DERMATOLOGY PRACTICAL & CONCEPTUAL
www.derm101.com

An eruption of yellow-red papules on the trunk, arms, and legs of an adult

L. Claire Hollins[1], Grace W. Weyant[2], Mark B. Gibbs[3], Elizabeth V. Seiverling[1,4]

1 Department of Dermatology, Penn State Health, Milton S. Hershey Medical Center, Hershey, PA, USA
2 Department of Pathology, Lancaster General Health, Lancaster, PA, USA
3 Lebanon VA Medical Center, Lebanon, PA, USA
4 Department of Family and Community Medicine, Penn State Health, Milton S. Hershey Medical Center, Hershey, PA, USA

Key words: dermoscopy, eruptive adult xanthogranuloma, xanthogranuloma, setting sun sign

Citation: Hollins LC, Weyant GW, Gibbs MB, Seiverling EV. An eruption of yellow-red papules on the trunk, arms, and legs of an adult. *Dermatol Pract Concept.* 2018;8(3):177-179. DOI: https://doi.org/10.5826/dpc.0803a05

Received: December 14, 2017; **Accepted:** February 26, 2018; **Published:** July 31, 2018

Funding: None.

Competing interests: The authors have no conflicts of interest to disclose.

All authors have contributed significantly to this publication.

Corresponding author: L. Claire Hollins, MD, 500 University Drive, Suite 100, Hershey, PA 17033 USA. Email: lhollins@pennstatehealth.psu.edu.

Case Presentation

A 68-year-old Caucasian man presented with a 6-month history of firm skin papules, starting on his face then gradually moving caudally, progressing to his arms, trunk, and legs (Figure 1). Many of his lesions were asymptomatic. However, a few of the papules were pruritic. The itching was alleviated by topical steroids.

On physical exam, multiple yellow-brown to red firm papules and small nodules were noted on the face, trunk, and proximal extremities. Dermoscopic examination demonstrated orange-yellow color surrounded by an erythematous border with occasional linear branched vessels (Figure 2).

Histopathologic evaluation of 4 of the lesions revealed spindle cells in the upper dermis (Figure 3). All 4 specimens had similar findings. Immunohistochemical studies showed positivity for CD163, as well as Factor XIIIa. There was no significant positivity for CD34. Laboratory data revealed a hemoglobin A1C of 9.3% and triglycerides of 461 mg/dL. An ophthalmology consult was obtained and no ocular pathology was found. A serum protein electrophoresis was negative. With this information, a diagnosis of adult eruptive xanthogranulomas was made.

Discussion

A xanthogranuloma (XG) is a type of non-Langerhans cell histiocytosis that most commonly presents in infancy and childhood; however, several cases have been described in adults. Xanthogranulomas present as dome-shaped papules or nodules on the skin. The condition typically presents with a solitary yellow to reddish-brown growth. However, several cases have been described of eruptive XGs in adults, or adult xanthogranulomas (AXGs) [1-3]. The differential diagnosis of XGs is extensive and is based on clinical appearance, associated comorbidities, histology, and immunohistochemistry.

Given the patient's elevated Hg A1C, eruptive xanthomas (EXs) were considered. In addition to diabetes mellitus, EXs

