

# Solitary orange papule on the back of a middle-aged man

Shamir Geller<sup>1</sup>, Melissa Pulitzer<sup>2</sup>, Patricia L. Myskowski<sup>1</sup>

<sup>1</sup> Dermatology Service, Department of Medicine, Memorial Sloan Kettering Cancer Center and Weill Cornell Medicine, New York, NY, USA  
<sup>2</sup> Department of Pathology, Memorial Sloan Kettering Cancer Center and Weill Cornell Medicine, New York, NY, USA

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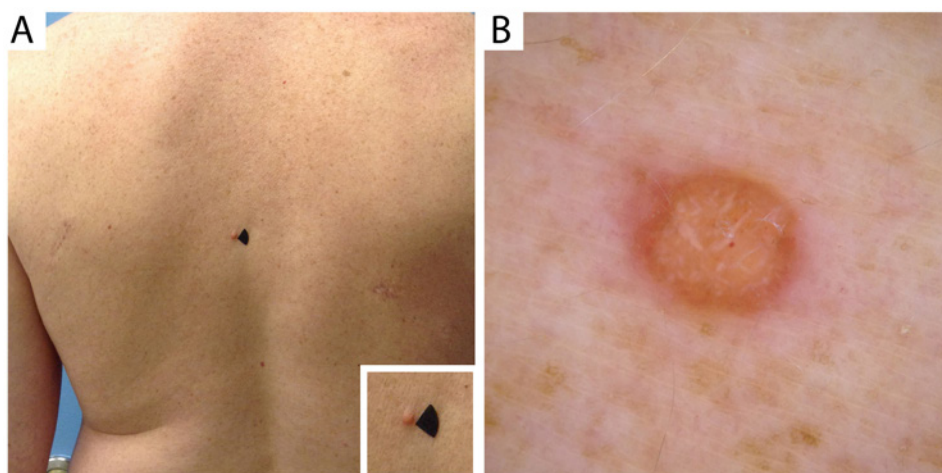
**Corresponding author:** Shamir Geller, MD, Dermatology Service, Memorial Sloan Kettering Cancer Center, 16 East 60th Street, New York, NY 10022, USA. Email: [shamirgeller@gmail.com](mailto:shamirgeller@gmail.com)

## The patient

A 51-year-old man with a history of recurrent primary cutaneous marginal zone lymphoma involving his back returned for a follow-up visit at our cutaneous lymphoma clinic. He complained of a several-month history of a new asymptomatic papule on the mid-thoracic back. He denied fevers, chills, night sweats or fatigue. Clinical examination identified a 3 to 4 mm red to orange-colored papule on the middle back (Figure 1A). Dermoscopic examination of the lesion revealed

an erythematous border encircling an orange-yellow area with white linear streaks and few dotted vessels (Figure 1B). A biopsy and histopathological examination performed to exclude recurrence of skin lymphoma demonstrated a nodular dermal infiltrate of histiocytes with vacuolated foamy xanthomatous cytoplasm and Touton-type multinucleated giant cells (Figure 2). The histiocytes stained positively for CD68 and were negative for Sox10.

### *What is your diagnosis?*



**Figure 1.** Clinical image of the red to orange-colored papule on the patient's upper back marked with a black triangular sticker (A). Polarized, non-contact dermoscopy images of the lesion revealing a structureless orange-yellow area with erythematous border. White linear streaks and few dotted vessels are seen in the center of the lesion; 10x original magnification (B). [Copyright: ©2018 Geller et al.]

## Answer

Adult-onset xanthogranuloma (juvenile xanthogranuloma presenting in an adult)

