Case Report

Ichthyosis with superimposed mycosis fungoides, a rare case of generalized erythema with malaise

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Abstract

Ichthyosiform eruption of mycosis fungoides is rare, and ichthyosis with superimposed mycosis fungoides is scarcely ever seen; only a few cases have been documented in the medical literature. Furthermore, the patients with ichthyosiform lesions may indicate the presence of an underlying systemic disease, paraneoplastic syndrome, or an atypical manifestation of cutaneous T-cell lymphoma. Thus, determining the accurate etiology is important to establish the correct diagnosis and subsequently facilitate its management. They should be evaluated thoroughly, and a skin biopsy is essential to rule out the possibility of ichthyosiform mycosis fungoides.

Keywords: Cutaneous T-cell lymphoma, erythroderma, exfoliative dermatitis, ichthyosis, mycosis fungoides

Introduction

Mycosis fungoides (MF) accounts for about 40 percent of cutaneous lymphomas, being the most frequent type of cutaneous T-cell lymphoma (CTCL).[1] The classic form of MF is defined as the patch, plaque, or tumor stage.[2] According to the World Health Organization European Organization for Research and Treatment of Cancer (WHO EORTC) updated version in 2018, there are three subtypes of MF, listed below: pagetoid reticulosis, granulomatous slack skin, and folliculotropic MF.[3] However, there are still some rare variants among MF, such as granulomatous MF, poikiloderma, hyper and hypo-pigmented MF, and solitary MF.[4-9] Amid them, ichthyosiform MF is not as well-understood as other forms of MF, and this particular manifestation as an ichthyosiform eruption of mycosis fungoides is scarcely ever seen, and only a few cases have been documented in the medical literature.[10,11] Moreover, ichthyosiform lesions may indicate the presence of an underlying systemic disease, paraneoplastic syndrome, or an atypical manifestation of cutaneous T-cell lymphoma.[12,13] Hence, determining the accurate etiology is important to establish the correct diagnosis and subsequently facilitate its management. We report a case presenting as ichthyosis with superimposed mycosis fungoides and generalized erythema with malaise.

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Case Report

A 74-year-old male with no obvious systemic underlying disease before, presented at Kaohsiung Medical University Hospital emergency room with generalized itchy erythema and malaise within one month's duration. He had no pre-existing dermatosis except dry skin since his childhood (his mother had similar dry skin), neither prior medical problems nor medication use before this episode. His vital signs were stable, but fatigue and malaise were noted. The physical examination showed diffuse erythematous scaling plaques with confluence over abdomen and extremities, especially deck chair sign noted on the abdomen, ichthyosis-like, xerosis and scaling spreading over the scalp, face, back, extremities, and periorbital area erythema [Figure 1]. After oral form antihistamine and topical ointment use, the patient was admitted to our dermatology ward the next day.

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After admission, we arranged several examinations for the more detailed survey, including laboratory data [Table 1], skin biopsy over the abdomen [Figure 2], chest X-ray [Figure 3], and whole-body PET (positron emission tomography) scan [Figure 4]. The laboratory data showed eosinophilia, immunoglobulin E elevated, C-reactive protein elevated, and abnormal lactate dehydrogenase, squamous cell carcinoma antigen; in contrast, the infection parameter, including Mycoplasma and Herpes simplex virus, were both negative, and autoimmune profile showed negative finding either. The chest X-ray revealed no specific finding. The pathology of skin biopsy over the abdomen revealed: epidermal acanthosis, spongiosis, and atypical lymphocytes exocytosis; also, the immunostain study of CD3 and CD4 showed scattered atypical T-cell lymphocytic infiltration and density of CD7 and CD8 lymphocytes was decreased, that is, the pathological result showed a predominance of CD3 and CD4 T-cells, which was compatible with mycosis fungoides. As for the PET scan to rule out the potentiality of systemic involvement of disease, it showed no obvious abnormal fluorodeoxyglucose (FDG) uptake in the internal organs; that is, the paraneoplastic syndrome was less likely.

According to the clinical features and pathology of skin biopsy, the diagnosis of ichthyosiform mycosis fungoides (IMF) has been established. The treatment goal of early-stage MF is to ensure the disease process is stable and prevent possible progression to overt MF. Our treatment plan is narrowband ultraviolet B phototherapy, and most crucially, we arrange examinations every three to six months to ensure that the character of the process is stable.

Discussion

Ichthyosiform eruption as a manifestation of mycosis fungoides is rare; furthermore, ichthyosis with superimposed mycosis fungoides is scarcely ever seen and just a few such cases have been presented in the medical literature.[10,11] Ichthyosiform MF can be subdivided into three types on the basis of the clinical findings: ichthyosiform lesions one and only, ichthyosiform eruption accompanied by classical MF, ichthyosiform eruption in combination with other atypical variants of MF.[10,11]

However, **our patient was separated from the above three categories. He presented with ichthyosis with superimposed mycosis fungoides**.[14] He had diffuse xerosis and scaling over the scalp, back, and extremities since his childhood, and his mother had similar dry skin. He presented at our emergency department with generalized erythematous scaling plaques and malaise, which was challenging to diagnose.

