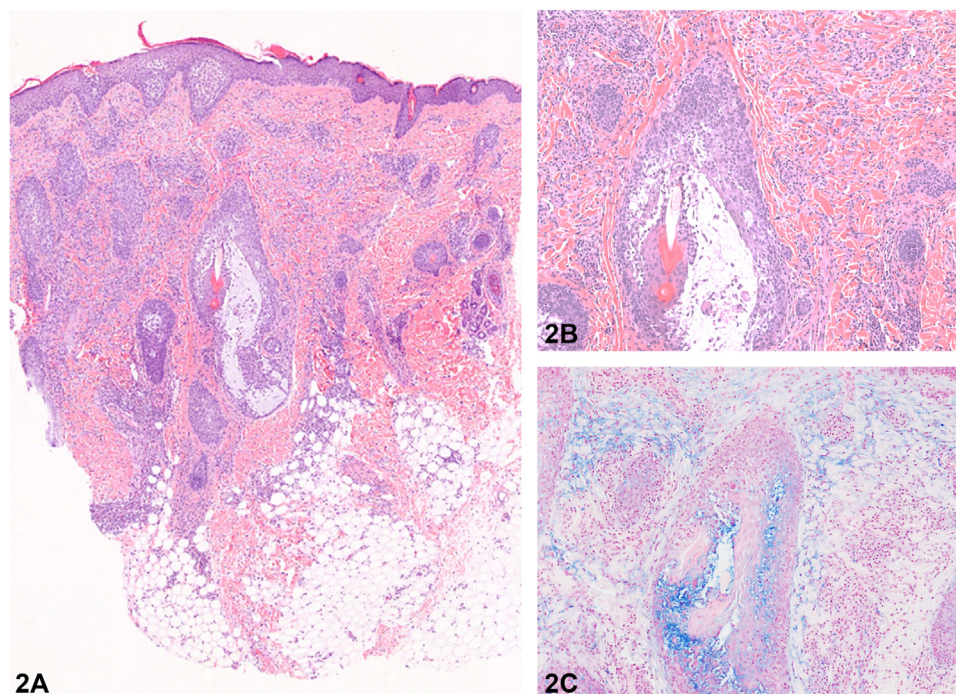
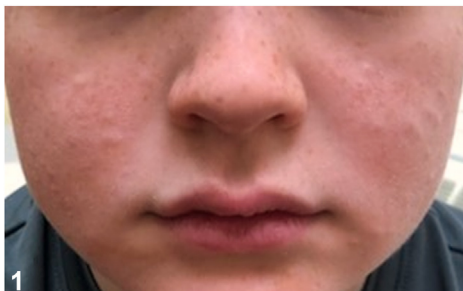


## Multiple skin-colored facial papules in a child



Michael L. MacGillivray, MD,<sup>a</sup> Ashley E. Sutherland, MD,<sup>a</sup> and Noreen M. Walsh, MD<sup>b</sup>

**Key words:** follicular mucinosis; multiple skin-colored facial papules; mycosis fungoides; pediatric.



An otherwise-well 9-year-old White male with a family history of acne presented with a 2-year history of firm dermal papulonodules on the anterior and lateral cheeks with no inflammation and no overlying epidermal change apart from scattered, coincidental ephelides (Fig 1). The forehead, back, and chest were spared. The

From the Division of Dermatology, Dalhousie University, Halifax, Nova Scotia, Canada<sup>a</sup>; Department of Pathology, Dalhousie University, Halifax, Nova Scotia, Canada.<sup>b</sup>

Funding sources: None.

IRB approval status: Not applicable.

Patient consent: We assure patient informed written consent via signature of their guardian was obtained.

Correspondence to: Michael L. MacGillivray, MD, Division of Dermatology, Dalhousie University, Suite 4-193 Dickson

Building, 5820 University Ave, Halifax, Nova Scotia, B3H 2Y9, Canada. E-mail: [michael.macgillivray@dal.ca](mailto:michael.macgillivray@dal.ca).

JAAD Case Reports 2022;27:85-7.

2352-5126

© 2022 by the American Academy of Dermatology, Inc. Published by Elsevier, Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

<https://doi.org/10.1016/j.jcdr.2022.05.046>

patient had no fever, weight loss, night sweats, and lymphadenopathy. A topical antibiotic, antifungal, and retinoid were trialed without effect. Laboratory tests were unremarkable. A 4-mm punch biopsy was performed, and representative sections are shown (Fig 2, A-C).

**Question 1: What is the most likely diagnosis?**

- A. Self-healing cutaneous mucinosis
- B. Fibrofolliculoma
- C. Syringomas
- D. Follicular mucinosis
- E. Angiofibromas

**Answer:**

**A.** Self-healing cutaneous mucinosis—Incorrect. Acute eruption of multiple papules, sometimes coalescing into linear infiltrated plaques on the head and neck, abdomen, and thighs. Often accompanied by systemic symptoms like fever, arthralgias, and myalgias. Spontaneous resolution over a period of 1-8 months is characteristic. Histologically, papular lesions show dermal mucin deposition.

**B.** Fibrofolliculoma—Incorrect. Presents as multiple small, skin-colored to hypopigmented papules involving the head, neck, and upper trunk. Histopathology displays slender strands of follicular mantle cells that emanate from a folliculosebaceous unit at the level of the isthmus. Occasionally, the stromal component of fibrofolliculomas can be rich in mucin, but this is distinct from the intraepithelial distribution of mucin in follicular mucinosis. In the setting of a fibrofolliculoma, strong consideration should be given to the possibility of Birt-Hogg-Dubé syndrome.

