Volume 19 Number 9
September 2013

Letter

Congenital agminated segmental nevi of the chest

Navid Ezra, MD,[1] Anita Haggstrom, MD[1]

Dermatology Online Journal 19 (9): 16

[1]Department of Dermatology, Indiana University School of Medicine, Indianapolis, IN

Correspondence:

Anita Haggstrom, M.D.
Associate Professor of Dermatology and Pediatrics
Email: ahaggstr@iupui.edu
Tel: (317) 944-7744

Introduction

Grouped patterns of pigmented lesions are infrequent. Of the several reports of agminated nevi, most have been Spitz nevi or blue nevi. The distribution of these nevi is often segmental, following a dermatome or the lines of Blaschko. Most segmental nevi are not agminated and develop early in childhood [1]. We describe a rare case of congenital agminated segmental nevi on the chest.

Key Words: congenital nevi, agminated nevi, segmental nevi

Case synopsis

A 21-year-old man was referred with numerous pigmented papules on the right side of his chest (Figure 1). The lesions had been present at birth and had not recently changed in size, color, or texture. The patient was well and had no family history of genetic disorders, melanoma, or dysplastic nevi. Examination showed numerous disseminated brown and skin-colored soft papules. Each measured up to 1 cm in diameter and most were covered with terminal hairs (Figure 2). The lesions were localized to the right side of the chest in an agminated distribution. Other body sites were unaffected.

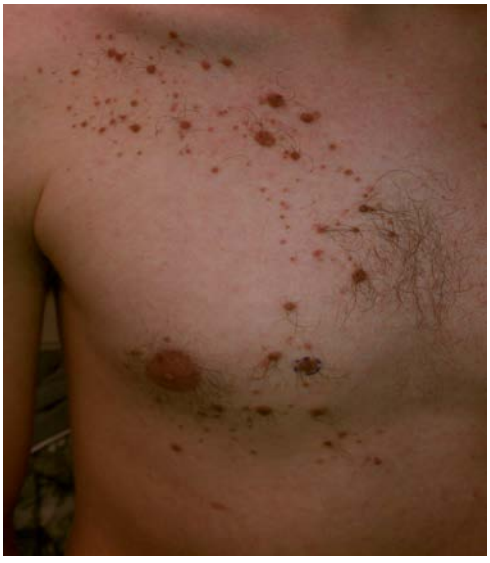

Figure 1. Congenital agminated segmental nevi of the chest. Numerous pigmented papules on the right side of his chest that had been present at birth and had not recently changed in size, color or texture.

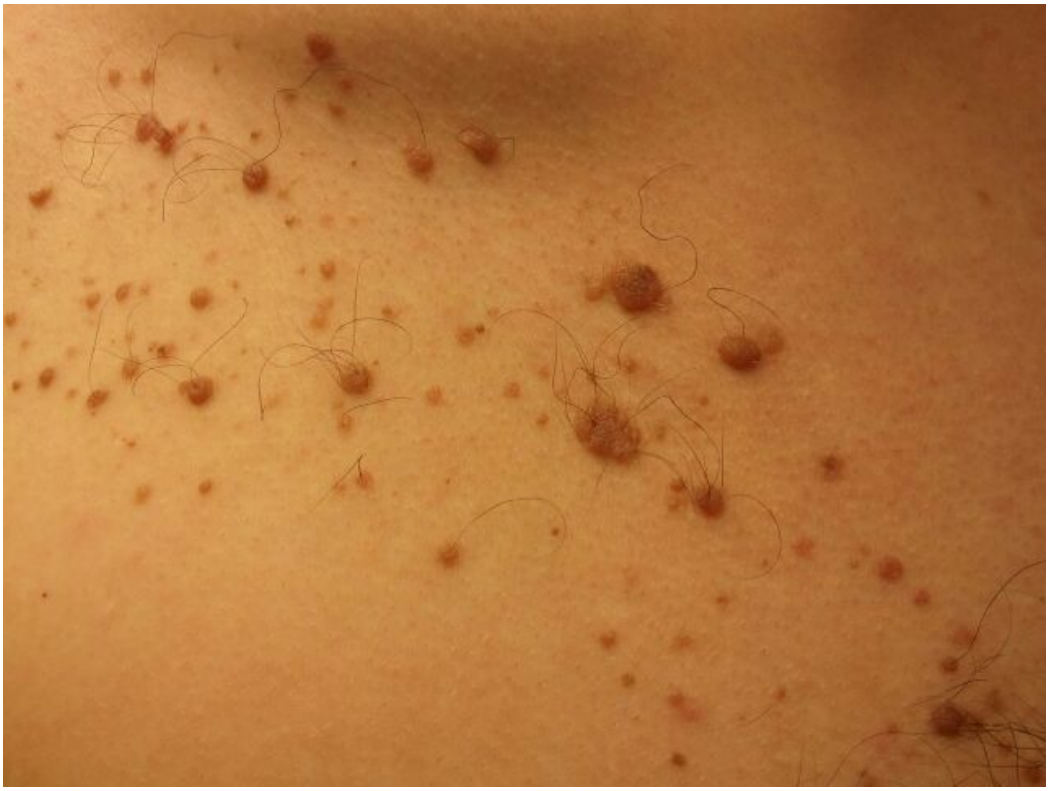

Figure 2. Congenital agminated segmental nevi of the chest. Numerous disseminated brown and skin-colored soft papules localized to the right side of the chest in an agminated distribution. Each measured up to 1 cm in diameter, and most were covered with terminal hairs.