Generalized erythema is a rare and severe dermatological manifestation of quite a few diseases, including pre-exist dermatosis, like psoriasis, atopic dermatitis, and drug hypersensitivity reactions; also, paraneoplastic syndrome or secondary to some malignancies was possible.[15,16] Above all, due to its systemic complications, including thermoregulatory disturbance, electrolyte imbalance, dehydration, high output heart failure, hypoalbuminemia, and septicemia, it is a **potentially life-threatening situation**.[17-19] Thus, we performed several surveys, including a skin biopsy to **find out the culprit**

Table 1: Laboratory data

Variable	Reference Range	On admission
Red cell count (per uL)	4,310,000-5,950,000	5,210,000
Hemoglobin (g/dL)	13.4-17.2	15.3
Hematocrit (%)	39.8-50.7	45.7
Mean corpuscular volume (fl)	83.4-98.5	87.7
Red cell distribution width (%)	11.7-14.7	14.3
Platelet count (per uL)	160,000-370,000	223,000
White cell count (per uL)	4,140-10,520	8,310
Differential count (%)		
Neutrophils	41.8-70.8	57.0
Lymphocytes	20.7-49.2	27.4
Monocytes	3.6-9.2	5.9
Basophils	0.2-1.5	0.4
Eosinophils	0.5-7.5	9.3
Band	0.0-5.0	0.0
C-Reactive Protein (mg/L)	<5.0	14.82
Natrium (mmol/L)	136-144	140
Potassium (mmol/L)	3.5-5.1	4.1
Blood urea nitrogen (mg/dL)	8.0-20.0	11.7
Creatinine (mg/dL)	0.64-1.27	1.25
Glutamic Oxaloacetic Transaminase (IU/L)	10-42	31
Glutamic Pyruvic Transaminase (IU/L)	10-40	30
Thyroid stimulating hormone (uIU/mL)	0.34-5.60	1.15
Free T4 (ng/dL)	0.61-1.12	0.68
Albumin (g/dL)	3.5-5.0	4.09
Erythrocyte sedimentation rate (mm/h)	0-15	14
Mycoplasma Immunoglobulin M (ratio)	<0.9	0.239 (Negative)
Herpes simplex virus Immunoglobulin M (ratio)	<0.8	0.105 (Negative)
Herpes simplex virus Immunoglobulin G (ratio)	<0.8	5.087 (Positive)
Immunoglobulin E (IU/mL)	<87.0	95.6
Complement 3 (mg/dL)	90-180	132
Complement 4 (mg/dL)	10.0-40.0	37.6
Antinuclear antibody	<1:40	<1:40 (Negative)
Anti double stranded DNA (IU/mL)	<10.0	1.3
Anti-Ro (EliAU/ml)	<7.0	0.3
Anti-La (EliAU/ml)	<7.0	<0.3
Lactate Dehydrogenase (IU/L)	98-192	321
Carcinoembryonic antigen (ng/mL)	0.00-5.00	2.82
Squamous cell carcinoma antigen (ng/mL)	0.0-1.5	49.2
Alpha fetoprotein (ng/mL)	<9.00	2.77
Beta human chorionic gonadotropin (mIU/mL)	<5.0	<0.6

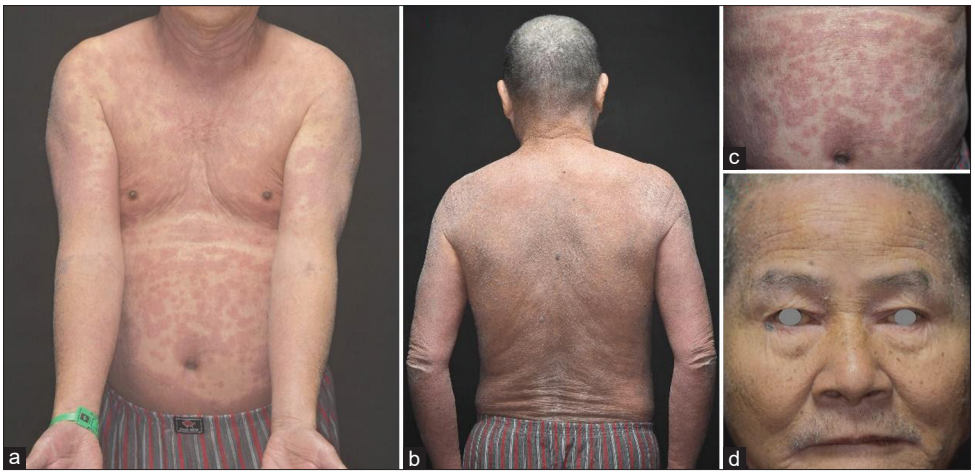

Figure 1: Skin presentation when admission (a): Diffuse erythematous scaling plaques with confluence over abdomen and extremities (b): Xerosis and scaling over the scalp, back, and extremities (c): Deck chair sign over the abdomen (d): Periorbital erythema