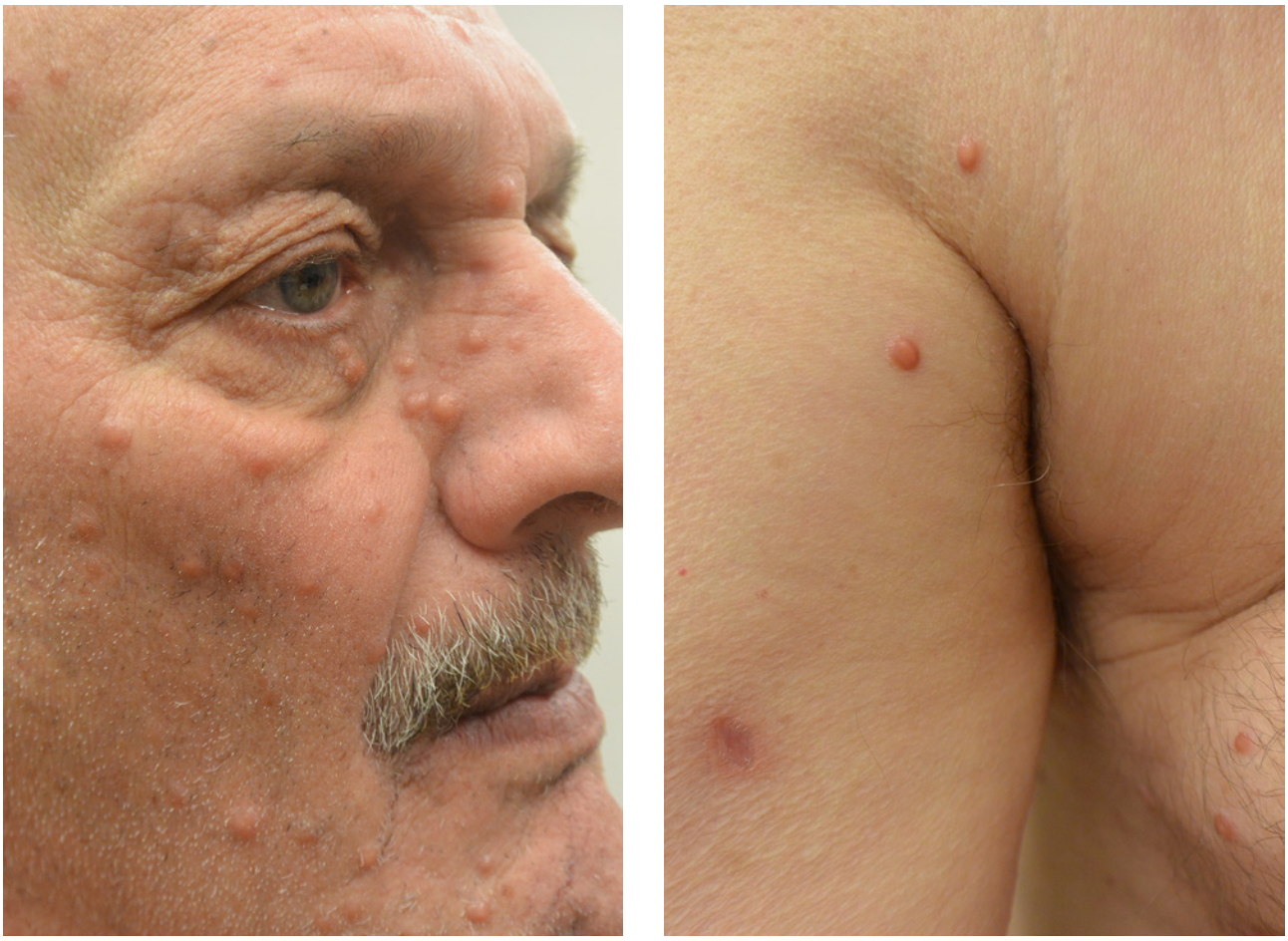

Figure 1. Yellow-red papules on the face, right arm, and trunk. [Copyright: ©2018 Hollins et al.]

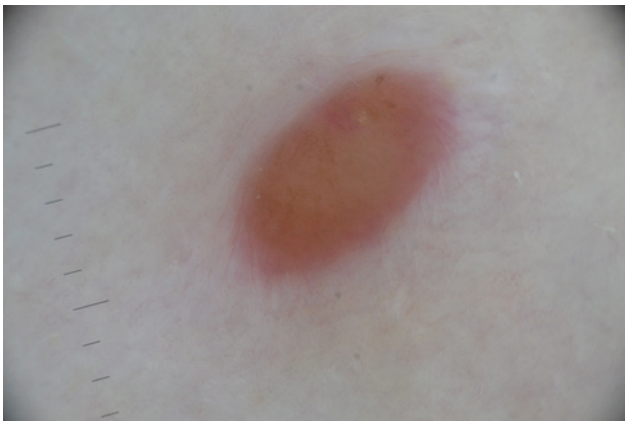

Figure 2. The "setting sun" sign of an adult xanthogranuloma on dermoscopy. [Copyright: ©2018 Hollins et al.]

