Folliculotropic Mycosis Fungoides Presenting with Secondary Cutis Verticis Gyrata in a Filipino Male*

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ABSTRACT

Mycosis fungoides (MF) is the predominant type of primary cutaneous lymphoma, accounting for at least 40%. It usually occurs in middle to late adulthood with the average age of diagnosis between 55 and 60 years. Folliculotropic MF is a variant that clinically presents with patches, plaques, and unusual hair loss within lesions. It predominantly involves the head and neck region. This usually presents with intense pruritus and commonly with secondary bacterial infection. Cutis verticis gyrata is a rare skin condition characterized by thickening and folding of the scalp, resulting in a furrowed or ridged appearance, and rarely associated with mycosis fungoides. A 46-year-old male, Filipino, presented with a 1-year history of generalized follicular papules, erythematous plaques, and subsequently presented with cutis verticis gyrata. Histopathological examination revealed findings consistent with folliculotropic MF. This was further confirmed by CD3, CD4, CD5 which revealed strongly positive immunohistochemical staining. To the best of our knowledge, there is currently only one published case of folliculotropic MF that presented with cutis verticis gyrata and only four cases of cutis verticis gyrata published locally.

Keywords: Mycosis fungoides, cutaneous lymphoma, cutis verticis gyrata, folliculotropic mycosis fungoides

INTRODUCTION

Mycosis fungoides, also known as classic MF, is the most prevalent primary cutaneous lymphoma.[1] It tends to occur more frequently in males typically between the ages of 55 and 60. It is believed that genetic factors, as well as prolonged exposure to allergens or carcinogens found in industries such as glass, pottery, paper, and wood, can predispose individuals to develop mycosis fungoides. The disease can be classified based on the appearance of skin manifestations, which include patches, plaques, or tumors. These skin lesions typically appear on non- sun exposed areas and can range from being asymptomatic to being associated with intense pruritus. In some cases, patients may also experience erythroderma, often with sparing of skin folds, described as the "deck chair" sign.[1] Additionally, patients may report symptoms such as fever, chills, weight loss, general discomfort, difficulty sleeping due to severe pruritus, and problems regulating body temperature.[1]

Folliculotropic MF is a variation that shares similarities with the classic form but primarily affects the head and neck area. It is identified by the infiltration of T-cells targeting hair follicles, potentially resulting in the degradation of hair follicles with or without mucinous involvement. In contrast to the classic form, folliculotropic MF displays follicular papules, acneiform lesions, indurated plaques, and tumors.[1]

Cutis verticis gyrata is a rare skin condition characterized by scalp thickening and folding, leading to a furrowed appearance commonly seen in diseases including neuropsychiatric pathologies, inflammatory, genetic, and endocrine pathologies.[2] It can be further divided into three distinct forms: primary essential, primary non-essential, and secondary.

We report a case of a Filipino male who presented with severe pruritus and widespread follicular papules, erythematous plaques, and nodules, along with localized hair loss, and convoluted folds and furrows on the scalp. The patient was diagnosed with folliculotropic MF based on examination of histopathological and immunohistochemical analysis. This case report will discuss the clinical features and relate the histopathological findings in a patient with folliculotropic MF and cutis verticis gyrata.

CASE REPORT

A 46-year-old male, Filipino, teacher from Batangas, with Fitzpatrick skin type IV presented with generalized papules and plaques. Lesions started one year prior to consultation when patient noted appearance of erythematous papules and plaques on the abdominal area. The lesions were associated with daily, intermittent pruritus graded 7/10. He had no known exposure to

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carcinogens from glass factories, potteries, mineral dust, wood burning, paper, and wood industry. He had no fever, weight loss, malaise. The lesions were noted to spread on the trunk and lower extremities with associated increased in pruritus despite application of potent steroids.

One month prior to consultation, along with persistence of previous lesions on the trunk and upper and lower extremities, the patient noted intensely pruritic tumors on the face and scalp. There was subsequent focal loss of hair on the scalp, face, and body. Impaired sweating was noted on the lesions.

The review of systems, past medical and family histories were unremarkable. He has been a non-smoker, non-alcoholic beverage drinker and denied illicit drug use. The patient worked as a teacher, living with his wife, niece, and nephew. He has no history of recent travels to other cities or provinces.

