Diagnosing juvenile xanthogranuloma with reflectance confocal microscopy

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Key words: juvenile xanthogranuloma; reflectance confocal microscopy.

INTRODUCTION

Juvenile xanthogranuloma (JXG) is a type of non-Langerhans cell histiocytosis, in which dendrocytes proliferate and accumulate in the dermis. JXG most frequently occurs in infants and children, presenting as a pink or yellow papule or nodule on the head and neck that spontaneously resolves over time.[1] A "setting sun" appearance, defined by a central orange-yellow papule or nodule surrounded by linear branched vessels and erythema, is the classic dermoscopic presentation of JXG. Microscopic evaluation is sometimes necessary to distinguish JXG from a congenital nevus, especially if pigmented. The use of in vivo reflectance confocal microscopy (RCM) can avoid an invasive procedure in pediatric patients.

CLINICAL PRESENTATION

A 12-year-old boy presented to our clinic with a lesion on his left cheek, which had been growing for 2 months. On physical examination, a 3.5- × 2.6-mm pink papule was seen among a background of ephelides (Fig 1).

DERMOSCOPIC APPEARANCE

Dermoscopy showed a homogenous yellow-white center surrounded by a network of erythema (Fig 1). Additionally, there was a scattered distribution of light brown uniform pigmentation with moth-eaten borders. Differential diagnosis included intradermal nevus, molluscum contagiosum, and JXG. Given this nonspecific pattern, microscopic evaluation was pursued.

Abbreviations used:

DEJ:	dermoepidermal junction
JXG:	juvenile xanthogranuloma
RCM:	reflectance confocal microscopy

CONFOCAL MICROSCOPY APPEARANCE

The patient and guardian opted for RCM over biopsy. Confocal images showed a well-circumscribed area with an irregular honeycomb pattern in the epidermis (Fig 2). At the dermoepidermal junction (DEJ), dermal papillae lacked a ringed-edged pattern (Fig 3). In the dermis, multiple large round and ovoid cells with a foamy cytoplasm were found in the center of the lesion (Fig 4). Also present in the dermis were multinucleated large cells (62 to 67 μm in size) with a hyperrefractile cytoplasm, forming a peripheral rim. Dermal and junctional nests were absent. The RCM diagnosis was JXG, with a recommended 1-year follow-up. Per the parent, the lesion remained unchanged in the 8 months since imaging.

DISCUSSION

RCM provides noninvasive imaging capability to a depth of 200 to 300 μm, with resolution comparable to histopathology.[2] It is a well-suited alternative for diagnosing JXG, as these lesions often appear in aesthetically sensitive areas, biopsies are challenging to perform in pediatric patients, and there is risk for scar expansion as the child grows.[3]

A combination of features on RCM allowed for diagnosis of JXG. The irregular honeycomb pattern was likely caused by disturbance from the underlying dermal tumor pressing up against the epidermis. Lack

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Funding sources: None.
Conflicts of interest: Babar K. Rao serves as a consultant for Caliber I.D. All other authors have no conflicts of interest to disclose.

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JAAD Case Reports 2020;6:975-6.
2352-5126

https://doi.org/10.1016/j.jdcr.2020.07.034

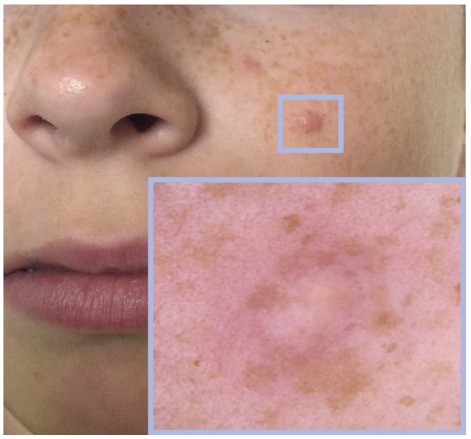

Fig 1. Clinical presentation. A 3.5- × 2.6-mm pink papule on the left cheek among a background of ephelides. Dermoscopic presentation (*inset*). A homogenous yellow-white center surrounded by a network of erythema and scattered light brown uniform pigmentation with moth-eaten borders.

