. Laboratory examination including routine blood examination, liver function, renal function, and blood sugar are within normal limit. The peripheral blood examination was not performed. From dermoscopy examination showed whitish scale with erosion. Skin sample for histopathology and immunohistochemistry examination was taken.

Patient diagnosed as erythroderma ec susp. Seborrheic dermatitis DDx psoriasis vulgaris. Patient treated with topical treatment urea cream 10% after taking a bath on upper part of body in the morning, and on lower part of body in the evening, hydrocortisone 2,5% on upper part of body in the morning, and on lower part of body in the evening. There was improvement after 2 weeks of therapy. Because the symptoms improved, the patient stopped the treatment.

From histopathological examination found epidermal hyperkeratosis, parakeratosis, and mild achanthosis. There was spongiosis in some areas with exocytosis of lymphocytes in the epidermis around the spongiosis areas. A section of the hair follicle is seen with inflammation of the perifollicle and an area of spongiosis in the epidermis above the hair follicle. There is lymphocyte infiltrate in the epidermis that form groups suspicious of a pautrier microabscess. In the upper dermis there are mild to moderate inflammatory cells consisting of lymphocyte infiltrate, an area with denser clusters of perivascular lymphocytes.

At first, the dermatopathologist suspect this as psoriasis vulgaris or dermatitis seborhoiec, but there is a suspicion of mycosis fungoides (MF). Immunohistochemistry examination is needed to confirm the diagnosis. From CD3 immunohistochemistry examination found there was dense clusters of CD3 (+) in the upper dermis, dermal papillae and some infiltrative cells into the epidermis (a feature of epidermotropism). These microscopic findings suitable for CTCL. Around 1 year later, patient complaint that the symptoms relapsed again. The laboratory examination revealed anemia, leucocytosis, and positive result of sezary cell in peripheral blood smear. The diagnosis of sezary syndrome was established. The patient then was consulted to internal department for chemotherapy.

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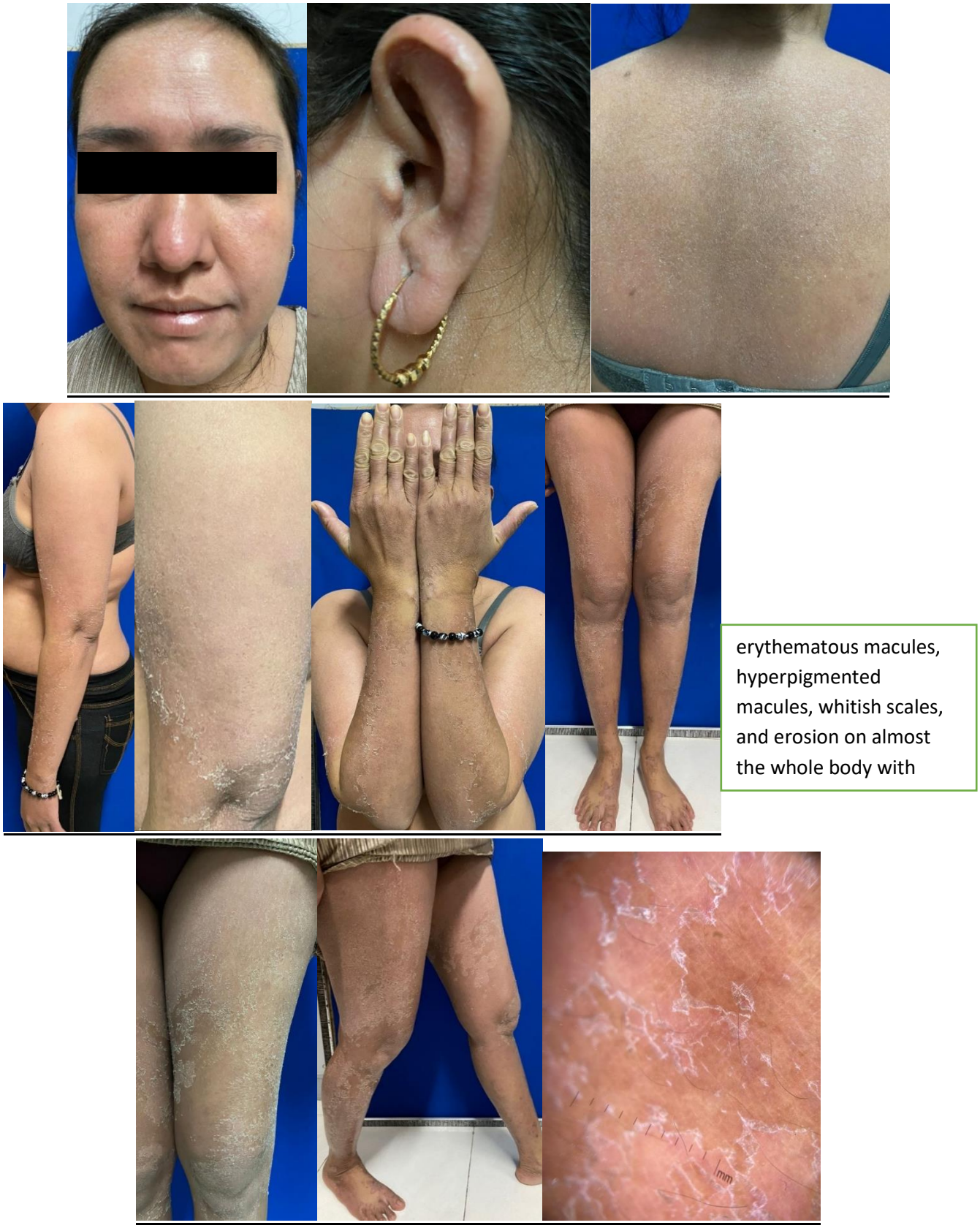

