Case Report

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An Unusual Presentation of Multiple Congenital Melanocytic Nevi with a Limb Distribution

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Key Words

Limb distribution

Congenital melanocytic nevus

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Abstract

We report a newborn in whom multiple small congenital melanocytic nevi (MN) were noted on the right side involving the scapular area, shoulder, upper arm and forearm. Such a limb distribution of small congenital MN has never been reported in the literature.

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Introduction

Melanocytic nevus (MN) is a common and benign skin disorder in children and adolescents. It is relatively rare in neonates with a prevalence rate of 1-2% and is classified as congenital if it is detected immediately after birth [1-3]. Congenital MN is most commonly solitary with less than 5% being multiple. Multiple lesions are more commonly associated with a large congenital MN [3, 4]. The presence of small congenital MN with multifocal involvement is rare, except in patients with a giant congenital MN in whom there may be numerous small lesions on the trunk and extremities away from the site of the giant one [5]. Here, we report an unusual case of multiple small congenital MN with a limb distribution.

Case Report

A 3,362-gram male baby was born to a 32-year-old gravida 1, para 1 mother at a gestational age of 38 weeks via vaginal route. The prenatal and postnatal courses were uneventful. The parents were nonconsanguineous.

At birth, many hyperpigmented flat or elevated skin lesions were noted over the right upper limb involving the scapular area, shoulder, upper arm, and forearm with sparing of the hand (fig. 1). No similar skin lesions were found in other areas of the body. The two arms were of similar circumference and length. Those skin lesions were of various sizes, ranging from 0.1 to 3 cm in diameter. They were irregularly shaped macules, papules, patches and plaques with

various colors, looking reddish, light brown, dark brown, grayish or blackish (fig. 2). The borders of the lesions were either sharply demarcated or merging imperceptibly with the surrounding skin. Some lesions showed an irregular border. In some lesions, the color was heterogeneous and color change was noted in some of them during the first days of life.

A skin biopsy was performed on the 6th day of life for fear of malignant change. Two specimens were taken and both showed intra-dermal nevi with the nevus cells extending to the deep dermis and the upper subcutis, which was consistent with the diagnosis of congenital MN. The nevus cells in the upper dermis were heavily pigmented and those in the lower dermis and upper subcutis were nonpigmented and somewhat smaller. No cellular atypia was noted.

The baby was kept under observation and was regularly followed up at our outpatient clinic for 1 year. His development was normal. The skin lesions showed no remarkable change in size, but some reddish nevi gradually turned brown. Further skin biopsy or any surgical intervention was refused.

Discussion

MN is a common skin disorder in children, but the appearance of multiple small congenital MN is rare. The distribution among these multiple small congenital MN does not usually display any particular pattern. A case with a linear arrangement of multiple small congenital MN has been reported [6]. However, the distribution of MN in that case was only a narrow band on the flexor aspect of the right arm. Another report described a case with a quadrant distribution of dysplastic nevus syndrome [7]. The pigmented lesions in this case, however, were acquired and demonstrated a quadrant distribution involving the left arm and the left upper thorax, contrary to the congenital presence of numerous nevi within a limb in our case. Thus, to the best of our knowledge, the limb distribution of multiple small congenital MN in our case seems to be unique.

The cellular or genetic basis of the limb distribution of MN in our case is unknown. It might be related to a somatic mutation oc-

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curing at an early stage of embryogenesis, as Effendy and Happle [6] suggested. Another issue that is not clear at present is the malignant potential of the lesions in this case. Although an increased risk of malignant change of giant congenital MN is well known [8, 9], the risk in relation to the small congenital MN is controversial [9, 10]. Since the presentation of our case is unique, there is no data available to predict the risk of malignant transformation in this type of congenital MN. The variegated color and irregular border of some nevi in this case did raise our doubt about their benign nature. However, biopsies taken from two lesions, one of which showing heterogeneous colors, failed to demonstrate any form of dysplasia or malignancy. Treatment of this case is challenging. Total excision of so many nevi which are closely aggregated in one limb is apparently difficult and impractical, if not impossible. The newly developed, short and ultra-short pulsed lasers such as Q-switched ruby laser or Q-switched Nd:YAG laser might have some therapeutic potential [11-13], although the removal of nevus cells is likely to be incomplete [14].

Fig. 1. A limb distribution of multiple congenital melanocytic nevi.

Fig. 2. The size and color of the nevi are greatly variable. Biopsies were taken from two lesions indicated by the arrowheads.

References

Rhodes AR: Neoplasms: benign neoplasias, hyperplasias and dysplasias of melanocytes; in Fitzpatrick TB, Eisen AZ, Wolff K, Freedberg IM, Austen KF (eds): *Dermatology in General Medicine*. New York, McGraw-Hill, 1993, vol. pp 996-1077.

Osburn K, Schosser RH, Everett VIA: Congenital pigmented and vascular lesions in newborn infants. *J Am Acad Dermatol* 1987;16: 788-792.
Roth ME, Grant-Kels JM: Important melanocytic lesions in childhood and adolescence. *Pediatr Clin N Am* 1991;38:791-809. Hoss DM, Grant-Kels JM: Significant melanocytic lesions in infancy, childhood, and adolescence. *Dermatol Clin* 1986;4:29-44. Reed WB, Becker SV7.
Becker SW Jr, Nickel WR: Giant pigmented nevi, melanoma and leptomeningeal melanocytosis. *Arch Dermatol* 1965;91:100-119.

Effendy I, Happle R: Linear arrangement of multiple congenital melanocytic nevi. *J Am Acad Dermatol* 1992;27:853-854.

Sterry W, Christophers E: Quadrant distribution of dysplastic nevus syndrome. *Arch Dermatol* 1988;124:926-929.

Greeley P\V, Middleton AG, Curtin JW: Incidence of malignancy in giant pigmented nevi. *PlastReconstrSwg* 1965;36:26-37.

Swerdlow AJ, English JSC, Qiao Z: The risk of melanoma in patients with congenital nevi: A cohort study. *J Am Acad Dermatol* 1995;32: 595-599.

10 Rhodes AR. Sober AJ, Day CL, Melski JW, Harrist TJ, Mihm Mc Jr, Fitzpatrick TB: The malignant potential of small congenital nevo-cellular nevi. *J Am Acad Dermatol* 1982;6: 230-241.

Tse Y, Levine VJ, McClain SA, Ashinoff R: The removal of cutaneous pigmented lesions with the Q-switched ruby laser and the Q-switched Nd:YAG laser: A comparative study. *J Dermatol Surg Oncol* 1994;20:795-800.

Taylor CR, Flotte TJ, Gange RW, Anderson RR: Treatment of nevus of Ota by Q-switched ruby laser. *J Am Acad Dermatol* 1994;30: 743-751.

Milgraum SS, Cohen ME, Auletta MJ: Treatment of blue nevi with the Q-switched ruby laser. *J Am Acad Dermatol* 1995;32:307-310.

Waldorf HA, Kauvar AN, Geronemus RG: Treatment of small and medium congenital nevi with the Q-switched ruby laser. *Arch Dermatol* 1996;132:301-304.

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