# A case of nevoid follicular mucinosis in a child

David M. Weiner, AB,[a] Ashley K. Clark, MD,[b] Amanda T. Moon, MD,[c] Paul L. Haun, MD,[b] and Sara S. Samimi, MD[b]
*Philadelphia, Pennsylvania*

***Key words:*** CTCL; cutaneous mucinoses; epidermal nevus; mycosis fungoides; nevoid follicular mucinosis.

## INTRODUCTION

Nevoid follicular mucinosis is a new diagnosis initially described by Tadini et al[1] in 2013 to represent a lesion with the Blaschkoid appearance of an epidermal nevus that exhibits follicular mucinosis (FM), or a pattern of mucin deposition within hair follicles, on histopathology. Such a presentation is quite rare, and to date, no other cases of nevoid FM have been reported. We describe herein a second case of a child with nevoid FM.

*Abbreviation used:*

FM: follicular mucinosis

## CASE REPORT

A 10-year-old boy presented with a lesion on his left cheek that first appeared around age 1. Clinical examination showed a linear and whorled plaque on the left cheek that extended to the nose and upper lip with small monomorphic folliculocentric papules, some with central keratotic plugging (Fig 1). His mother reported that the lesion had been growing slowly and proportionally with the patient over time, with waxing and waning of follicular prominence, plugging, and associated inflammation. Since age 8, however, the follicular changes had become more prominent, and the lesion had become more pruritic. The patient had otherwise been developing normally.

Prior treatment with topical steroids and UV-B light therapy had produced partial improvement, but the lesion worsened with cessation of each treatment. The lesion was biopsied at ages 1, 4, and 9 and repeatedly demonstrated multifocal FM with a focally elevated CD4:CD8 ratio (Fig 2). The biopsies also demonstrated an associated perifollicular lymphocytic infiltrate with newly noted lymphocyte atypia on the most recent biopsy. T-cell receptor gene rearrangement testing was performed on the last biopsy and was negative. Given the Blaschkoid distribution, lack of other cutaneous findings, early age of onset, and FM on histopathology, the patient was diagnosed with nevoid FM. Treatment with hydroxychloroquine and tazarotene gel initially produced some clinical improvement. However, the pruritus and accentuated follicular plugging recurred. The patient is currently being treated with isotretinoin 10 mg daily and ultraviolet B light therapy. His nevoid FM has since been stable, with some slight thickening on the left upper lip.

## DISCUSSION

This lesion had the clinical appearance of a congenital epidermal nevus as it began early in the patient's life and was distributed along Blaschko's lines. Epidermal nevi are hamartomatous proliferations of the epidermis that usually develop *in utero* or during early childhood. They tend to follow a Blaschkoid distribution along the lines of embryonic cell migration that occur in skin development.[2] Epidermal nevi containing mucin are usually categorized as either mucinous nevi, with mucin throughout the papillary dermis, or mucinous eccrine nevi, with mucin around proliferating eccrine structures.[3] However, this lesion contained mucin exclusively within hair follicles.

FM is a pathologic description of this follicular pattern of mucin deposition, originally named

From the Perelman School of Medicine[a] and the Department of Dermatology[b], University of Pennsylvania; Division of Pediatrics, Section of Dermatology, Children's Hospital of Philadelphia.[c]

Funding sources: None.
IRB approval status: Not applicable.

Correspondence to: Sara S. Samimi, MD, Department of Dermatology, University of Pennsylvania, 3400 Civic Center Blvd, Philadelphia, PA 19104. E-mail: sara.samimi@pennmedicine.upenn.edu.

