A Persistent "Pimple" in a 5-Year-Old Girl

A 5-year-old girl presented for evaluation of a slowly enlarging, asymptomatic "pimple" below the nose that appeared 3 months before presentation. The child was otherwise healthy, and the mother had a history of thyroid cancer. On physical examination, subjacent to the nasal columella was a 4-mm, apricot-colored, dome-shaped, firm papule with overlying telangiectasias (Figure 1). Dermoscopy revealed a conspicuous pink-red rim in a "setting sun" pattern (Figure 2). Given the family history of cancer, the parents were concerned about the possibility of malignancy and elected to proceed with a skin biopsy. A shave biopsy of the lesion demonstrated a dense infiltrate with multiple giant cells amidst an abundance of lipidized histiocytes and scattered inflammatory aggregates composed of lymphocytes and eosinophils (Figure 3). Based on these findings, a diagnosis of juvenile xanthogranuloma (JXG) was established.

JXG is an uncommon, self-limiting non-Langerhans' cell histiocytosis that occurs predominantly in infants (71% of cases occurring within the first year of life) with a slight male preponderance.[1] Whether JXG is a reactive or neoplastic process is not currently established, although it is believed to develop after an infectious or physical insult that in turn triggers a granulomatous histiocytic reaction.[2] It has also been proposed that lesions may arise owing to increased intracellular synthesis of cholesterol within dermal macrophages.[2] Clinically, JXGs typically appear as well-demarcated papules or nodules whose color varies based on the stage of the lesion.[3] Early lesions display a pink-red color with a yellow hue, whereas well-established ones characteristically feature a yellow-brown color with or without telangiectasias.[3] JXG have a predilection for arising on the face and neck during infancy with spontaneous regression ensuing within the first several years of life.[3] The dermoscopic appearance of JXG can vary based on its stage of maturation; however, the setting-sun appearance is most

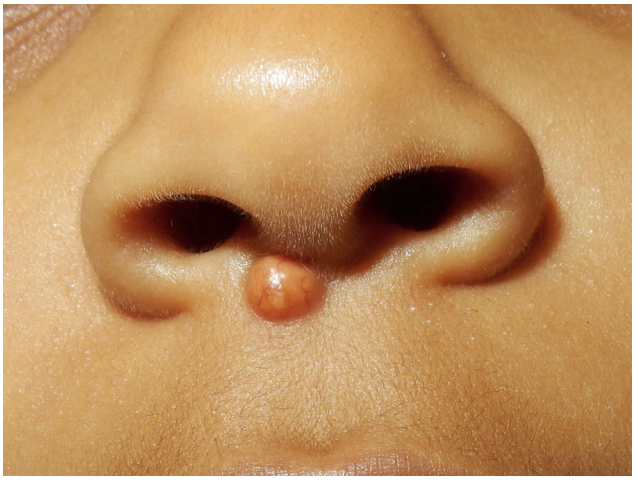

Figure 1. A 4-mm, apricot-colored, dome-shaped, firm papule with overlying telangiectasias below the nasal columella.

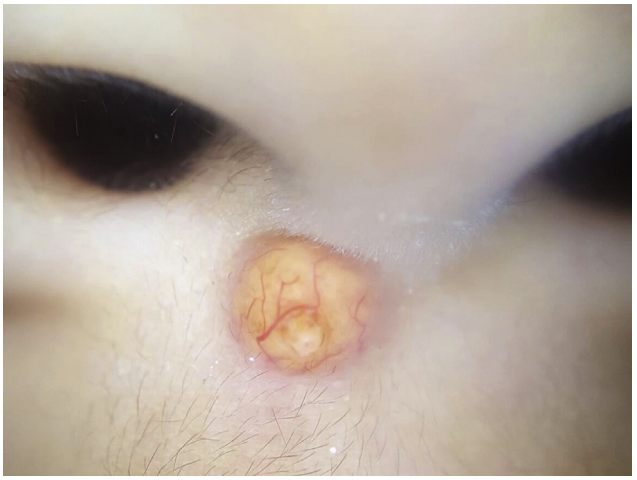

Figure 2. Pink-red rim in a "setting-sun" pattern seen by dermoscopy.

