Published in final edited form as:

J Am Acad Dermatol. 2019 June ; 80(6): 1704–1711. doi:10.1016/j.jaad.2019.01.062.

Follicular mucinosis in patients with hematologic malignancies other than mycosis fungoides: A clinicopathologic study.

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Abstract

Background: Follicular mucinosis (FM), defined by mucin accumulation within follicular epithelium, may occur in mycosis fungoides (MF). FM without MF is occasionally reported in systemic hematologic malignancies, and may be diagnostically challenging.

Objective: To describe clinicopathological characteristics of FM in patients with hematologic malignancies other than MF.

Methods: Clinical data and histopathology were analyzed in patients with FM and hematologic malignancies diagnosed between 1994 and 2017.

Results—Eighteen patients with FM and systemic hematologic malignancies without cutaneous T-cell lymphoma (CTCL) were identified, nine of them were post hematopoietic stem cell transplantation (HSCT). No non-CTCL associated FM cases (n=46, 37 biopsies) developed CTCL during mean follow-up of 4.3 years. Of CTCL associated with FM (n=44, 31 biopsies) MF was the most common subtype (n=38), although other CTCLs were identified. FM in patients with non-CTCL hematologic malignancies differed clinically from MF-associated FM, presenting most frequently with erythematous papules ($P < .0001$), without plaques ($P < .0001$), without alopecia ($P = .001$), and without histopathologically identified epidermal exocytosis ($P = .013$).

Limitations: A retrospective study in a single cancer center.

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Conflict of interest disclosures: The authors have no conflict of interest to declare.

IRB approval status: Reviewed and approved by MSKCC IRB; IRB #17–341

Conclusions: FM can present in systemic hematologic malignancies, including post-HSCT. Papular lesional morphology and histopathology may help to distinguish these cases from MF.

Keywords

cutaneous T-cell lymphoma; CTCL; mycosis fungoides; MF; follicular mucinosis; hematopoietic stem cell transplantation; CD30+ lymphoproliferative disorders; histopathology

Introduction

Follicular mucinosis (FM) is defined by the accumulation of mucin within follicular epithelium that can result in follicular damage. Clinically, FM presents as follicular prominence, acneiform eruption and focal hair loss. FM in association with various systemic conditions including hematological malignancies, hematopoietic stem cell transplantation (HSCT), and other malignant and benign skin disorders¹ has been occasionally reported. However, it is typically reported in the context of cutaneous T-cell lymphomas (CTCL), specifically folliculotropic mycosis fungoides (MF).^{2,3} It is a matter of controversy whether FM, especially when seen in a generalized eruption, may represent an early variant or a precursor stage of MF.^{3,4} FM may develop as a primary process that is considered idiopathic. Idiopathic FM is an uncommon benign disorder that is typically reported in children and young adults with locally distributed follicular-based papules, patches or plaques. MF-associated FM is more commonly described in older patients with a diffusely distributed eruption.^{3,5–8} MF-associated FM and idiopathic FM frequently look similar clinically, and their histopathologic features, immunohistochemical phenotype and T-cell receptor gene-rearrangement status overlap, thus diagnosis is based on a combination of clinical, histopathologic, immunohistochemical and molecular data.^{3,5,9–12}

In this study, we sought to examine our experience with FM in cancer center patients, specifically focusing on the clinicopathological features of FM in patients with non-MF hematologic malignancies.

Methods

Patients selection and clinical data collection

The study was approved by the Memorial Sloan Kettering Cancer Center (MSKCC) Institutional Review Board. A free text search of histopathology reports from the MSKCC Darwin patient database was performed for the following terms: ('skin' OR 'cutaneous') AND ('mucin' OR 'mucinosis' OR 'mucinoso' OR 'mucinosum'). All resulted cases that were diagnosed from January 1994 through July 2017 were evaluated by reviewing their pathology reports and medical records. Patients without a confirmed histopathological diagnosis of FM by a dermatopathologist at MSKCC or who were never seen by a physician at MSKCC were excluded. Clinical data including diagnoses, demographics, presentation and follow-up was collected. CTCL diagnoses were confirmed by clinical, histologic, immunohistochemical and molecular attributes as per the International Society for Cutaneous Lymphomas (ISCL) and the European Organization for Research and Treatment of Cancer (EORTC) guidelines.^{2,13,14} History of hematologic malignancy and HSCT prior

to FM detection was noted. We excluded three patients and three biopsies that showed an incidental finding of FM in a biopsy of a nevus / non-melanoma skin cancer with no correlation to an eruption or other clinical or histological findings. The included cases were grouped into a CTCL-associated FM group (MF-associated and other CTCL types subgroups) and a non-CTCL associated FM group (hematologic malignancy-associated and no hematologic malignancy subgroups). The MF-associated and the hematologic malignancy-associated FM cases were compared.

