

Cutaneous mucinosis of infancy



Swapna C. Reddy, MD,^a Jayson R. Baman, BS,^b Clinton S. Morrison, MD,^c and Glynis A. Scott, MD^d
Rochester, New York

Key words: Alcian blue; cutaneous mucinosis of infancy; cobblestoned plaque; mucin.

We present a case of a 4-year-old fair-skinned girl, with no significant medical history, who presented to the outpatient clinic of an academic medical center with an asymptomatic, flesh-colored, cobblestoned plaque on her lateral left thigh (Fig 1). Per the family's report, the lesion had grown significantly since it was first noticed 2 years ago, with current measurements of 7.5 cm by 6.5 cm at the time of clinical presentation. The patient denied pain, tenderness, or pruritus at the site, and there was no history of drainage or infection. Further interrogation found no family history of cutaneous malignancies and no history of trauma to the area. The clinical differential diagnosis included lymphangioma, granuloma annulare, and scleromyxedema.

An incisional biopsy of the lesion was performed after obtaining informed consent from the patient's mother. Sections showed increased mucin, highlighted by alcian blue stains, confined to the dermis. The subcutis was normal (Fig 2). There was no evidence of abnormal lymphatic pattern or interstitial histiocytes. A diagnosis of cutaneous mucinosis of infancy (CMI) was made. The patient and her family were assured of the benign nature of CMI. She will be monitored for any acute changes to the lesion, but she is not being treated with any pharmacologic intervention or further surgical action at this point.

DISCUSSION

CMI was recognized as a distinct clinical entity in 1980.¹ Since then, only 8 cases have been reported.²⁻⁵ The histologic appearance of CMI shares features with other cutaneous mucinoses, including localized lichen myxedematosus, papular lichen myxedematosus, acral

Abbreviations used:

CMI: cutaneous mucinosis of infancy
SHJCM: self-healing juvenile cutaneous mucinosis



Fig 1. Flesh-colored cobblestone plaque seen on left thigh. Healing biopsy site is visible near the center of the lesion.

persistent papular mucinosis, and self-healing juvenile cutaneous mucinosis (SHJCM). Unifying characteristics of these conditions include the absence of thyroid disease and the absence of rheumatologic disease.^{4,6} Given the rarity of CMI, there are currently insufficient data to establish if CMI is in its own class of disease or if it is simply a member of the pediatric papular mucinosis family. The reported cases describe age

From the Department of Dermatology, University of Rochester Medical Center/Strong Memorial Hospital^a; the University of Rochester School of Medicine & Dentistry^b; and the Departments of Plastic Surgery^c and Dermatology and Pathology,^d University of Rochester Medical Center.

Funding sources: None.

Conflicts of interest: None declared.

Correspondence to: Swapna C. Reddy, MD, 601 Elmwood Ave, Box 697, Rochester, NY 14642. E-mail: Swapna_Reddy@urmc.rochester.edu.

JAAD Case Reports 2016;2:250-2.

2352-5126

© 2016 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/>).

<http://dx.doi.org/10.1016/j.jidcr.2016.04.005>

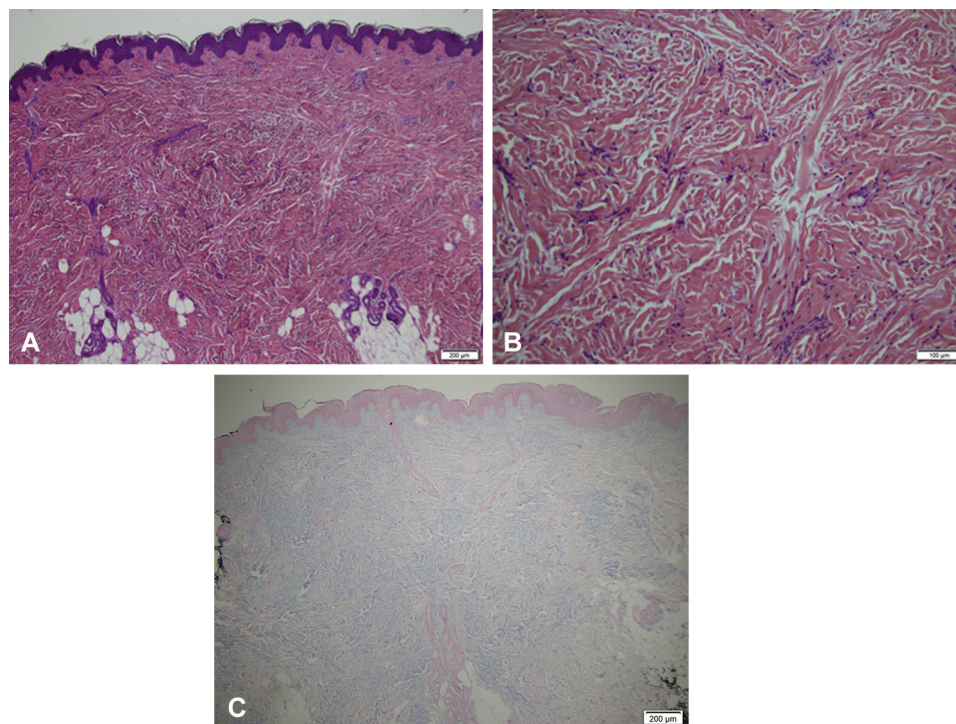


Fig 2. Sections show a subtle thickening of the reticular dermis with widening of the interstitial spaces, seen in low power (**A**) and high power (**B**). **C**, Alcian blue stains highlight the increased mucin.

and superficial mucinous deposition as common traits in the disease, but there are hardly robust data to establish diagnostic criteria.²

The clinical presentation of CMI and SHJCM are distinctly different, which aids in discriminating between these 2 mucinoses of infancy and childhood. Unlike SHJCM, mucin in CMI is confined to the dermis, and there is no accompanying fibroblastic proliferation.