Figure 4A. Clinical presentation of the patient.

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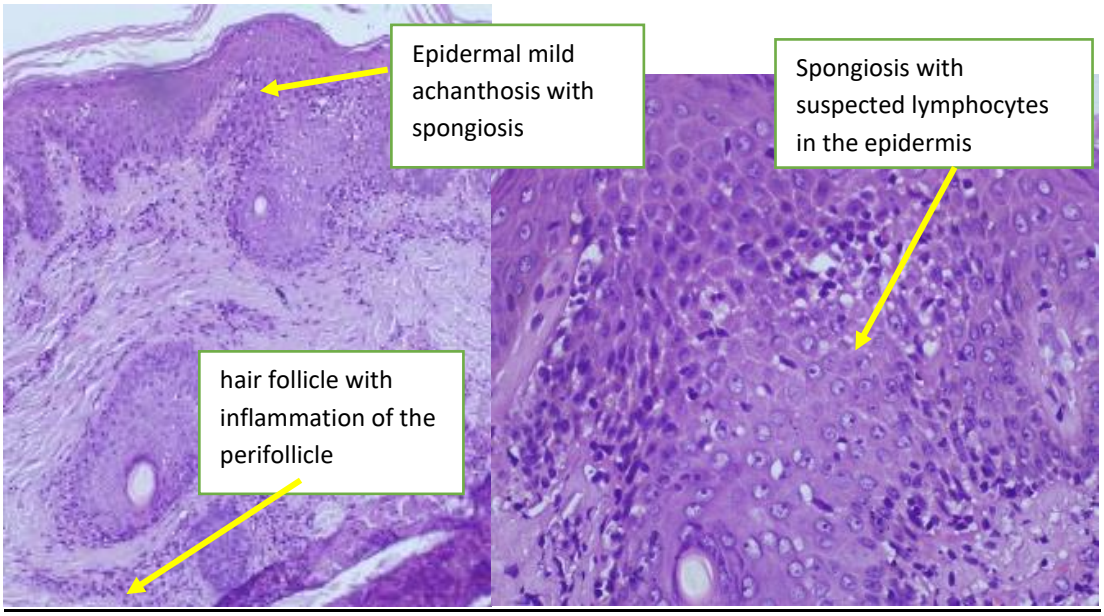