Histopathologic analysis

Available skin biopsy specimens showing FM were reviewed by a dermatopathologist (MP) who was blinded to the clinical diagnoses. Hematoxylin and eosin stained slides were evaluated for the following histologic features as previously performed in the literature^{9,10}: amount of mucin filling the hair follicles (graded between 1 to 4); presence of dermal mucin; presence of perifollicular lymphoid infiltrate, band-like architecture of lymphoid infiltrate, epidermal lymphocytic exocytosis; presence of lymphocytic atypia characterized by enlarged hyperchromatic nuclei with irregular contours; extent and location of eosinophils (graded as 0 none; + some; ++ abundant; localization in the hair follicle and/or the dermis).

Statistical analysis

Qualitative data were expressed as frequencies and percentages and quantitative data as means, standard deviations and ranges. Clinicopathologic features were compared between the two groups of patients with MF-associated FM and patients with FM and non-CTCL hematologic malignancy using two-sample t-test, and chi-square or Fisher's exact tests. Statistical analyses were performed using IBM SPSS Statistics software (version 24). All tests were 2-sided and P values less than .05 were considered statistically significant.

Results

A total of 90 patients with FM were included in the study: 46 patients (51%) without CTCL and 44 (49%) with CTCL. A total of 68 skin biopsies showing FM from 55 of the 90 patients (61%) were available for pathologic review. Demographics and initial clinical diagnosis for all patients are summarized in Table I. Non CTCL-associated FM patients were followed at our institution for periods ranging from 2 months to 21 years after FM was diagnosed (mean 51.7 months) and none developed CTCL during their follow-up.

18 patients (20%) had a diagnosis of non-CTCL hematologic malignancy at presentation. Clinical and histological features of FM in patients with non-CTCL hematologic malignancies are summarized in Table II. The majority of the patients in this group were males (14/18, 78%) and their mean age at diagnosis was 50.8 years. Compared to MF-associated FM, a higher percentage of non-white patients was noticed in the non-CTCL hematologic malignancy group (47% versus 16%) while no differences were found for gender or age between the groups. FM in patients with non-CTCL hematologic malignancies presented most frequently as erythematous papules in 83% (15/18) on the head and neck in 78% (14/18) (as seen in Fig 1). Less frequently it involved the trunk in 61% (11/18), presenting only occasionally as plaques, patches or macules and in none of the cases as

tumors/nodules. The clinical presentation differed significantly from patients with MF-associated FM, who presented with plaques in 76% (29/38, $P < .001$), papules in 13% (5/38, $P < .001$) and involvement of the lower extremities in 66% (25/38, $P = .023$). In addition, none of the patients with non-CTCL hematologic malignancy presented with alopecia compared to 40% of the MF-associated FM patients (15/38, $P = .001$).

Histopathologically, the incidence of epidermal exocytosis was lower in FM cases in patients with hematologic malignancies (2/16, 13%) compared to FM cases in MF patients (14/28, 50%, $P = .013$) while all other studied histopathology features didn't show significant differences between the groups (Table II).

Of the 18 patients with non-CTCL hematologic malignancy, nine patients had undergone HSCT prior to diagnosis of FM and nine had no history of HSCT (Table III). In eight of the patients with hematologic malignancy and no HSCT the malignancy was active at the time of FM diagnosis and one patient was in remission after he was treated with chemotherapy. In three patients, FM developed while being treated with chemotherapy. In two cases the FM eruption preceded the diagnosis of the hematologic malignancy: in the first, chronic diffuse pruritic erythematous patches were reported five years prior to the diagnosis of B-cell lymphoplasmacytic lymphoma and in the second patient, an erythematous pruritic eruption on the trunk and extremities coincided with the appearance of lymphadenopathy a few months before he was diagnosed with Hodgkin's lymphoma, nodular sclerosing subtype. In the nine patients who presented with FM and hematologic malignancy post-HSCT, FM onset was between less than a month and up to 7 years after HSCT.

28 patients with FM had no associated CTCL or hematologic malignancy and had variable presentations. Three patients had history of solid tumors (prostate cancer, gastrointestinal Stromal Tumor (GIST) and Kaposi's sarcoma) and two patients had HIV infection. All other patients were otherwise well and two of them have been reported by us elsewhere.⁸

Within the group of CTCL-associated FM, six cases had CTCL types other than MF (Table I). Clinicopathological correlation in a case with FM and primary cutaneous CD30+ lymphoproliferative disorders (LPD) is shown in Fig 2.