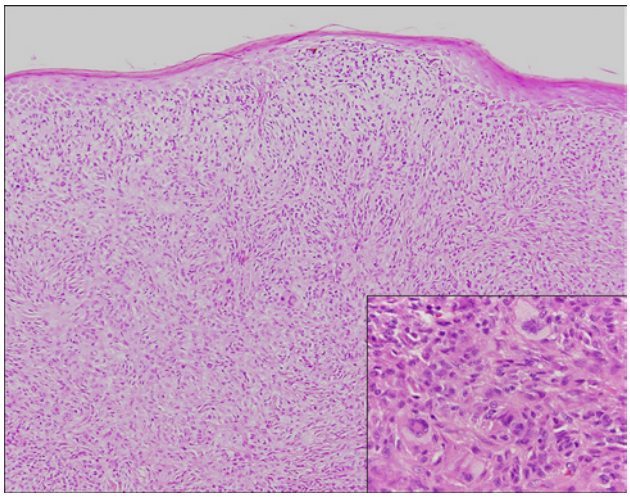

Figure 3. Dermal proliferation of predominantly spindle-shaped histiocytes (hematoxylin and eosin [H&E] staining ×100). Inset: Foamy histiocytes and Touton giant cells are also present (H&E ×400). [Copyright: ©2018 Hollins et al.]

are associated with dramatically increased low-density lipoprotein (LDL) levels and hypertriglyceridemia. Our patient had LDL levels around 100 at the time of biopsy. His triglycerides were only moderately elevated at 461 mg/dL, whereas serum triglyceride levels typically exceed 1500 to 2000 mg/dL in EX, leading us to favor other diagnoses.

Two other diagnoses included in the differential are necrobiotic xanthogranulomas (NXGs) and progressive nodular histiocytosis (PNH), both of which can mimic eruptive AXG. To rule out NXGs, a serum protein electrophoresis was ordered and found to be negative for monoclonal gammopathy. Additionally, while NXG can be seen on the trunk and extremities, it is typically seen primarily in the periorbital region, and our patient had papules over his trunk and

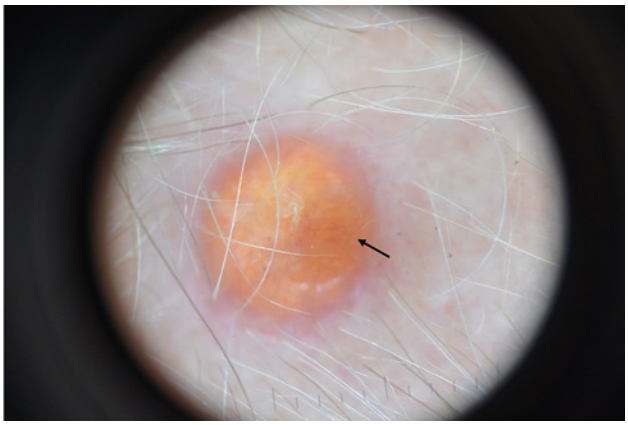

Figure 4. Linear, branched vessels on the edge may mimic basal cell carcinoma on dermoscopy. [Copyright: ©2018 Hollins et al.]

extremities, in addition to his face. Histopathology was also not consistent with NXG. Finally, NXG can have ocular involvement, which we ruled out with an ophthalmologic examination. PNH was thought to be a less likely diagnosis, given the clinical features that are usually seen with this disease, such as facies leonina, and its progressive nature. Our patient has normal facies and to date has not experienced progression of the disease, but will continue to be monitored. Finally, generalized eruptive histiocytosis was also on the differential as a type of symmetric papular eruption of the skin on the trunk and proximal extremities. However, this diagnosis was less likely given the histopathologic findings. Our patient had multiple foamy spindle-shaped histiocytes and Touton giant cells.