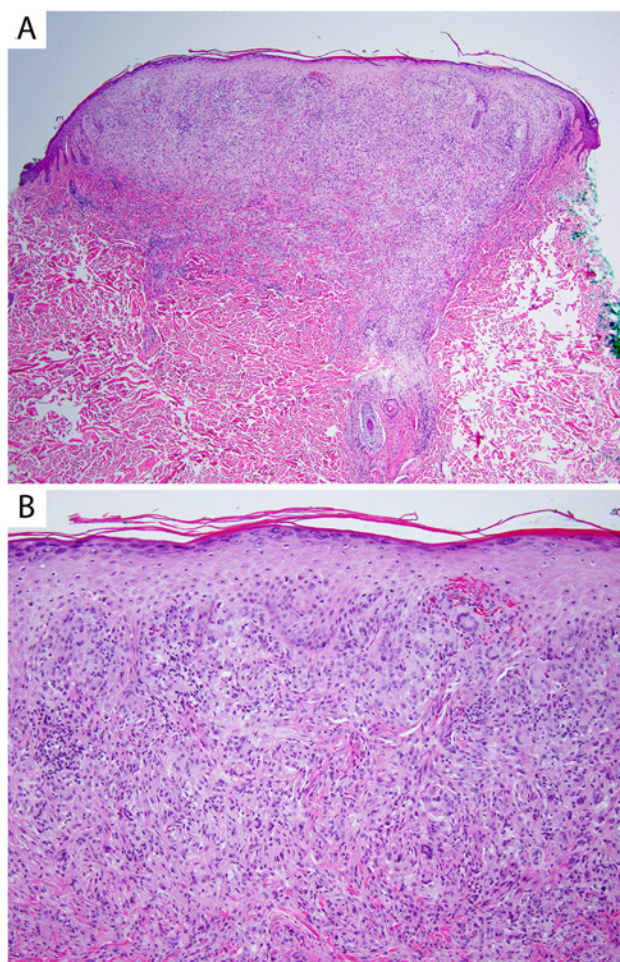
## Discussion

Juvenile xanthogranuloma (JXG) is the most common non-Langerhans cell histiocytosis, which presents either as a single lesion or as multiple asymptomatic pink to red-brown dome-shaped papules or nodules (2 mm to 2 cm in diameter) that become yellow-brown as they mature. JXG most often affects infants and young children; however, it can appear in adults in their 20s and 30s and can even present in the elderly. JXG's course is usually self-limited in the pediatric population and treatment is not necessary, although, in adults, in whom the term adult-onset xanthogranuloma is favored, it may not regress spontaneously [1].

Histologically, JXG shows a dense histiocytic infiltration involving the dermis but sometimes extending subcutaneously. Monomorphic histiocytes with eosinophilic cytoplasm are seen in the early lesions, while in mature JXG the histiocytes acquire a foamy and xanthomatous appearance, Touton giant cells are identified, and lymphocytes, eosinophils, and plasma cells can be found throughout the infiltrate. Regressing lesions show a proliferation of fibroblasts and fibrosis.

The dermoscopic pattern of JXG was first described in 2007 by Palmer et al., who described a characteristic orange-yellow “setting sun” appearance [2]. Other dermoscopic features that have been reported in JXG cases include clouds of pale yellow globules, subtle pigment network, whitish streaks, and branched linear or dotted vessels [3,4]. Song et al. studied the correlation between dermoscopic appearance and histopathological findings and found that the dermoscopic features correlated with the histologic level of maturation of the JXG lesions. The setting sun appearance was found in early evolutionary and in fully developed JXG. In fully developed lesions, the surrounding erythema decreased, and yellow globules became more evident as the histiocytes transformed to more xanthomatous cells, while in late regressive lesions, prominent whitish streaks were identified and were suggested to correspond to fibrosis [5]. It was suggested that dermoscopy might be helpful in differentiating JXG from other conditions that appear as solitary yellow-orange lesions [6,7].

The main differential diagnoses of JXG include sebaceous tumors [8], xanthomatous dermatofibroma, basal cell carcinoma, granulomatous dermatoses, and nonpigmented Spitz nevus [4,9]. Table 1 shows the dermoscopic features of all such conditions. In the present case, the clinical diagnosis



**Figure 2.** Histopathology. Circumscribed nodular dermal infiltrate effacing rete ridges and elevating and attenuating epidermis; hematoxylin-eosin stain, 40x original magnification (A). Foamy histiocytes and Touton-type multinucleated giant cells (asterisks) comprise the majority of the infiltrate. Lymphocytes percolate through the histiocytic tumor; hematoxylin-eosin stain, 100x original magnification (B). [Copyright: ©2018 Geller et al.]

of cutaneous B-cell lymphoma was highly suspected, but the dermoscopic setting sun pattern was quite different from the typical dermoscopic features of the salmon-colored area and linear-irregular vessels typically seen in such a type of cutaneous lymphomas [10,11] (Table 1).

Reflectance confocal microscopy (RCM) and high-definition optical coherence tomography (HD-OCT) features of JXG and adult-onset xanthogranuloma were reported recently. Highly refractive atypical large cells of variable diameter were seen in the dermis, and some of them had pleomorphic nuclei, corresponding to Touton-type giant cells that were seen in histopathologic examination [9,12].

JXG and adult-onset xanthogranuloma can present with characteristic dermoscopic features of an orange-yellow structureless pattern with an erythematous border (setting sun sign) and white streaks. Dermoscopy can be helpful in diagnosing these benign skin conditions in a noninvasive manner.

**TABLE 1. Dermoscopic clues for the differential diagnosis of the studied solitary orange colored papule.**

Diagnosis	Dermoscopic Features
Juvenile xanthogranuloma [5]	Erythematous border circling an orange-yellow area (“setting sun”), white linear streaks and linear/branched vessels
Basal cell carcinoma [9]	Arborizing vessels with a pink background, ulcerations, leaf-like structures, blue-gray ovoid nests and white shiny streaks
Cutaneous B-cell lymphoma [10]	Salmon-colored background/area and serpentine (linear-irregular) vessels
Granulomatous dermatoses [13,14]	Structureless yellow-orange area with linear or branching vessels
Sebaceous tumors [8]	<ul style="list-style-type: none"> <li>• Radially arranged, elongated crown vessels surrounding structureless yellow areas</li> <li>• Yellow comedo-like globules and branching arborizing vessels</li> </ul>
Nonpigmented Spitz nevus [15]	Regularly distributed dotted vessels (coiled, hairpin or linear-irregular in elevated/nodular nevi) with reticular depigmentation (inverse white network)
Xanthomatous dermatofibroma [4]	Homogeneous yellow area with peripheral delicate pigment network

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