Discussion

A skin biopsy showing mucin accumulation within the follicular epithelium raises a concern for MF versus a less worrying diagnosis of idiopathic FM. These two entities and the challenge of distinguishing between them are well described in the literature. There is limited data on FM in the context of systemic and cutaneous disorders other than MF, and these conditions are poorly characterized. Mir-Bonafé *et al* reported on a series of thirteen cases with FM associated with nonlymphoid skin conditions, mainly inflammatory disorders, emphasizing that FM is a reactive histological pattern that can be seen unrelatedly to CTCL.¹ In our study of FM in a population of cancer center patients, FM was unrelated to CTCL in 51% of the cases and none of these 46 patients developed CTCL after a mean follow-up of 4.3 years. Our findings support the concept that FM should not be considered as a variant of MF or a precursor stage of CTCL as previously suggested.^{3,4}

20% of the FM cases in our cohort were seen in patients with history of a systemic hematologic malignancy and of those patients, half were post-HSCT. The association of FM and hematological malignancies, mostly HL^{15–17}, AML¹⁸, and CLL^{19,20} has been reported in small case-series and case reports and FM has been rarely described after HSCT^{21–23}. In our study FM occurred in association with various systemic hematological malignancies and HSCT. This group of 18 patients with non-CTCL hematologic malignancy were mostly males who generally presented with an eruption of papular morphology, only rarely with plaques, and without alopecia. Histopathologically, dense perifollicular infiltrates, epidermal exocytosis and bandlike infiltrate were uncommon. Patients with FM post-HSCT were predominantly non-white males and all presented with a papular eruption, mostly involving the head and neck and the trunk. Erythematous follicular-based papules were discrete (Fig 1A); were not clustered or linearly arranged; had no waxy firm appearance; and the surrounding skin was not shiny or indurated as frequently seen in papular mucinosis.²⁴ Furthermore, microscopically, mucin was deposited in the hair follicles (Fig 1B–C) and only rarely was seen in the dermis. Three similar cases of FM in male patients post-HSCT for hematologic malignancies have been previously reported in the literature: a 48-year-old post autologous HSCT for multiple myeloma²², a 19-year-old after allogeneic HSCT for acute lymphoblastic leukemia²¹ and a 17-year-old after allogeneic HSCT for AML²³. The first two patients presented with a papular eruption involving the face or the head, neck and upper trunk 3–5 weeks after transplant. The third case appeared with patches and follicular prominence on his trunk and extremities 9 months post-transplant. Interestingly, the two former patients were diagnosed with mild acute cutaneous GVHD diagnosed before presenting with FM, while in our cohort only one patient had history of chronic GVHD. Our findings and the previously reported cases suggest that FM is a reactive pattern that can be found in papular eruptions, can appear between a month and up to several years post transplantation regardless of the HSCT type, is transient in most cases and not associated with CTCL. Additional cases should be studied to evaluate the possible association of FM post-HSCT and GVHD.

The exact mechanism underlying the deposition of mucin selectively within the follicular unit is unknown. Follicular keratinocytes have been postulated to be the source of mucin and an etiologic role has been proposed for cell-mediated immune mechanisms and possible stimulation by cytokines released from perifollicular T lymphocytes.⁹ This might be supported by a possible association in our study between FM and alteration of cell-mediated immune mechanisms in patients post HSCT or underlying hematologic malignancy.

Recognition of the possible occurrence of non-CTCL FM in patients with hematologic malignancies and post-HSCT is important to spare these patients from unnecessary diagnostic and therapeutic strategies. History of lymphoma and other hematological disorders is occasionally reported in patients with MF. In a recent study, six of 203 patients with folliculotropic MF had history of hematologic malignancies²⁵ and in our study three of the 44 patients with MF-associated FM had history of hematologic neoplasms. Our findings suggest that certain clinical characteristic and histological features may support the diagnosis of non-CTCL FM in patients with hematologic malignancies.

FM is a common histological finding in folliculotropic MF and it has been reported in 78% of the cases included in a recent study.²⁵ FM can also be found in biopsies of cutaneous lymphomas other than MF, either CTCLs or cutaneous B-cell lymphomas^{26,27}. The finding of FM in primary cutaneous CD30+ LPD is unusual and 6 cases have been reported in the literature so far and were initially misdiagnosed as folliculotropic MF.²⁸ FM has been reported also in the setting of cutaneous lesions of adult T-cell leukemia lymphoma²⁹. In our study we report four additional cases of CD30+ LPD with FM as well as CD4+ small/medium CTCL and cutaneous peripheral T-cell lymphoma, unspecified, suggesting that the presence of FM in CTCL types other than MF may not be a rare condition as previously thought.