We favored the diagnosis of eruptive AXGs, described previously as the presence of more than 5 yellowish papules and nodules over the body [2]. AXG usually occurs in men, at a ratio of 1.6:1, and more than 90% of the time, the lesions are found on the trunk, followed by the head and neck. It has been reported on the extremities, albeit rarely. The demographic fits our patient well, and the biopsied specimen confirms the diagnosis of AXG.

Because XG in adults is uncommon, the diagnosis might not be immediately obvious. Dermoscopy can be employed to help differentiate it from other diagnoses on the differential, such basal cell carcinoma or sebaceous hyperplasia [4]. On dermoscopy, XGs, particularly the juvenile type, have been described as looking like a "setting sun" [5]. This orange-yellow color is not typical of the yellow "popcorn" appearance of sebaceous hyperplasia. The vessels in sebaceous hyperplasia also differ from XGs. Sebaceous hyperplasia has crown vessels [4]. Furthermore, XGs may have what look like arborizing vessels, as seen in basal cell skin cancer. However, XGs will have a more yellow-orange hue on dermoscopy than one might find in a basal cell carcinoma (BCC) and may also have clouds of pale yellow dispersed through the papule (Figure 1). In one study, 90.9% of confirmed XGs had this "setting sun" finding [5]. If suspicion is still high for a BCC, look for other findings of BCC such as leaf-like structures, blue-gray ovoid nests or globules, spoke-wheel structures, shiny white areas, or ulceration.

Conclusions

Our case represents the somewhat rare diagnosis of AXG. The diagnosis was suspected based on dermoscopy and clinical morphology and was confirmed by histopathology (and laboratory ruled out other, similar diagnoses). XGs will usually resolve in children, while only approximately half of adult XGs regress [1]. Our patient was treated with shave removal of symptomatic XGs with good results; however, isotretinoin can also be used, with possible recurrence of lesions with discontinuation. Currently, the etiology of AXG in otherwise healthy adults is unclear. Dermoscopy can help with diagnosis and will show a characteristic yellow-orange hue, called the "setting sun" sign, and is sometimes accompanied by linear, branched vessels around the border (Figure 4). This can be reassuring to both patients and physicians to help rule out skin carcinoma in which the color is not yellow and vessels tend to arborize across the lesion. Finally, while most cases of adult eruptive XGs are idiopathic, reports have linked them with hematologic and rarely solid organ malignancy; therefore, close monitoring of patients with eruptive AXGs is warranted [6].

References

1. Saad N, Skowron F, Dalle S, Forestier JY, Balme B, Thomas L. Multiple adult xanthogranuloma: case report and literature review. *Dermatology.* 2006;212(1):73-76. doi: 10.1159/000089027.
2. Chang SE, Cho S, Choi JC, et al. Clinicohistopathologic comparison of adult type and juvenile type xanthogranulomas in Korea. *J Dermatol.* 2001;28(8):413-418. doi: 10.1111/j.1346-8138.2001.tb00002.x.
3. Whitmore SE. Multiple xanthogranulomas in an adult: case report and literature review. *Br J Dermatol.* 1992;127(2):177-181. doi: 10.1111/j.1365-2133.1992.tb08053.x.
4. Lovato L, Salerni G, Puig S, Carrera C, Palou J, Malvehy J. Adult xanthogranuloma mimicking basal cell carcinoma: dermoscopy, reflectance confocal microscopy and pathological correlation. *Dermatology.* 2010;220(1):66-70. doi: 10.1159/000264670.
5. Song M, Kim SH, Jung DS, Ko HC, Kwon KS, Kim MB. Structural correlations between dermoscopic and histopathological features of juvenile xanthogranuloma. *J Eur Acad Dermatol Venereol.* 2011;25(3):259-263. doi: 10.1111/j.1468-3083.2010.03819.x.
6. Vadeboncoeur S, Provost N. Multiple xanthogranulomas in an adult: known entity, new association. *J Cutan Med Surg.* 2016;20(5):474-477. doi: 10.1177/1203475416640795.