CMI presents as a plaque or localized grouping of papules in young children. The most common areas for these localized lesions are the upper extremities and trunk.⁴ Lesions on the thigh, as seen in our patient, were similarly noted in a recent report.³ Nearly all cases are idiopathic, although the case of a child whose father had similar cutaneous lesions during his childhood may suggest a genetic or multifactorial element.² Even though our patient's lesions were first noticed at 2 years of age, other lesions associated with CMI have presented at birth.⁵

Mucin, which is produced by fibroblasts and normally accumulates in small quantities as part of the skin's extracellular matrix, is important in regulating fluid balance and contributing to the extracellular scaffold. For unclear reasons, patients with the aforementioned mucinoses experience an abnormal deposition of mucin, which results in clinically appreciated cutaneous lesions.

The abnormal deposition of mucin may be the primary etiology of a cutaneous mucinosis, as in the case of CMI. In addition, abnormal extracellular mucin accumulation can be the result of a secondary disease process in the context of a systemic condition. This finding is perhaps most widely recognized in the findings of exophthalmos and pretibial myxedema in the setting of Graves' disease hyperthyroidism. In this scenario, autoantibodies induce fibroblasts to produce and deposit glycosaminoglycans, a variant of mucin.⁷ Fibroblast overstimulation, hypersensitivity reactions, viral infections, and neoplastic changes are among the proposed mechanisms of cutaneous mucinoses.^{8,9} A working model of the pathophysiologic mechanism of CMI has not been proposed.

CMI has been classically considered a persistent, stable cutaneous condition, although reports of dynamic changes in its lesions, including the expanding nature of our own patient's lesion, highlight our evolving understanding of this condition.^{3,4}

Given their similar cutaneous findings and pathology, CMI and SHJCM are considered related disorders. Although mucin accumulation in the dermis is a recognized phenomenon in both cases, the clinical course differs between these diseases. The cutaneous lesions in CMI are known to persist for many years. In addition, CMI has not been shown to be associated

with systemic symptoms. The lesions in SHJCM, however, tend to present in older patients (most cases present at age 8 years or older), are self-resolving, and classically occur on the face or hands. In addition, the cutaneous findings in SHJCM have been associated with systemic symptoms such as arthralgias, myositis, and lymphocytosis.^{10,11} Histologically, SHJCM has a fibroblastic reactive proliferation involving the dermis and subcutis, with mucin deposited throughout the dermis and subcutis.

This patient's presentation is particularly interesting in that many of the lesion's characteristics are unusual for this disease, including the expanding nature of the lesion and its location on the lower extremity. We describe this patient's case to highlight the importance of recognizing this initially concerning, yet ultimately benign, cutaneous lesion.

REFERENCES

1. Lum D. Cutaneous mucinosis of infancy. *Arch Dermatol*. 1980; 116:198-200.
2. Podda M, Rongioletti F, Greiner D, et al. Cutaneous mucinosis of infancy: is it a real entity or the paediatric form of lichen myxoedematosus (papular mucinosis)? *Br J Dermatol* 2001; 144:590-593.
3. Mir-Bonafe JM, Vicente-Villa A, Sunol M, Soria X, Gonzalez-Ensenat MA. Cutaneous Mucinosis of Infancy: A Rare Congenital Case with Coexisting Progressive, Eruptive, and Spontaneously Involuting Lesions. *Pediatr Dermatol*. 2015; 32(6):e255-e258.
4. Chen CW, Tsai TF, Chang SP, Chen YF, Hung CM. Congenital cutaneous mucinosis with spontaneous regression: an atypical cutaneous mucinosis of infancy? *Clin Exp Dermatol* 2009;34: 804-807.
5. McGrae JD Jr. Cutaneous mucinosis of infancy. A congenital and linear variant. *Arch Dermatol*. 1983;119:272-273.
6. Rongioletti F, Rebora A. Updated classification of papular mucinosis, lichen myxoedematosus, and scleromyxedema. *J Am Acad Dermatol*. 2001;44:273-281.
7. Bahn RS. Graves' ophthalmopathy. *N Engl J Med*. 2010;362: 726-738.
8. Pandya AG, Sontheimer RD, Cockerell CJ, Takashima A, Piepkorn M. Papulonodular mucinosis associated with systemic lupus erythematosus: possible mechanisms of increased glycosaminoglycan accumulation. *J Am Acad Dermatol*. 1995; 32:199-205.
9. Wriston CC, Rubin AI, Martin LK, Kossard S, Murrell DF. Plaque-like cutaneous mucinosis: case report and literature review. *Am J Dermatopathol*. 2012;34:e50-e54.
10. Carder KR, Fitzpatrick JE, Weston WL, Morelli JG. Self-healing juvenile cutaneous mucinosis. *Pediatr Dermatol*. 2003;20: 35-39.
11. Cowen EW, Scott GA, Mercurio MG. Self-healing juvenile cutaneous mucinosis. *J Am Acad Dermatol*. 2004;50:S97-S100.