Limitations of our study include its retrospective design and being performed in a single cancer center.

In conclusions, FM can be seen in association with hematologic malignancies and HSCT and the clinical presentation and histological features in such cases may help to distinguish it from CTCL-associated FM.

Acknowledgments

Funding sources: This research was funded in part through the NIH/NCI Cancer Center Support Grant P30 CA008748.

Abbreviations

AML	acute myeloid leukemia
CLL	chronic lymphocytic leukemia
CTCL	cutaneous T-cell lymphoma
EORTC	European Organization for Research and Treatment of Cancer
FM	follicular mucinosis
GVHD	graft versus host disease
HL	Hodgkin's lymphoma
HSCT	hematopoietic stem cell transplantation
ISCL	International Society for Cutaneous Lymphomas
LPD	lymphoproliferative disorder
MF	mycosis fungoides
MSKCC	Memorial Sloan-Kettering Cancer Center

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Capsule Summary

- Follicular mucinosis in association with systemic hematologic disorders other than mycosis fungoides is poorly characterized.
- Clinicopathological features of follicular mucinosis in the setting of hematopoietic stem cell transplantation and diverse systemic hematologic malignancies differ from mycosis fungoides-associated cases and follicular mucinosis is not necessarily a precursor to cutaneous T-cell lymphoma.

Fig 1. Follicular mucinosis in a 50-year-old patient with history of mantle cell lymphoma post autologous hematopoietic stem cell transplantation. **A**, Clinical features. Erythematous papules on the face. **B and C**, Histopathology. Dermal atypical lymphohistiocytic infiltrate with follicular mucinosis. (Hematoxylin-eosin stain, original magnification, B, $\times 10$; C $\times 40$).

Fig 2.

Primary cutaneous CD30+ lymphoproliferative disorder with follicular mucinosis in a 71-year-old patient. **A**, Clinical features. Erythematous ulcerated papule on the neck. **B and C**, Histopathology. Dense dermal and subcutaneous atypical lymphoid infiltrate with eosinophils and follicular mucin. (Hematoxylin-eosin stain, original magnification, B, ×10; C ×40).

Table I.

Patients' demographics and clinical diagnoses at presentation

	CTCL-associated FM, N = 44 N (%) [*]	Non-CTCL-associated FM, N = 46 N (%) [*]
Age at diagnosis,		
years, mean $\pm$ SD,	55.0 $\pm$ 17.0	44.3 $\pm$ 16.4
(range)	(16–87)	(11–77)
Sex		
Male	26 (59)	26 (57)
Female	18 (41)	20 (43)
Race		
White	37 (84)	29 (63)
African American	3 (7)	4 (9)
Asian	0 (0)	4 (9)
Hispanic	3 (7)	6 (13)
N/A	1 (2)	3 (6)
Diagnosis		
CTCL	44 (100)	0 (0)
	mycosis fungoides- 38 (86)	
	primary cutaneous CD30+ lymphoproliferative Disorders - 4 (10)	
	primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorder - 1 (2)	
	peripheral T-cell lymphoma, unspecified - 1 (2)	
Hematological malignancies	3 (7)	18 (39)
	Burkitt lymphoma - 1	Hodgkin's lymphoma - 3
	chronic lymphocytic leukemia - 1	chronic lymphocytic leukemia - 3
	follicular lymphoma - 1	acute myeloid leukemia - 2
		mantle cell lymphoma - 2
		diffuse large B-cell lymphoma - 2
		myelodysplastic syndrome - 2
		multiple myeloma - 2
		chronic myeloid leukemia - 1
		lymphoplasmacytic lymphoma - 1
HSCT	0 (0)	9 (20)
		autologous HSCT: 5
		allogeneic T-cell depleted HSCT: 2
		double umbilical cord and haploidentical HSCT: 2
HIV	1 (2)	2 (4)

^{*} N (%) except where indicated.

Table II.