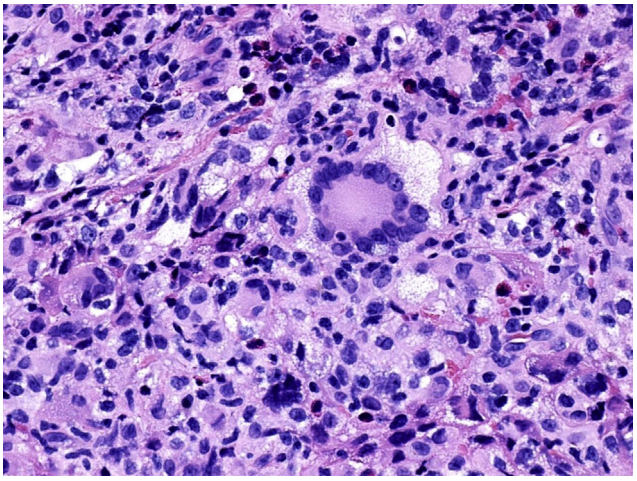

Figure 3. Shave biopsy demonstrating a dense infiltrate with multiple giant cells amidst an abundance of lipidized histiocytes and scattered inflammatory aggregates composed of lymphocytes and eosinophils. Hematoxylin-Eosin 40× (original magnification x400).

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suggestive and is characterized by an erythematous border encompassing the orange-yellow lesion.[4] Although rare, JXG may be associated with potentially serious extracutaneous manifestations, particularly in patients with multiple skin lesions.[3] The most common extracutaneous site involved is the iris, especially in young children, with possible complications including intraocular hemorrhage, glaucoma, and loss of vision.[3] Mortality is rare, but can occur owing to liver failure or central nervous system disease in patients with multiple extracutaneous lesions.[5] Additionally, JXG has been associated with the development of juvenile chronic myelogenous leukemia, particularly in patients with concomitant neurofibromatosis type 1.[6]

Most cutaneous lesions spontaneously regress; therefore, therapy is not usually required, although surgical removal can be pursued for cosmetic reasons.[2] Ophthalmic examination is recommended for patients under 2 years old with multiple, small (<10 mm) lesions.[7] In conclusion, although an uncommon entity, clinicians should consider JXG in young children presenting with single or multiple pink-red or yellow-brown papules or nodules, especially those with a setting-sun pattern on dermoscopy. ■

Katheryn A. Bell, BA
Department of Dermatology
Georgetown University School of Medicine
Washington, DC

Kalyani Marathe, MD
Department of Dermatology
University of Cincinnati and
Cincinnati Children's Hospital
Cincinnati, Ohio

Katherine T. Burke, MD
Michael A. Cardis, MD
Department of Dermatology
Medstar Washington Hospital Center
Georgetown University
Washington, DC

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Milia-Like Idiopathic Calcinosis Cutis: When Waiting Is the Best Option

A 6-month-old boy was referred to our department for the presence of diffuse multiple whitish, pseudomilia-like papules on the skin of the extremities since birth. The patient was well-appearing and afebrile, with normal vital signs. The examination revealed multiple, yellow-white, firm, small, subcutaneous nodules and papules (**Figure 1**, A-C). The lesions were present only on the extremities, arms, and legs. The lesions were in different stages, with a precise evolution process toward extrusion of a whitish material, erosion, and healing (**Figure 1**, A). This clinical presentation was consistent with a diagnosis of calicinosis cutis. To confirm our diagnosis, we performed an incisional biopsy. Histopathologic examination showed the accumulation of well-defined, dense, basophilic material surrounded by fibrous tissue in the dermis (**Figure 2**, A, B). Biochemical studies, including serum calcium, phosphate, parathyroid hormone, vitamin D, and organic acid levels were within the normal range, distinguishing idiopathic calcinosis cutis from metastatic, dystrophic, and iatrogenic calcinosis. Based on pathologic and clinical findings, the patient was diagnosed with milia-like idiopathic calcinosis cutis (MICC).[1-3]

Characteristically, MICC lesions appear as multiple whitish to skin colored, firm, tiny milia-like papules, mostly on the hands and feet. Lesions may have an erythematous halo and may perforate, causing transepidermal calcium elimination. The differential diagnosis is with molluscum, warts, milia, or inclusion cysts. Although definitive confirmation of MICC is based on histopathologic diagnosis, dermoscopy may be helpful in the differential diagnosis. Since the first description in 1978, numerous reports confirmed a strong association between MICC and Down syndrome[2,3]; however, there are a small

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