**C.** Syringomas—Incorrect. Clinically manifests as multiple small, firm, skin-toned papules at any site of the body but are prone to occur in the periorbital area, especially the eyelids. Histopathology demonstrates a proliferation of epithelioid cells with pale or pinkish cytoplasm forming nests and tubules of relatively uniform size.

**D.** Follicular mucinosis (FM)—Correct. Histopathology shows abundant, focally coalescent mucin within the follicular epithelium along with a surrounding perivascular and interstitial mixed inflammatory infiltrate composed of lymphomononuclear cells and abundant eosinophils.<sup>1</sup>

**E.** Angiofibromas—Incorrect. Usually appear as a small skin-toned to red papule. If multiple, should consider an underlying disorder such as tuberous sclerosis, particularly if they present in childhood on the bilateral cheeks, nasolabial folds, nose, and chin.

Histopathology consists of a dermal proliferation of plump or stellate fibroblasts in a collagenous stroma with an increase in the number of thin-walled, dilated blood vessels.

**Question 2: Which histopathologic stain would detect the mucin deposition present in follicular mucinosis (Fig 2, C)?**

- A. Masson trichrome
- B. Alcian blue (pH = 2.5)
- C. PAS-D
- D. Giemsa
- E. Prussian blue

**Answer:**

**A.** Masson trichrome—Incorrect. Stains collagen blue-green and smooth muscle red.

**B.** Alcian blue (pH = 2.5)—Correct. Stains acid mucopolysaccharides blue.

**C.** PAS-D—Incorrect. Stains neutral mucopolysaccharides red.

**D.** Giemsa—Incorrect. Has many uses, including highlighting myeloid and mast cell granules purplish-blue.

**E.** Prussian blue—Incorrect. Stains ferric ions to form a deep blue color. Useful to distinguish hemosiderin from melanin.

**Question 3: Which cutaneous neoplasm has been most commonly associated with FM?**

- A. Squamous cell carcinoma
- B. Tricholemmal carcinoma
- C. Mycosis fungoides (MF)
- D. Basal cell carcinoma
- E. Primary cutaneous follicle center lymphoma

**Answer:**

**A.** Squamous cell carcinoma—Incorrect. Can be rarely associated with histologic deposition of mucin, termed secondary mucinosis. Not associated with FM.

**B.** Tricholemmal carcinoma—Incorrect. Rare malignant counterpart of trichilemmoma with no associated

FM. Histopathology shows infiltrative aggregations of pale keratinocytes with outer root sheath differentiation, cytologic atypia, and brisk mitotic activity.

**C.** MF—Correct. Two main clinicopathologic variants of FM have been proposed: a primary benign idiopathic form that occurs mostly in children and young adults, with a tendency to resolve spontaneously within 2 months to 2 years, and a secondary form occurring in older patients and associated with cutaneous lymphoma, mainly MF and Sézary syndrome.<sup>2</sup> Incel Uysal et al<sup>3</sup> and references therein demonstrate the rare occurrence of MF in children with FM. The differentiation between primary FM and MF-associated FM is difficult, and there is no single reliable distinguishing criterion.<sup>1</sup> Detection of clonal T-cell gene rearrangements can be seen in both settings. Ultimately the clinical evolution of the disease is most informative.

**D.** Basal cell carcinoma—Incorrect. Secondary mucinosis commonly observed but does not involve hair follicles.

**E.** Primary cutaneous follicle center lymphoma—Incorrect. FM has occurred concomitantly with primary cutaneous follicle center lymphoma in only 1 case report of a man in his 60s.<sup>4</sup> Geller et al<sup>5</sup> and references therein note other non-cutaneous T-cell lymphomas, many not primary to the skin,

that may have FM as an associated feature, but their co-occurrence is rarer than FM with MF.

#### Abbreviations used:

FM: follicular mucinosis  
MF: mycosis fungoides

#### Conflict of interest

None disclosed.

#### REFERENCES

1. Rongioletti F, Rebora A. Cutaneous mucinoses: microscopic criteria for diagnosis. *Am J Dermatopathol*. 2001;23(3):257-267. <https://doi.org/10.1097/00000372-200106000-00022>
2. 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. <https://doi.org/10.1111/j.1600-0560.2009.01338.x>
3. Incel Uysal P, Bozdogan O, Atilan A, Yalcin B. Juvenile-onset early-stage mycosis fungoides—associated follicular mucinosis: a case report. *Am J Dermatopathol*. 2018;40(8):e112-e114. <https://doi.org/10.1097/DAD.0000000000001114>
4. Garrido MC, Riveiro-Falkenbach E, Rodriguez-Peralto JL. Primary cutaneous follicle center lymphoma with follicular mucinosis. *JAMA Dermatol*. 2014;150(8):906-907. <https://doi.org/10.1001/jamadermatol.2013.9060>
5. Geller S, Gomez CJ, Myskowski PL, Pulitzer M. Follicular mucinosis in patients with hematologic malignancies other than mycosis fungoides: a clinicopathologic study. *J Am Acad Dermatol*. 2019;80(6):1704-1711. <https://doi.org/10.1016/j.jaad.2019.01.062>