DERMATOLOGY PRACTICAL & CONCEPTUAL
www.derm101.com

Asymptomatic cluster of violaceous papules

Reza Yaghoobi[1], Karan Lal[2], Amir Feily[3]

1 Department of Dermatology, Jundishapur University of Medical Sciences, Ahvaz, Iran
2 New York Institute of Technology College of Osteopathic Medicine, New York, NY, USA
3 Skin and Stem Cell Research Center, Tehran University of Medical Sciences, Tehran, Iran

Citation: Yaghoobi R, Lal K, Feily A. Asymptomatic cluster of violaceous papules. Dermatol Pract Concept 2015;5(3):3. doi: 10.5826/dpc.0503a03

Corresponding author: Amir Feily, MD, Skin and Stem Cell Research Center, Tehran University of Medical Sciences, Tehran, Iran. Email: Dr.feily@yahoo.com

Case report

A 56-year-old male patient was seen in the dermatology clinic for a 25-year history of an asymptomatic cluster of violaceous papules in a coalescing pattern on the right lateral thigh (Figure 1). No other significant findings were found on physical exam and all laboratory results were within normal demographic ranges. The patient reported to have hypothyroidism that was diagnosed one year ago and trigeminal neuralgia for which he was taking carbamazepine and calcium for five years' duration. An incisional biopsy from one of the lesions involving lesion and peri-lesional skin was performed.

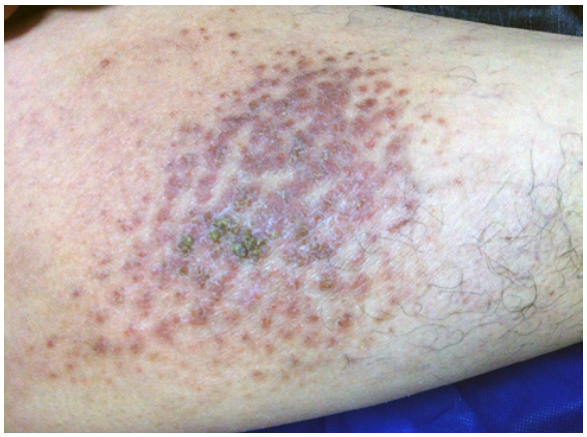

Figure 1. Multiple violaceous papules on the right lateral thigh. [Copyright: ©2015 Yaghoobi et al.]

Discussion

Dermatopathological examination revealed hyperkeratosis, an acanthotic epidermis with elongation of rete ridges, and a superficial to deep dermal inflammatory infiltrate (Figure 2). An Alcian blue stain at a pH of 2.5 was performed indicating the presence of mucin within the papillary dermis (Figure 3). Clinicopathologic examination confirmed the diagnosis of mucinous nevus.

Mucinous nevus is a benign hamartomatous lesion, first described by Redondo Bellian in 1993, is a rare type of primary cutaneous mucinosis [1]. Its name stems from the lesion's striking appearance to a nevus and the presence of dermal mucin [2]. Histologically, two types of MN exist: connective tissue nevus of the proteoglycan (CTNP), and the

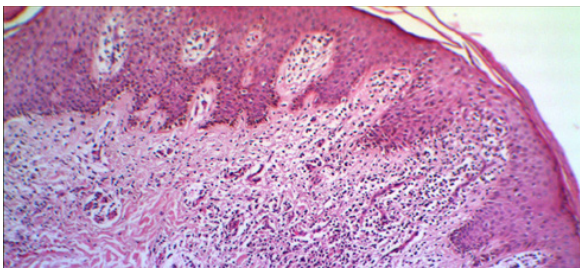

Figure 2. Hematoxylin and eosin (100X) showing hyperkeratosis, acanthosis along with elongation of rete ridges, and a superficial to deep dermal inflammatory infiltrate. [Copyright: ©2015 Yaghoobi et al.]

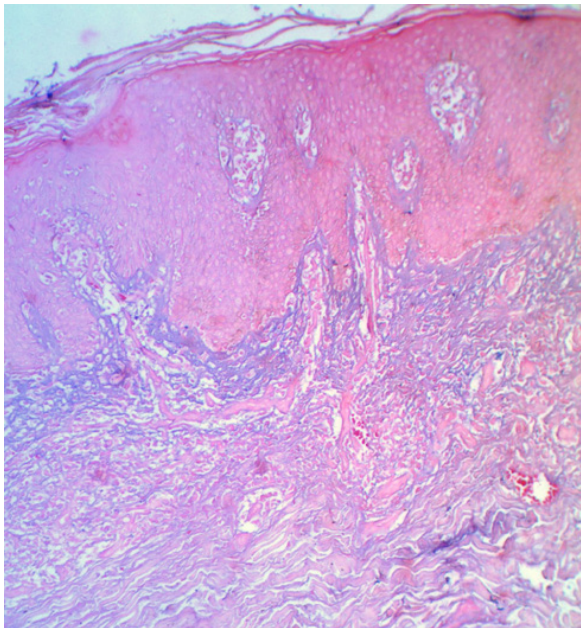

Figure 3. Alcian blue stain (100X) showing purple mucin deposits within the papillary dermis. [Copyright: ©2015 Yaghoobi et al.]

combined epidermal connective tissue nevus of the proteoglycan. CTNP is classified by the presence of dermal mucin whereas combined epidermal nevus of the proteoglycan has features consistent with epidermal nevi, hyperkeratosis, elongation of rete ridges, and an acanthotic epidermis [3]. A case has been reported of combined epidermal CTNP, with mature adipocytes within the superficial dermis that was misdiagnosed as nevus lipomatosis superficialis indicating the importance of Alcian blue staining in suspicious lesions [4].