Comparison of clinical parameters and histopathologic features among patients with non-CTCL hematologic malignancy associated follicular mucinosis and MF-associated follicular mucinosis

Clinical Parameter	MF-associated FM N = 38 patients, N (%)*	Hematologic malignancy-associated FM N = 18 patients, N (%)*	P value
Age at diagnosis , years, mean +/- SD	54.0 +/- 16.6	50.8 +/- 14.9	.494
Sex			
Male	21 (55)	14 (78)	.104
Female	17 (45)	4 (22)	
Race			
White	31 (82)	9 (50)	.022
African American	3 (8)	1 (6)	
Asian	0 (0)	4 (22)	
Hispanic	3 (8)	3 (17)	
N/A	1 (2)	1 (5)	
Type of lesions⁺			
Papules	5 (13)	15 (83)	<.0001
Plaques	29 (76)	3 (17)	<.0001
Macules	0 (0)	2 (11)	.099
Patches	23 (61)	6 (33)	.057
Nodules / Tumors	6 (16)	0 (0)	.162
Follicular prominence	6 (16)	5 (28)	.305
Alopecia	15 (40)	0 (0)	.001
Distribution⁺			
Head and Neck	31 (82)	14 (78)	.732
Trunk	29 (76)	11 (61)	.239
Upper Extremities	16 (42)	7 (39)	.819
Lower Extremities	25 (66)	6 (33)	.023
Unilesional	6 (16)	1 (6)	.409
Pruritus	26 (68)	11 (61)	.763
Histologic Features	N = 28 biopsies, N (%)	N = 16 biopsies, N (%)	
Follicular mucin			
Grade 1	6 (21)	2 (13)	.732
Grade 2	13 (47)	7 (44)	
Grade 3	5 (18)	5 (31)	
Grade 4	4 (14)	2 (13)	
Dermal mucin	1 (4)	1 (6)	>.999
Lymphoid infiltrate			
Perifollicular infiltrate			.141
None	0 (0)	1 (6)	
Some	21 (75)	14 (88)	
Dense	7 (25)	1 (6)	

	MF-associated FM	Hematologic malignancy-associated FM	P value
Epidermal exocytosis	14 (50)	2 (13)	.013
Bandlike infiltrate	12 (43)	4 (25)	.236
Lymphoid cytologic atypia	22 (79)	11 (69)	.492
Eosinophils			
None	8 (29)	2 (13)	.457
Some	12 (42)	9 (56)	
Abundant	8 (29)	5 (31)	
In dermis	14 (70)	11 (79)	.704
In dermis & hair follicle	6 (30)	3 (21)	

Table III

Clinical and histological features of FM in non-CTCL hematologic malignancy patients post-HSCT and with no HSCT

	FM in hematologic malignancy patients post-HSCT	FM in hematologic malignancy patients and no HSCT
<u>Clinical features</u>	N = 9 patients, n/N (%)*	N = 9 patients, n/N (%)*
Age at diagnosis , years, mean +/- SD	48.3 +/- 16.3	53.3 +/- 4.6
Sex (male : female)	7:2 (78:22)	7:2 (78:22)
Race		
White	3/9 (33)	6/9 (67)
African American	1/9 (11)	0/9 (0)
Asian	4/9 (44)	0/9 (0)
Hispanic	1/9 (11)	2/9 (22)
N/A	0/9 (0)	1/9 (11)
Type of lesions		
Papules	9/9 (100)	6/9 (67)
Plaques	0/9 (0)	3/9 (33)
Macules	1/9 (11)	1/9 (11)
Patches	1/9 (11)	5/9 (56)
Nodules / Tumors	0/9 (0)	0/9 (0)
Follicular prominence	4/9 (44)	1/9 (11)
Alopecia	0/9 (0)	0/9 (0)
Distribution⁺		
Head and Neck	8/9 (89)	6/9 (67)
Trunk	6/9 (67)	5/9 (56)
Upper Extremities	5/9 (56)	2/9 (22)
Lower Extremities	3/9 (33)	3/9 (33)
Unilesional	0/9 (0)	1/9 (11)
Pruritus	5/9 (56)	6/9 (67)
<u>Histologic features</u>	N = 7 biopsies, n/N (%)*	N = 9 biopsies, n/N (%)*
Follicular mucin		
Grade 1	1/7 (14)	1/9 (11)
Grade 2	2/7 (29)	5/9 (56)
Grade 3	4/7 (57)	1/9 (11)
Grade 4	0/7 (0)	2/9 (22)
Dermal mucin	1/7 (14)	0/9 (0)
Lymphoid infiltrate		
Perifollicular infiltrate	7/7 (100)	8/9 (89)
Epidermal exocytosis	0/7 (0)	2/9 (22)
Bandlike infiltrate	2/7 (29)	2/9 (22)
Lymphoid cytologic atypia	6/7 (86)	5/9 (56)
Eosinophils		

	FM in hematologic malignancy patients post-HSCT	FM in hematologic malignancy patients and no HSCT
None	1/7 (14)	1/9 (11)
Some	2/7 (29)	7/9 (77)
Abundant	4/7 (57)	1/9 (11)

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