JAAD Case Reports 2021;18:29-32.
2352-5126



https://doi.org/10.1016/j.jdcr.2021.10.011

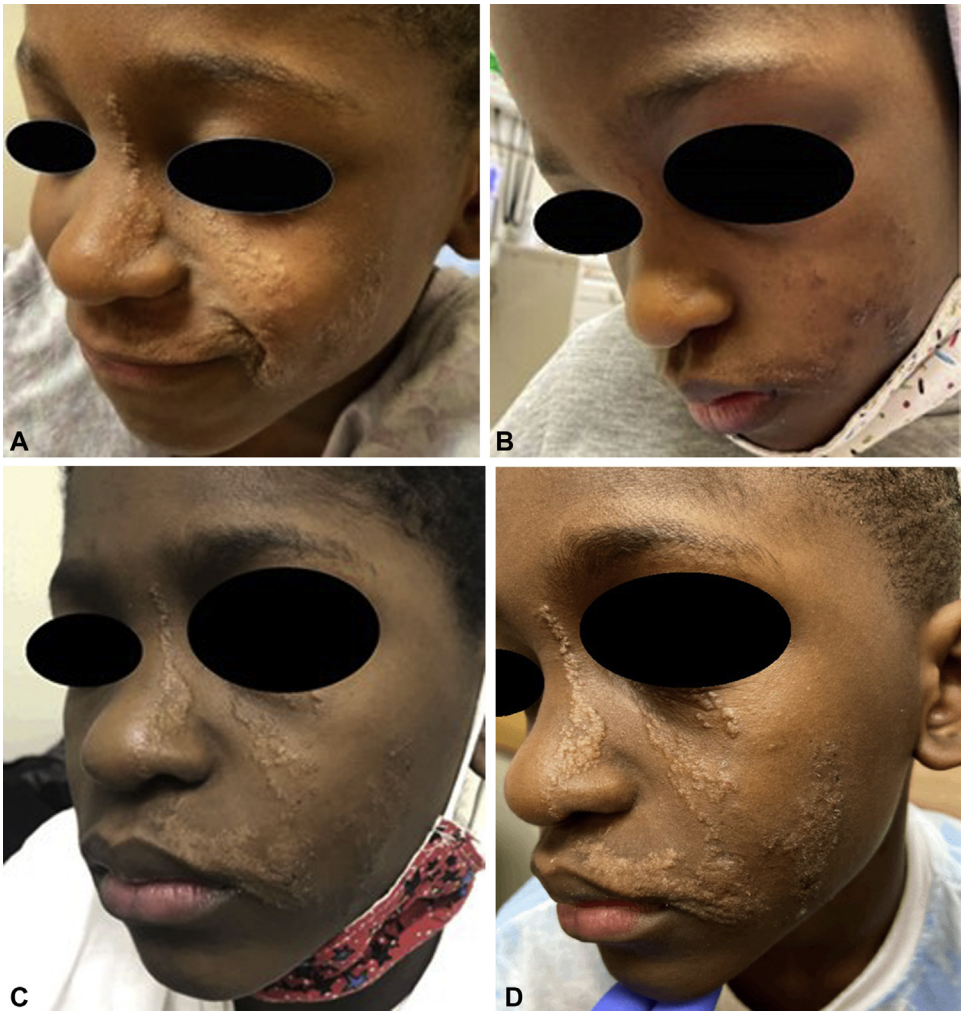


**Fig 1.** Clinical images of nevoid follicular mucinosis (FM). **A**, Lesion on presentation: Linear and whorled plaque on left cheek with extension onto left lateral nose and left upper lip with somewhat edematous small monomorphic papules, some with central keratotic plugging. **B**, Initial improvement after hydroxychloroquine and tazarotene gel: Reduced induration and follicular plugging. **C**, Subsequent progression: Recurrence of accentuated follicular plugging. **D**, Stable nevoid FM: With slight thickening on left upper lip.

alopecia mucinosis by Pinkus[4] in 1957 and later termed follicular mucinosis by Jablonska[5] in 1959. The etiology of this reaction pattern is somewhat unclear, but one hypothesized trigger of follicular mucin production is stimulation of follicular keratinocytes by cytokines from adjacent T lymphocytes.[6] FM usually presents as erythematous plaques, sometimes with follicular prominence and localized alopecia.[7]

The Blaschkoid appearance of this lesion did not align with the traditional clinical features of FM. However, the new entity termed nevoid follicular mucinosis encompasses such cases of nevoid skin lesions distributed along Blaschko's lines with FM on histopathology.[1] This condition was first described by Tadini et al[1] in 2013 in a 6-year-old girl with an early-onset linear skin lesion along her face, trunk, and arm. She was initially diagnosed with an epidermal nevus but subsequently demonstrated FM on biopsy without T-cell clonality. She was ultimately diagnosed with nevoid follicular FM to denote the combination of the lesion's nevoid appearance and FM pattern on histopathology.