Clinically the lesions may be congenital [1] or may develop later on in life [5,6,7], as they did in our patient and can vary in presentation most often appearing as papules, plaques, or even a pedunculated mass [8] in either a rare zosteriform pattern or a more commonly a unilateral linear nevoid pattern. The most common location for these lesions is the lower back4; however, cases have been reported on the abdomen [9], thighs [10], and face [11].

The pathogenesis of mucinous nevus remains uncertain; however, many theories exist regarding the origin of the mucin. The most supported theory is that the mucin is overproduced by abnormal fibroblasts within the superficial dermis [12] Characteristically, due to positive staining with Alcian blue, at a pH of 2.5, and not at 0.5, mucin is considered to be of hyaluronic acid origin [5].

Due to the subtleties in its presentation and histology, differential diagnoses to consider include: nevoid follicular mucinosis [13], cutaneous mucinosis of infancy [14], cutaneous focal mucinosis [14], epidermal nevus [14], nevus lipomatosis superficialis [4], papular mucinosis [14], and the various connective tissue nevi. Other features such as dermal inflammation may be present as seen on the hematoxylin and eosin stained specimen from the biopsy of our patient's lesion. Although this moderately dense inflammatory infiltrate most likely represents exogenous irritation from pruritus, it may distract pathologists to focus on the inflammatory lesions as opposed to the background features favoring mucinous nevus. For this reason, biopsy with dermatopathological evaluation is necessary with the addition of Alcian blue staining at a pH of 2.5 for identification of dermal mucin, along with Von Geison staining to determine the quantitative extent of elastin fibers within the papillary and reticular dermis.

The benign nature of these lesions requires only excision-based therapy. Some authors have suggested the use of carbon dioxide laser therapy only for MN classified as combined epidermal CTNP [15].

References

1. Redondo Bellon P, Vazquez-Doval J, Idoate M, Quintanilla E. Mucinous nevus. J Am Acad Dermatol 1993;28 (5 Pt 1):797-98.
2. Perez-Crespo M, Lopez-Navarro N, Betlloch I, et al. Acquired and familial mucinous nevus. Int J Dermatol 2011;50(10):1283-85.
3. Rongioletti F, Rebora A. Mucinous nevus. Arch Dermatol 1996;132 (12):1522-23.
4. Song BH, Park S, Park EJ, et al. Mucinous nevus With fat. Am J Dermatopathol 2012;34(8):E146-48.
5. Brakman M, Starink TM, Tafelkruyer J, Bos JD. Linear connective tissue naevus of the proteoglycan type ('naevus mucinosus'). Br J Dermatol 1994;131(9):368-70.
6. Uitto J, Santa Cruz DJ, Eisen AZ. Connective tissue nevi of the skin. Clinical, genetic, and histopathologic classification of hamartomas of the collagen, elastin, and proteoglycan type. J Am Acad Dermatol 1980; 3(5):441-61.
7. Suhr KB, Ro YW, Kim KH, et al. Mucinous nevus: report of two cases and review of the literature. J Am Acad Dermatol 1997;37(2 pt 2):312-3.
8. Lim JH, Cho SH, Kim HO, Kim CW, Park YM. Mucinous naevus with atypical features. Br J Dermatol 2003;148(5):1064–66.
9. Sahara LY, Bastian BC, Bruckner AL. Coalescing, nevoid papules in an infant. Arch Dermatol 2005;141(7):897– 902.
10. Chen CW, Tsai TF, Chen YF, Hung CM. Familial mucinous nevus. Pediatr Dermatol 2008;25(2):288–9.
11. Nayal B, Mathew M, Kumar P, Nair RP. Mucinous naevus: a rare variant of connective tissue naevus. J Interdiscipl Histopathol 2013;1(2):89-92.
12. Chui HH, Chang WY, Chen GS. Mucinous nevus—a case report and review of literature. Dermatol Sinica 2007;25:147-52.
13. Tadini G, Boldrini MP, Brena M, et al. Nevoid follicular mucinosis: a new type of hair follicle nevus. J Cutan Pathol 2013;40 (9):844-47.
14. Esteves T, Ferreira L, Viana I, Bordalo O. Brown plaques on the lower back. Dermatology Online Journal 2010;16(6):11.
15. Chi CC, Wang SH, Lin PY. Combined epidermal-connective tissue nevus of proteoglycan (a type of mucinous nevus): a case report and literature review. J Cutan Pathol 2009;36(7):808-11.