Two shave biopsies were performed to rule out atypia, one of the upper chest and one of the lower chest. Histologic examination was consistent with congenital melanocytic dermal nevi. The epidermis was normal and numerous pigmented nevus cell nests, with a partly neuroid pattern, were present in the upper and mid-dermis. These nests were located around blood vessels and hair follicles and between collagen fibers.

Discussion

Agminated nevi have been reported both as congenital and acquired lesions. They have been reported to follow a Blaschkoid or linear distribution. This linear distribution may be owing to a clonal outgrowth of cells, which carry a major gene responsible for the development of melanocytic nevi [2]. This clone is thought to originate from a somatic mutation at an early stage of embryogenesis [3,4]. Acquired agminated linear nevi may develop along the course of peripheral nerves following cutaneous injury [5], after intense exposure to sunlight, or following radiotherapy [6]. The relationship with cutaneous innervation by peripheral nerves suggests that developing melanocytes are under neural control [7].

Most segmental nevi are not agminated [1] and arise because of perturbations in the proliferation, migration, and differentiation of embryological precursors, including melanocytes [8,9]. Development of numerous acquired melanocytic nevi in a segmental distribution suggests mosaicism for a mutation in a nevus-susceptibility gene [10]. Cutaneous mosaicism often manifests as lines of Blaschko, a checkerboard pattern, or a phylloid (leaf-like) pattern [3]. Luo et al. recently described a case of segmentally arranged, agminated nevi that exhibit intranevic BrafV600E concordance, suggesting a ‘‘driver’’ mosaic model for the development of these lesions [11].

Our patient differs from previously reported cases in the location and distribution of his nevi. His lesions are on the chest and follow the lines of Blaschko. In a 2005 report by Bragg et al., the presence of an underlying dysplastic nevus syndrome phenotype in 4 of 5 cases of agminated acquired melanocytic nevi raised the possibility that agminated nevi arise as a consequence of postzygotic loss of heterozygosity and, thus, may represent a type 2 segmental manifestation of the atypical mole syndrome phenotype [12]. Whether or not congenital agminated segmental nevi undergo malignant change is unknown, but long-term follow-up is recommended.

References

1. Happle R. Segmental lesions are not always agminated. Arch Dermatol 2002;138(6):838. [PMID:12056974]

2. Sterry W, Christophers E. Quadrant distribution of dysplastic nevus syndrome. Arch Dermatol 1988;124(6):926-9. [PMID:3377521]
3. Happle R. Mosaicism in human skin. Understanding the patterns and mechanisms. Arch Dermatol 1993;129(11):1460-70. [PMID:8239703]
4. Effendy I, Happle R. Linear arrangement of multiple congenital melanocytic nevi. J Am Acad Dermatol 1992;27(5 Pt 2):853- [PMID:1469144]
5. Pariser RJ, Bluemink GG. Acquired linear dermal melanocytosis. Nerve course distribution. Arch Dermatol 1982;118(2):125- [PMID:7059215]
6. Lancer HA, Muhlbauer JE, Sober AJ. Multiple agminated spindle cell nevi: unique clinical presentation and review. J Am Acad Dermatol 1983;8(5):707-11. [PMID:6863631]
7. Brunner M, Vardarman E, Megahed M, Ruzicka T. Congenital agminated segmental naevi. Br J Dermatol 1995;133(2):315-6. [PMID:7547407]
8. Misago N, Takahashi M, Kohda H. Unilateral dysplastic nevi associated with malignant melanoma. J Dermatol 1991;18(11):649-53. [PMID:1800531]
9. Sun BK, Tsao H. X-chromosome inactivation and skin disease. J Invest Dermatol 2008;128(12):2753-9. PMID:18509358
10. Yu X, Nagai H, Nishigori C, Horikawa T. Acquired unilateral melanocytic nevi in otherwise normal skin. Dermatology 2008;217(1):63-5. [PMID:18401177]
11. Luo S, Chaplin AC, Langley RG, Njauw CN, Duncan LM, Miller RA, et al. Agminated segmental nevi demonstrating intranevic concordance of BRAF status. J Invest Dermatol 2011;131(3):788-90. [PMID:21160499]
12. Bragg JW, Swindle L, Halpern AC, Marghoob AA. Agminated acquired melanocytic nevi of the common and dysplastic type. J Am Acad Dermatol 2005;52(1):67-73. [PMID:15627083]