Cutaneous examination showed multiple, well-defined, discrete erythematous papules, plaques, and nodules on the head with hair loss noted on the eyebrows. There were well-circumscribed convoluted folds and deep furrows observed on the scalp with focal hair loss noted on the folds. The patient also presented with grouped follicular erythematous papules and indurated plaques on the chest, abdomen, and bilateral upper and lower extremities. Generalized erythema of the trunk and extremities was noted, with well to ill-defined areas of sparing on the folds of the abdomen, axillary areas, antecubital areas, and legs. Focal desquamation was observed on the palms and soles, while no nail changes or lesions on the oral mucous membranes were present (Figure 1). Furthermore, no palpable cervical and axillary lymphadenopathy was detected.

Blood chemistries revealed elevated triglycerides and HDL (high-density lipoprotein). A complete blood count showed leukocytosis, while the red blood cells appeared normal in size, shape, and color on the peripheral blood smear. No nucleated red blood cells or evidence of inclusions were observed. The WBC distribution was appropriate, with no reactive or immature forms detected. The platelets appeared normal in number and morphology. Furthermore, urinalysis findings and chest radiography were normal.

Punch biopsies were taken on different sites. The biopsy from a nodule on the scalp (Figure 2) showed that the epidermis is flat with grenz zone underneath it. Throughout the dermis are infiltrates of atypical lymphocytes with hyperchromatic nuclei and irregular nuclear contours. There were plenty of atypical mitotic figures noted. Similar cells in a lichenoid pattern was appreciated in the biopsy taken from the erythematous plaque on the abdomen. The follicles showed deposits of mucin on alcian blue staining (Figure 3). The atypical cells stained positive with CD3, CD4, CD5, and CD45 and were negative for CD20, CD8, CD7, CD30, confirming that this is a T-cell lymphoma (Figure 4).

Correlation of the patient's clinical features and the histopathological results established the diagnosis of folliculotropic MF. The patient was prescribed a combination of clobetasol propionate 0.05% ointment and petroleum jelly, to be applied twice daily for one month which offered slight reduction in size of lesions with a significant decrease in pruritus, and better sleeping patterns. The patient was referred to the Medical Oncology department for further evaluation and treatment. However, he opted not to comply due to his personal preference.

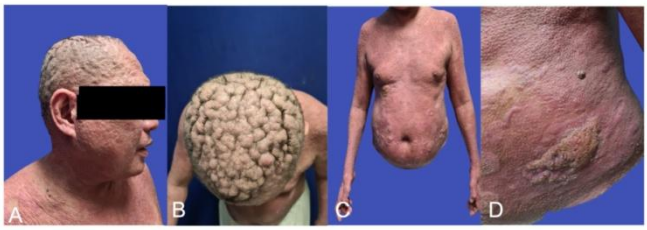

Figure 1. Clinical presentation of the patient showed A) pale erythematous papules and nodules on head, B) well-circumscribed convoluted folds and deep furrows on the scalp with focal hair loss noted on the folds, C) generalized erythema with well to ill-defined areas of sparing on the folds of the abdomen, axillary areas, antecubital areas and, D) grouped follicular erythematous papules with indurated plaques on the chest, abdomen.

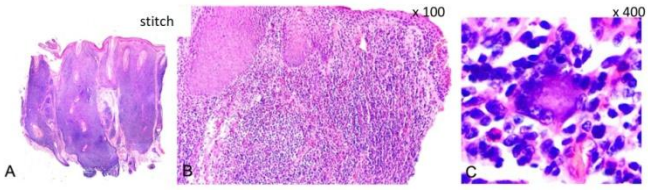

Figure 2. Photomicrograph of the nodule from the scalp showed A) a flattened side with grenz zone beneath it. Throughout the dermis are dense infiltrates of lymphocytes in a nodular pattern. B) Focal areas showed epidermotrophism of lymphocytes. C) Close up showed atypical lymphocytes with numerous eosinophils. H&E stain; original magnification: A) stitch view B) x 100 C) x400