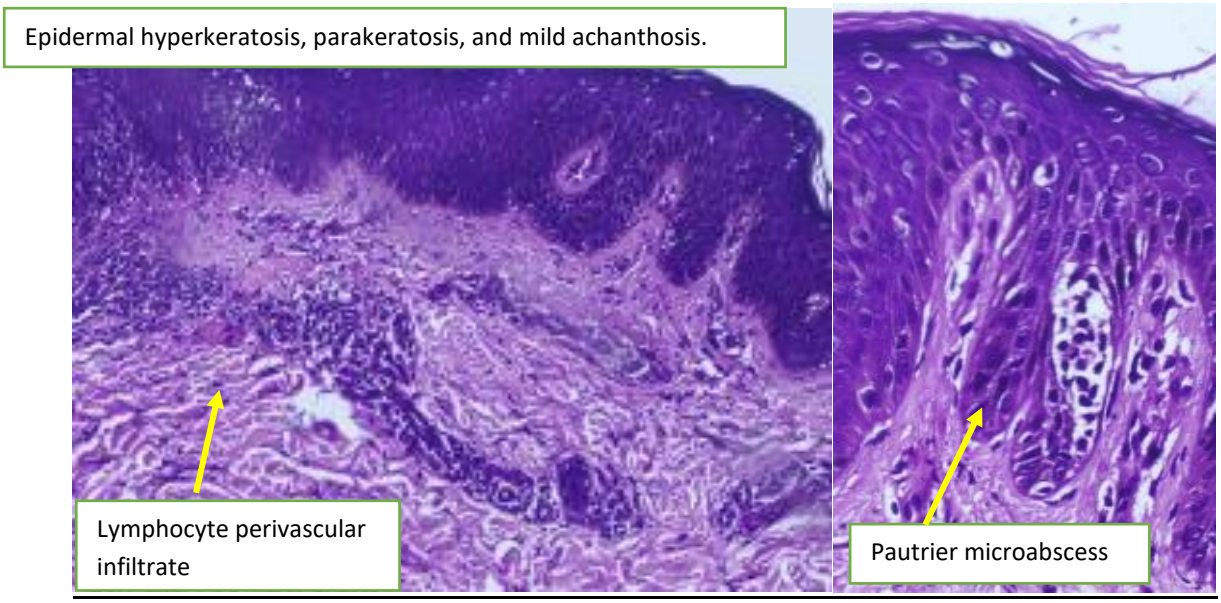

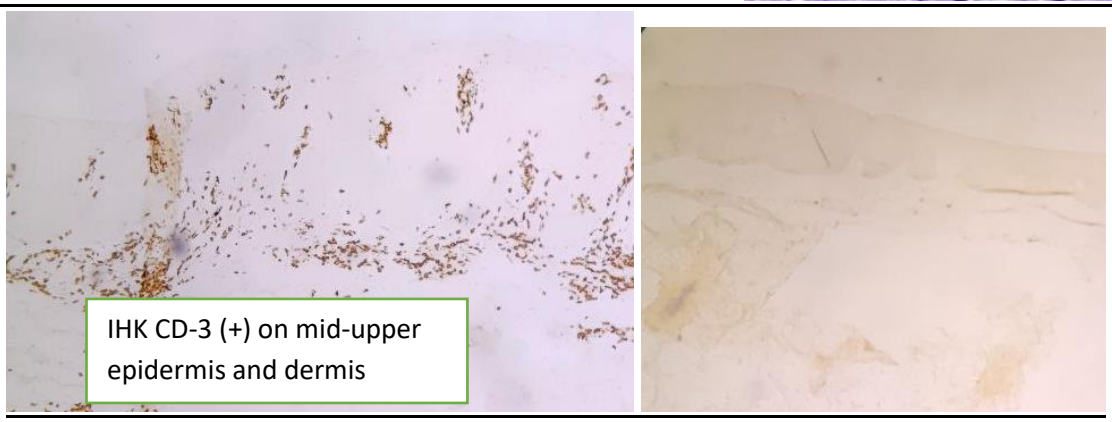

Figure 4B. Histopathology and Immunohistochemistry of the patient.

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3. DISCUSSION

From a retrospective study on the characteristics of erythroderma patients admitted to the Dermatology and Venereology Department at Dr. M. Djamil Padang General Hospital during the period of 2017-2019 (unpublished), it was found that the most common etiology of erythroderma was seborrheic dermatitis at 21 cases (32%), predominantly occurring in males over the age of 60. This differs from a study conducted by Barboza AC et al., where the most common etiology of erythroderma was pre-existing skin diseases: psoriasis vulgaris (25-50%), spongiotic dermatitis (5.12-25.3%) (atopic dermatitis 9%, contact dermatitis 9%, and seborrheic dermatitis 4%). These cases were predominantly observed in males with an average age of 52.57 years.[8-9] Another study from Turkey conducted by Askin et al. (2020) concluded that mycosis fungoides represents 12.8% of the cases as the etiology of erythroderma.[5]

In this report, we reported a case series consisted of four patients of Erythrodermic CTCL in M Djamil Hospital Padang from 2021-2023. Patient no. 1 is 85 years old male with E-MF that initially diagnosed as erythroderma ec. DS. Patient no. 2 is 65 years old male with E-MF that initially diagnosed as erythroderma ec. DS. Patient no. 3 is 70 years old male with Sézary syndrome that misdiagnosed as erythroderma ec AD and psoriasis vulgaris. Patient no. 4 is 33 years old female with Sézary syndrome that initially diagnosed as erythroderma ec. DS. Three out of four reported patients were over the age of 60, while only one of them was under 40 years old (33 years old). The initial suspected etiology of erythroderma in three of them was dermatitis seborrheic (DS).