FM is usually classified as either primary idiopathic, a benign form, or secondary FM, which can occur together with various other conditions including cutaneous lymphoma, hematologic malignancy, inflammatory disorders, and infections.[7] The

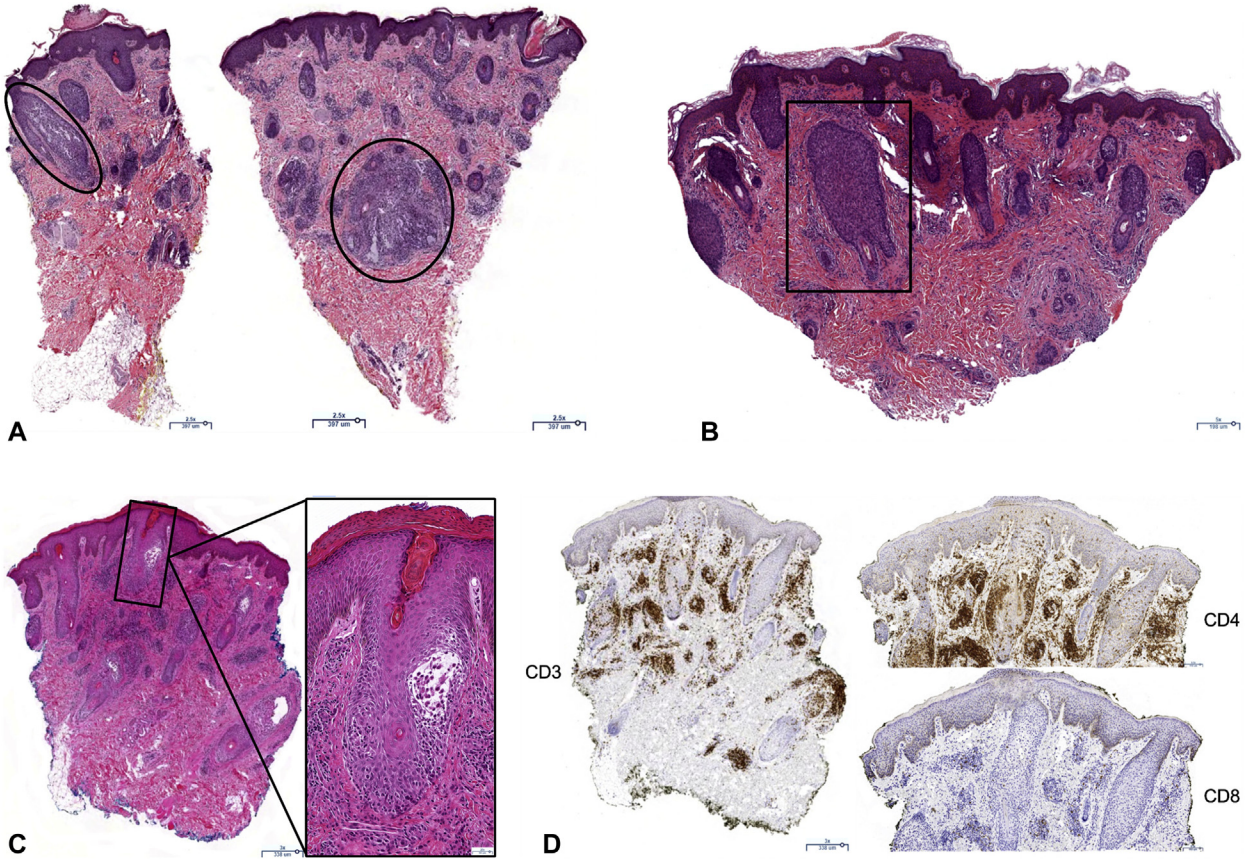


**Fig 2.** Histopathologic images of nevoid follicular mucinosis (FM) at ages 1, 4, and 9. **A**, Findings at age 1 (original magnification: ×2.5): Multifocal FM (*black circles*) with perifollicular and perivascular lymphocytic inflammation. Epidermis with mild acanthosis, spongiosis, and focal hyperkeratosis with serum scale. **B**, Findings at age 4 (original magnification: ×5): Stable multifocal FM (black square) with perifollicular and perivascular lymphocytic inflammation. **C**, Findings at age 9 (original magnification: ×3): Stable FM (*black square*) with follicular lymphocytic inflammation showing some atypical cytologic features (see box on the right for magnified view; original magnification: ×20). Overlying epidermis with spongiosis and parakeratosis. **D**, Findings at age 9, immunohistochemistry (original magnification: ×3): Left: CD3 highlights lymphocytes within follicular epithelium but not the overlying epithelium. Right: The intrafollicular T cells show an elevated CD4:CD8 ratio.

majority of FM cases are associated with lymphoma, most commonly cutaneous T-cell lymphoma.[7] In particular, folliculotropic mycosis fungoides may present with FM as well as an atypical lymphocytic infiltrate of hair follicles, and it often occurs in the head and neck area.[8] Before making a diagnosis of nevoid FM in this patient, it was therefore important to investigate the possibility of mycosis fungoides-associated FM. The clinical appearance and progression of the lesion as well as the results of T-cell receptor gene rearrangement testing and immunohistochemistry were carefully considered.