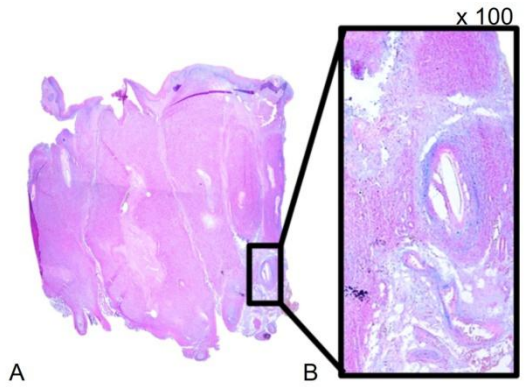

Figure 3. Alcian blue stain showed the presence of follicular mucinosis. (Original magnification A) stitch view B) x 100)

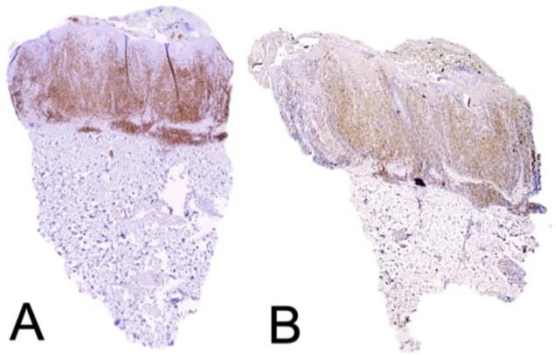

Figure 4. Immunohistochemical staining. A) CD3 and B) CD4 stain showed strong cytoplasmic staining and membrane accentuation. (original magnification A) and B) stitch view)

DISCUSSION

According to the 2016 revision of the World Health Organization[3], folliculotropic MF, the most frequent variant, is characterized by invasion of hair follicles by atypical T-cells. It comprises only 5% of all lymphomas. Other variants include pagetoid reticulosis, granulomatous slack skin, and hypopigmented MF. Currently, there are only a few published studies on folliculotropic MF.

Early diagnosis is challenging due to unspecific signs and symptoms. Folliculotropic MF affects men more than women, lesions commonly seen on the head and neck region.[1] However, a study by Gerami et al.[4], showed that it is not uncommon for it to manifest on the trunk. Studies conducted by Van Doorn et al.[5] and Gerami et al.[4] characterizing folliculotropic MF cases showed that patients presented with erythroderma, patches, plaques, papules, nodules and tumors, acneiform lesions, madarosis, scalp scarring alopecia, cutis verticis gyrata. These lesions are associated with pruritus and commonly develop secondary bacterial infection[1]. Our patient exhibited erythroderma with intensely pruritic erythematous follicular papules and plaques on the head, trunk, extremities and hair loss but had no episode of secondary bacterial infection which is consistent with the characteristic skin lesions of folliculotropic MF. As the disease advances, bulky tumor masses form, and the face may take on a leonine appearance.[4] Our patient exhibited with well-defined cutis verticis gyrata with localized hair loss specifically on these folds.

Findings from available literature, including reports by Gerami et al.[4], Van Doorn et al.[5], Muniesa et al.[6], and Demirkesen et al.[7], identify alopecia, papules, plaques, tumors, and nodules as the most common clinical features of folliculotropic MF. Our patient's presentation aligns with these descriptions, except for the presence of acneiform lesions and leonine facies. Interestingly, among these studies, only one case has been reported with cutis verticis gyrata, underscoring its rarity even within the folliculotropic MF variant. To the best of our knowledge, the present case represents only the second documented case worldwide and the first reported in the Philippines.

Cutis verticis gyrata is an uncommon skin disorder characterized by excessive growth of the skin on the scalp or face, resulting in folds resembling the convolutions of the cerebral cortex. It has been suggested that elevated levels of growth hormone (GH) and insulin-like growth factor 1 (IGF-1) play a role in its pathogenesis. Our patient subsequently developed cutis verticis gyrata. Chamli et.al described the three distinct forms of cutis verticis gyrata: primary form either essential or non-essential, and secondary form.[2]

Primary essential cutis verticis gyrata affects the skin without any associated underlying conditions. On the other hand, primary non-essential cutis verticis gyrata is often associated with neuropsychiatric abnormalities and ophthalmological abnormalities. It is also the most common form. Our patient did not have any associated conditions previously mentioned.