In patient no. 1, we had tried to exclude any possibility that could be the etiology of erythroderma such as drug eruption, psoriasis, atopic dermatitis, contact dermatitis, systemic diseases and mycosis fungoides. The suspicion of dermatitis seborrheic (DS) arose due to the patient's history of head dandruff during their youth and the observed accentuation of skin involvement in the scalp and upper chest. No clinical findings supporting the suspicion of mycosis fungoides, including Sézary cells, were found. The histopathology examination did not reveal any findings indicative of dermatitis seborrheic (DS). Instead, the examination showed epidermotropism and a lichenoid reaction with acanthosis, which shifted the suspicion towards mycosis fungoides.

In patient no. 2, we had tried to rule out various potential causes of erythroderma such as drug eruption, psoriasis, atopic dermatitis, contact dermatitis, systemic diseases, and mycosis fungoides. The suspicion of dermatitis seborrheic (DS) arose due to the patient's history of dandruff 5 years ago, which was accompanied by the appearance of red patches with itchy scales on the face, chest, and upper back. The intermittent history of dandruff over the past 5 years raised suspicions of seborrheic dermatitis in this patient. Furthermore, the lesions in this patient appeared thicker on the forehead, around the nasolabial area, and around the eyebrows. Althrough patient had history of weight loss and the BMI was underweight, the suspicion to mycosis fungoides was not first suspected because the sezary cell in peripheral blood examination was negative. The consideration of seborrheic dermatitis as the etiology of the erythroderma also arises from the histopathological findings. Typically, seborrheic dermatitis would show spongiotic reaction in histopathology. However, in this case, the presence of acanthosis with interface dermatitis and atrophied hair follicles raised the possibility of mycosis fungoides.

In patient no. 3, the initial diagnosis of atopic dermatitis as the etiology of erythroderma was based on the patient's clinical history, including symptoms of frequent sneezing in the morning, intermittent conjunctivitis, and dryness on the skin. There was no suspicion of mycosis fungoides at that time. The patient was treated with several systemic medications, including methotrexate and cyclosporine, but there was no significant improvement in the condition. After several months of treatment without significant improvement, a biopsy was performed, which revealed a psoriasiform reaction consistent with psoriasis vulgaris rather than atopic dermatitis. Following the diagnosis of psoriasis vulgaris, the patient was treated with secukinumab. Unfortunately, after two weeks of treatment, the patient's symptoms worsened instead of improving. Histopathology examination then performed again. The examination revealed a psoriasiform reaction with interface dermatitis and lymphocyte infiltrate at the basal epidermis, indicating epidermotropism. These findings raised suspicion of mycosis fungoides. To confirm the diagnosis, immunohistochemistry examination using CD3 was conducted, which confirmed the presence of abnormal T-cell lymphocytes consistent with mycosis fungoides.

In patient no. 4, various possibilities for the etiology of erythroderma were considered and excluded. The suspicion of dermatitis seborrheic (DS) arose from the

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patient's history of head dandruff in her youth and the observed accentuation of skin involvement in specific areas such as the forehead, behind the ear, and upper back. No clinical findings supported the suspicion of mycosis fungoides initially. Upon histopathological examination, the dermatopathologist noted findings consistent with psoriasis vulgaris, including acanthosis, as well as findings suggestive of dermatitis seborrheic, such as spongiosis reaction and perifollicular inflammation. However, there was also evidence of lymphocyte infiltrate in the epidermis, forming Pautrier microabscesses. These additional findings raised the suspicion of mycosis fungoides. To confirm the diagnosis, CD3 examination was conducted, which confirmed the presence of abnormal T-cells consistent with mycosis fungoides.

The description of CTCL includes a cellular infiltrate with variable epidermal changes, such as irregular epidermal acanthosis with focal orthokeratosis and parakeratosis. Spongiosis is occasionally present, although usually mild. The papillary dermis shows fibrosis with thickened collagen bundles and scattered melanophages. These findings were also observed in all of the patient reported. The histological features of MF-erythroderma appear to be less pronounced compared to the findings in MF patch and plaque stages, making a histopathological diagnosis alone more challenging.[13-14] These minimal histopathological findings initially raised suspicions of erythroderma with seborrheic dermatitis, atopic dermatitis, or psoriasis vulgaris.