In this case, the CD4:CD8 ratio was elevated, and atypical lymphocytes were present. However, T-cell receptor gene rearrangement/polymerase chain reaction testing was negative. A diagnosis of nevoid FM was made based on the notable Blaschkoid distribution, the stable FM on histopathology with negative gene rearrangement test, the early age of onset, and the benign long-term clinical appearance without development of additional cutaneous lesions elsewhere.

This is the second reported case of nevoid FM. Notably, several biopsies spanning 9 years of clinical follow-up were available, allowing for histopathologic observation of this rare lesion over time. In this case, the biopsies consistently demonstrated FM with an associated inflammatory infiltrate and recent

cytologic atypia. We will continue to monitor this patient to evaluate for any further clinical or histopathologic evolution of the lesion.

**Conflicts of interest**

None disclosed.

**REFERENCES**

1. Tadini G, Boldrini MP, Brena M, Pezzani L, Marchesi L, Rongioletti F. Nevoid follicular mucinosis: a new type of hair follicle nevus. *J Cutan Pathol*. 2013;40(9):844-847.
2. Brandling-Bennett HA, Morel KD. Epidermal nevi. *Pediatr Clin North Am*. 2010;57(5):1177-1198.
3. Rongioletti F. Primary paediatric cutaneous mucinoses. *Br J Dermatol*. 2020;182(1):29-38.
4. Pinkus H. Alopecia mucinosa; inflammatory plaques with alopecia characterized by root-sheath mucinosis. *AMA Arch Derm*. 1957;76(4):419-424. discussion 424—426.
5. Jablonska S, Chorzelski T, Lancucki J. Mucinosis follicularis. Article in German. *Hautarzt*. 1959;10:27-33.
6. Rongioletti F, De Lucchi S, Meyes D, et al. Follicular mucinosis: a clinicopathologic, histochemical, immunohistochemical and molecular study comparing the primary benign form and the mycosis fungoides-associated follicular mucinosis. *J Cutan Pathol*. 2010;37(1):15-19.
7. Khalil J, Kurban M, Abbas O. Follicular mucinosis: a review. *Int J Dermatol*. 2021;60(2):159-165.
8. Emge DA, Lewis DJ, Aung PP, Duvic M. How to discern folliculotropic mycosis fungoides from follicular mucinosis using a pediatric case. *J Cutan Med Surg*. 2018;22(3):336-340.