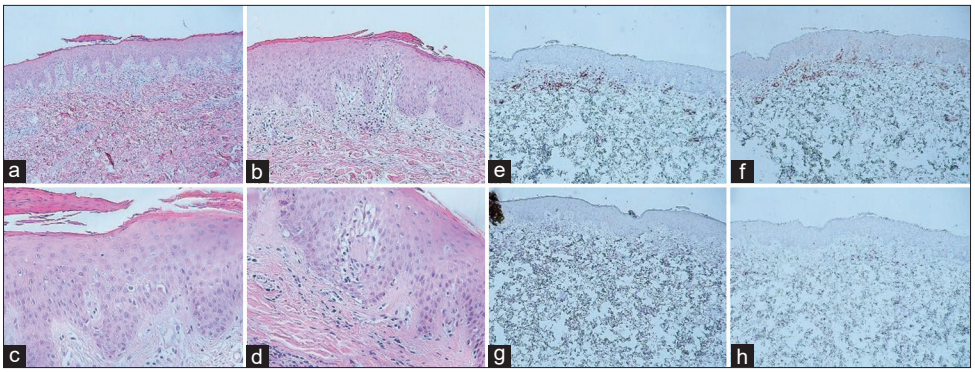

Figure 2: Pathology of skin biopsy over abdomen:(a-d): Epidermal acanthosis, spongiosis, and atypical lymphocytes exocytosis (e-h): Predominance of CD3 and CD4 T-cells with CD7 and CD8 T cell deficiency(e): CD3: Positive (f): CD4: Positive (g): CD7: Negative (h): CD8: Negative

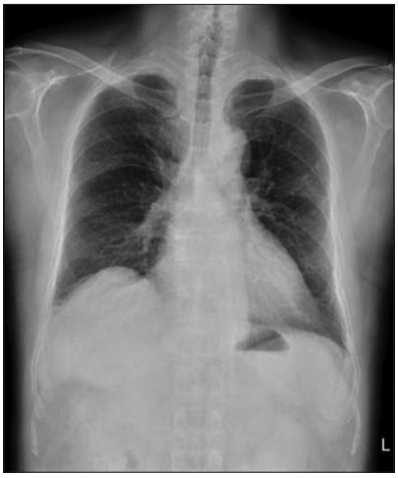

Figure 3: Chest X-ray: No obvious cardiopulmonary lesion

underlying cause, and finally, the diagnosis of ichthyosis with superimposed mycosis fungoides was established.

To sum up, ichthyosiform mycosis fungoides (IMF) is a rare type of early MF, and **patients with ichthyosiform lesions may indicate the presence of an underlying systemic disease, paraneoplastic syndrome, or an atypical manifestation of cutaneous T-cell lymphoma**[12,13]; therefore, determining the accurate etiology is important to obtain the correct diagnosis and subsequently facilitate its management. They should be evaluated thoroughly, and a skin biopsy is essential to rule out the possibility of IMF. Last but not the least, **ichthyosis is associated with some malignancies**, including lymphoma, breast cancer, and lung cancer[12,13]; thus, **exclusion of a neoplasm behind this pattern is aprecedence**. **Ichthyosis may indicate the presence of malignancies via finding of our case**, which is manifested as general erythema and later diagnosed as ichthyosiform mycosis fungoides. Thus, this case adds to the literature on clinical features of ichthyosis with superimposed mycosis fungoides and reminds us of the importance of **defining the relationship between ichthyosis and potential malignancies, which may** be the potential future research.

Conclusion

Generalized erythema is a rare and severe dermatological manifestation, and it is a potentially life-threatening situation. Since its possible fatal complications, such as electrolyte imbalance, dehydration, heart failure, even sepsis, and possible

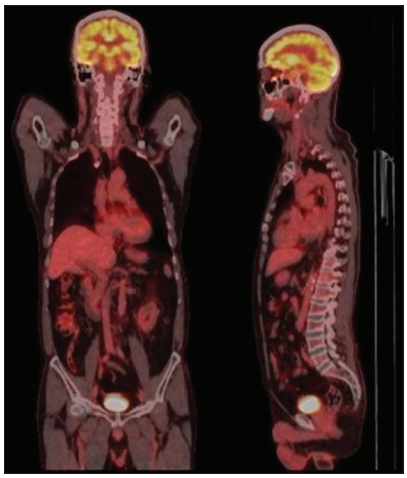

Figure 4: PET scan:Physiological FDG distribution in the brain, mediastinum, heart, liver, kidneys, and urinary bladder. No obvious abnormal FDG uptake in the internal organs

paraneoplastic syndrome are secondary to some malignancies, we should **lay emphasis on it and perform several surveys, including a skin biopsy to find out the culprit underlying cause so as to manage it properly and immediately**. **Ichthyosis with superimposed mycosis fungoides is one of the causes of generalized erythema** and may be **encountered by primary care physicians**; the treatment goal of early-stage mycosis fungoides is to ensure the disease process is stable and prevent possible progression to overt mycosis fungoides.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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