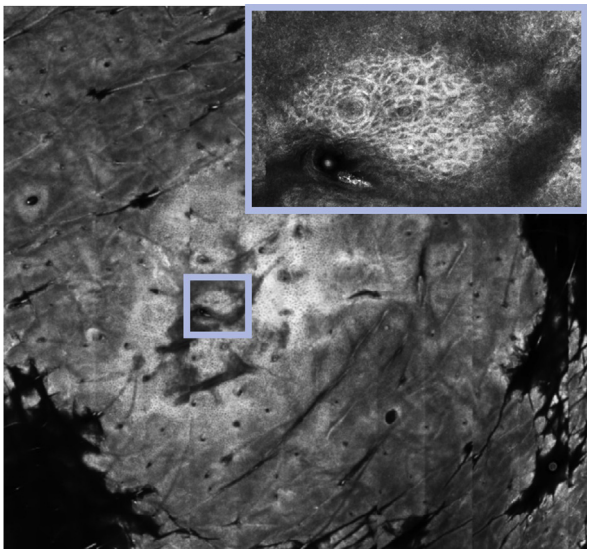

Fig 2. RCM mosaic at the epidermis. Well-circumscribed area with an irregular honeycomb pattern.

of melanocytic nests and a ringed-edged pattern at the DEJ ruled out a nevus. Additionally, the absence of round refractile cystic areas in the dermis eliminated the possibility of molluscum contagiosum. Instead, large round and ovoid cells with a foamy cytoplasm corresponded to xanthomatous histiocytes. Other large, multinucleated cells, with a hyperrefractile peripheral rim, mimicked Touton giant cells, as described in the literature.[4,5] Similar to histopathology, the presence of foam cells and Touton giant cells in a granulomatous formation is sufficient to make the diagnosis of xanthogranuloma on RCM. This finding shows the viability of in vivo RCM as an alternative to biopsy in similar cases of JXG.

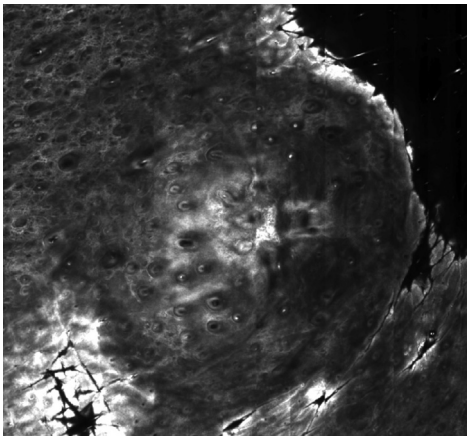

Fig 3. RCM mosaic at the DEJ.

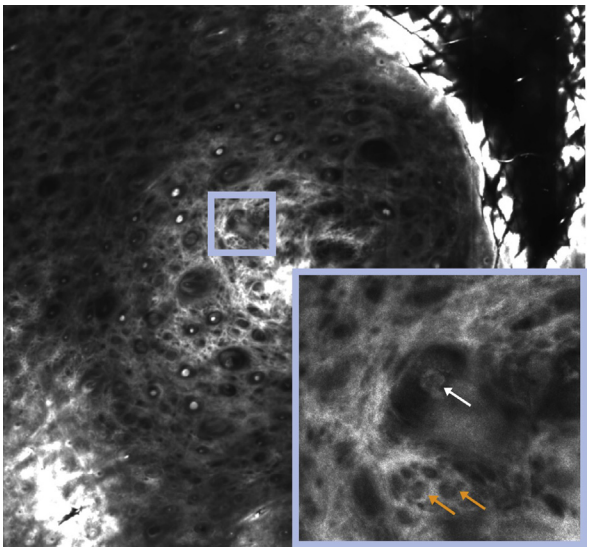

Fig 4. RCM mosaic at the dermis. The inset displays round and ovoid cells with a foamy cytoplasm in the center of the lesion corresponding to xanthomatous histiocytes (*orange arrows*). Large cells are present in the dermis containing multiple nuclei and a bright peripheral cytoplasm, mimicking Touton giant cells in histology (*white arrow*).

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