Secondary cutis verticis gyrata occurs when there are modifications in the skin trophicity due to associated abnormalities or underlying conditions such as liver diseases, paraneoplastic syndromes (associated with certain types of cancers), inflammatory dermatoses (psoriasis and eczema), genetic conditions (neurofibromatosis, Noonan syndrome, Turner syndrome etc.), endocrine disorders (diabetes mellitus, myxedema etc.) and iatrogenic causes such as using minoxidil (a medication for hair loss). Our patient presented with the secondary form of cutis verticis gyrata from the folliculotropic MF.

In total, 36 published cases of cutis verticis gyrata were identified in the literature. Of these, 10 were primary essential, 3 primary non-essential, and 23 secondary. Notably, only three cases have been reported in the Philippines, underscoring the rarity of this condition in the local setting. A detailed summary of these published cases is provided in Supplementary Table 1.

Histologically, folliculotropic MF is characterized by the presence of infiltrates surrounding the hair follicles, ranging from perifollicular to diffuse patterns. These infiltrates consist of T cells with cerebriform and hyperchromatic nuclei. In most cases, there is follicular mucinosis, which can be visualized through Alcian blue, or colloidal iron staining as seen in our patient. While infiltration of the follicular epithelium (folliculotropism) may occur, concurrent infiltration of the epidermis (epidermotropism) is uncommon in folliculotropic MF.[8] Our patient exhibited a dense, diffuse, nodular dermal infiltrate of small to medium-sized atypical lymphocytes and eosinophils. This infiltration was associated with folliculotropism, leading to focal flattening of some areas of the epidermis. Additionally, there were observed epidermotropism and superficial perivascular infiltrates, all consistent with the histological characteristics of folliculotropic MF.

Histopathologic patterns namely early follicular MF and advanced follicular MF were recently recognized. Early follicular MF is characterized by follicular papules, patches, plaques, acneiform lesions associated with a good prognosis. On the other hand, advanced follicular MF is characterized by infiltrated, follicular plaques or tumors associated with a poor prognosis[10] which is similar with our patient. Moreover, studies have shown that patients with denser and deeper infiltrates, syringotropism, as well as the presence of eosinophils and plasma cells typically exhibit an aggressive form of the disease.[10]

Immunohistochemical stainings CD3 and CD20 were utilized to determine whether it is of T or B cell lineage. Since it was positive for CD3, it indicated a T-cell origin. In folliculotropic MF, increased CD4:CD8 ratio is typically observed, which is consistent with our patient. Insignificant staining of CD8 helps rule out CD8-positive lymphoproliferative disorders. The loss of CD7, as seen in our patient, is often the first pan T cell marker to be lost in MF, and it is associated with a poorer prognosis. CD30 staining which showed insignificantly positive in our patient, was conducted to exclude the possibility of large cell transformation and indicates a worse prognosis. The CD45 stain, which is positive in our patient, is expected to be significant in hematolymphoid cells and pronounced staining in tumor cells is associated with a poorer prognosis.

Staging plays a crucial role in determining the appropriate treatment and prognosis of patients with mycosis fungoides. The International Society for Cutaneous Lymphomas (ISCL) and the European Organization of Research and Treatment of Cancer (EORTC) proposed a revised clinical staging system in 2007, based on the TNMB classification. Early-stage disease (IA-IIA) is characterized by patches and/or plaques, while advanced-stage disease (IIB-IV) involves tumors, erythroderma, or nodal, blood, and/or visceral involvement.[11] Based on the limited information in our patient, it can be inferred that the patient's minimum stage classification is stage III, as he presented with erythroderma at the time of diagnosis.

There are various treatment options available for folliculotropic MF, depending on the extent of the disease. These options range from skin-directed therapies such as topical corticosteroids, imiquimod, and reiquimod, as well as local and total electron beam therapy and phototherapy. Systemic therapies can also be utilized like retinoids, interferons, targeted monoclonal antibodies (e.g., alemtuzumab, brentuximab vedotin, mogamulizumab), and single-agent chemotherapy (methotrexate, pegylated liposomal doxorubicin, gemcitabine, and pentostatin).