Erythroderma can occur at any stage of MF's evolution. The distinction between Sézary syndrome (SS) and MF-erythroderma is based on the absence of a significant number of "Sézary cells" in the peripheral blood. Differentiating erythroderma from Sézary syndrome/CTCL can be challenging, and physicians must consider clinical findings along with histological and immunological features. There is no single clinical presentation pathognomonic of Sézary syndrome/CTCL. The presence of atypical lymphoid band-like or perivascular infiltrates in the papillary dermis or the presence of Pautrier microabscesses may suggest but are not pathognomonic of Sézary syndrome/CTCL.[12-14]

Sezary syndrome is rare, with an estimated annual incidence of 30 to 40 cases, accounting for approximately 5% of all newly reported CTCL cases. Although little is known about the epidemiology of Sézary syndrome, overall, most patients with CTCL are diagnosed between the ages of 45 and 70 years (with an average age of 63 years).[14] Patient number 3 and 4 was diagnosed with Sézary syndrome at the age of 70 and 33 years old respectively.

CONCLUSION

We present a case series of four patients diagnosed with erythrodermic CTCL. Diagnosis of erythrodermic CTCL requires comprehensive histopathological examination and immunohistochemistry. While there are no specific histopathological features pathognomonic for this diagnosis, a meticulous analysis of histopathology samples is crucial. The possibility of CTCL/mycosis fungoides should be considered in all patients presenting with erythroderma.

REFERENCES

1. Khaled A, Sellami A, Fazaa B, Kharfi M, Zeglaoui F, Kamoun MR. Acquired erythroderma in adults: a clinical and prognostic study. J Eur Acad Dermatol Venereol. 2010;24:781-8.
2. Usatine RP, Smith MA, Chumley HS, Mayeaux EJ Jr. Erythroderma. In: Usatine RP, Smith MA, Chumley HS, Mayeaux EJ Jr. eds. The Color Atlas of Family Medicine, 2 ed. New York, NY: McGraw-Hill; 2013.
3. Ozkoca D, Uzuncakmak TK. Clinical Approach to Erythroderma. J Turk Acad Dermatol. 2022; 16(2):33-35
4. Sterry W, Muche JM. Erythroderma. In : Bolognia JL, Jorizzo Jl, Rapini RP, Horn TD, Mascaro JM, Mancini JA, Salasche JE, Saurat JH, Sting G eds. Dermatology. Ist Edition. Spain : Mosby, 2003: 165-74
5. Askin O, Altunkalem RN, Uzuncakmak TK, Toplu FŞ, Engin B. Erythroderma: A clinicopathological study of 47 cases from 2018 to 2020. Dermatol Ther 2020;33:e14342.
6. Tan GFL, Kong YL, Tan ASL, Tey HL. Causes and features of erythroderma. Ann Acad Med Singap 2014;43:391-394.
7. Grant-Kels JM, Fedeles F, Rothe MJ. Exfoliative dermatitis. In: Fitzpatrick's Dermatology in General Medicine. 8th ed. New York: McGraw Hill Medical; 2012.
8. Li J, Zheng HY. Erythroderma: a clinical and prognostic study. Dermatology. 2012;225: 154-62
9. Barboza AC, Candiani O, Ruelas ME. A Practical Approach to the Diagnosis and Treatment of Adult Erythroderma. Actas Dermosifiliogr. 2018: 1-10

10. Bagot M, Rudolf S. Cutaneous Lymphoma. In: Fitzpatrick's Dermatology in General Medicine. 9th ed. New York: McGraw Hill Medical; 2017.
11. Weinstock MA, Horm JW. Population-based estimate of survival and determinants of prognosis in patients with mycosis fungoides. Cancer 1988;62:1658-61
12. Wieselthier, Janet S. and Howard K. Koh. "Sézary syndrome: diagnosis, prognosis, and critical review of treatment options." Journal of the American Academy of Dermatology 22 3 (1990): 381-401.
13. Miyagaki T, Sugaya M. Erythrodermic cutaneous T-cell lymphoma: how to differentiate this rare disease from atopic dermatitis. J Dermatol Sci. 2011 Oct;64(1):1-6.
14. Patterson JW. (2020). Weedon's skin pathology 5th edition. Ch 42: Cutaneous Infiltrate – Lymphomatous and Leukemic. Pg 1236. Edinburgh: Churchill Livingstone/Elsevier.