Considering that the patient is classified as at least stage III, the National Comprehensive Cancer Network clinical practice guidelines (NCCN-CPG)[12] in Oncology for primary cutaneous lymphoma recommend systemic therapy in combination with skin-directed therapy. Combination therapy such as extracorporeal plasmapheresis plus interferon alpha and/or a retinoid may also be considered. Ten-year survival prognosis for patients with FMF differ according to the stage and presentation: 72% in skin-limited early stages, 28% in skin-limited advanced stages, and 2% in FMF with extracutaneous localizations at first presentation.[13]

CONCLUSION

We reported an exceptionally rare case of folliculotropic MF presenting with secondary cutis verticis gyrata in a Filipino male. To the best of our knowledge, this is only the second documented case worldwide with this presentation. Our case also represents the 4th documented case of cutis verticis gyrata in the Philippines, highlighting its rarity. Accurate diagnosis requires close clinical, histopathologic, and immunohistochemical correlation.

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SUPPLEMENTARY TABLE 1

Case	Age/Sex	CVG Type	Associated Condition(s)	Country
1	19 / M	Primary Essential	None	France – Alzaid et al. 2021
2	22 / F	Primary Essential	None	France – Alzaid et al. 2021
3	25 / M	Primary Essential	None	France – Alzaid et al. 2021
4	31 / M	Primary Essential	None	France – Alzaid et al. 2021
5	42 / M	Primary Essential	None	France – Alzaid et al. 2021
6	14 / F	Primary Essential	None	France – Alzaid et al. 2021
7	28/M	Primary Essential	None	Brazil – Yang et al 2014
8	25 / M	Primary Essential	None	Morocco – Mai et al 2019
9	53/ M	Primary Essential	None	Bosnia – Zeljko 2016
10	28 / M	Primary Essential	None	Tunisia – Chamli et al 2022
11	35/M	Primary Non Essential	Schizophrenia	India – Nachinolcar et al 2021
12	32 / M	Primary Non Essential	Epilepsy	Tunisia – Chamli et al 2022
13	15/M	Primary Non Essential	Epilepsy	Saudi – Aldawsari et al 2025
14	60 / F	Secondary	Infiltrating ductal carcinoma (breast)	Pakistan –Rahman et al. 2012
15	25 / M	Secondary	Cerebriform intradermal nevus (CIN)	China – Zeng & Guo 2021
16	46 / F	Secondary	CIN	USA – Fronek et al. 2021
17	28 / M	Secondary	Blue nevus, pigmented nodules	India – Sarkar et al. 2014
18	45 / M	Secondary	CIN	Mexico – Huerta et al. 2014
19	22 / M	Secondary	CIN	India – Ghosh et al. 2012
20	14 / M	Secondary	CIN	India – Tagore et al. 2011
21	17 / F	Secondary	CIN	China – Zhang et al. 2011
22	38 / F	Secondary	Combined blue and congenital nevus	Germany–Muhlhoff et al. 2010
23	43 / F	Secondary	Intradermal nevus	Brazil – Bonalumi et al. 2010
24	48 / M	Secondary	Intradermal nevus	Spain – Alcántara et al. 2010
25	16 / M	Secondary	Neurofibroma	France – Alzaid et al. 2021
26	34 / M	Secondary	Dissecting cellulitis	France – Alzaid et al. 2021
27	67 / F	Secondary	CIN	France – Alzaid et al. 2021
28	38 / M	Secondary	Lipoedema	France – Alzaid et al. 2021
29	29 / M	Secondary	Folliculitis fibrosis	France – Alzaid et al. 2021
30	44 / M	Secondary	Chronic folliculitis	France – Alzaid et al. 2021
31	51 / M	Secondary	Acromegaly	France – Alzaid et al. 2021
32	54 / F	Secondary	Endocrine pathology	Tunisia – Chamli et al 2022
33	18/M	Secondary	Congenital melanocytic nevi	Philippines – Sison et al 2017
34	0/M	Secondary	Probably Noonan Syndrome	Philippines – Cain 2018
35	28/M	Secondary	Primary pachydermoperiostosis	Philippines – Zamora et al 2022
36	28/M	Secondary	Psoriasis	USA – Le Nguyen et al 2016