CASE REPORT

Congenital Dermal Melanocytosis on the Foot: A Case Report and Review of the Literature

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Dermal melanocytosis is a common pigmented skin disease, characterized by an increased number of ectopic melanocytes in the dermis. Rare variants of dermal melanocytosis that do not belong to these four typical groups—nevus of Ota, nevus of Ito, blue nevus, and Mongolian spots—are called dermal melanocyte hamartoma, or congenital dermal melanocytosis (CDM) as it mostly appears from birth. We report a case of CDM on the foot of a young woman with a literature review of previously reported cases of CDM. (*Ann Dermatol* 31(2) 213~216, 2019)

-Keywords-

Congenital, Dermis, Melanocytes, Nevus, blue, Pigmentation disorders

INTRODUCTION

Dermal melanocytosis is a common pigmented skin disease, characterized by an increased number of ectopic melanocytes in the dermis¹. It is usually divided into four main groups: nevus of Ota, nevus of Ito, blue nevus, and Mongolian spots. Rare variants of dermal melanocytosis

that do not belong to these four typical groups are called dermal melanocyte hamartoma, also referred as congenital dermal melanocytosis (CDM) because it mostly appears from birth²⁻⁵. Here, we report a case of CDM on the foot with a literature review of previously reported cases of CDM.

CASE REPORT

A 22-year-old woman presented with an asymptomatic, confluent, and pigmented skin lesion on the right foot dorsum since her birth. No specific changes occurred, including color and texture, except that the lesion enlarged in proportion to the growth of the body. She denied any significant medical problems and family history of pigmented disorders. On physical examination, mottled, confluent, and blue-gray macules and patches were observed on the right foot dorsum (Fig. 1A). We received the patient's consent form about publishing all photographic materials. Dermoscopic examination revealed a steel blue or gray structureless area surrounded by mottled brown globules (Fig. 1B). Skin biopsy demonstrated increased scattered melanocytes in the dermis without nest formation. Spindle and epithelioid melanocytes with melanin pigment were randomly oriented and dispersed among the collagen bundles. Immunohistochemical staining for Melan A showed positive staining for the melanocytes (Fig. 2). Based on the clinical and pathologic findings, the patient was diagnosed with CDM. She has been treated with three sessions of Q-switched Nd:YAG 1,064 nm laser without remarkable improvement.

DISCUSSION

Embryologically, melanocytes are derived from neural crest

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Fig. 1. (A) Mottled, confluent, blue-gray macules surrounding darker patches on the right dorsum of the foot. (B) Steel blue or gray structureless area surrounded by mottled brown globules on dermoscopy.

Fig. 2. (A~C) Increased and scattered pigmented melanocytes without nest formation throughout the dermis. Scattered and elongated melanocytes among collagen bundles without certain orientation (H&E; A: $\times 100$, B: $\times 200$, C: $\times 400$). (D) Melanocytes were positively stained for Melan-A (MART-1, $\times 200$).

cells originating from the ectoderm⁵. Dermal melanocytosis has been suggested to occur due to the arrest of melanocyte migration, resulting in increased number of melanocytes in the dermis⁵. Except for blue nevus, the other types of dermal melanocytosis share similar histopathologic findings and differ only in the concentration and location of melanocytes. The histopathologic finding shows elongated dermal melanocytes scattering between collagens¹. Clinically, the types of dermal melanocytosis, such as nevus of Ota or Ito, are distinguished by their unique location—trigeminal or acromioclavicular distribution⁵. Dermal melanocyte hamartoma is a rare dermal melanocytosis pattern that does not fit into the typical classification of dermal melanocytosis^{2,5}. In the present case, the pigmented lesion distribution differed from the nevus of Ota, nevus of Ito, and Mongolian spots. Additionally,

the lesion was not matched to blue nevus, which shows a high concentration of dermal melanocytes in the dermis¹. Several cases of CDM have been previously reported (Table 1)^{1,4,6-10}. Among the 10 cases including ours, 6 were reported at adults. Female predominance (male:female=2:8) was observed similar to other dermal melanocytosis, such as the nevus of Ota. Segmental or dermatomal distribution was observed in five cases, localized pattern in four cases, and generalized form in one case. Among the cases with a localized pattern, the lesions were confined to the trunk or upper extremities^{3,9,10}. CDM developing as an isolated patch on the foot similar to our case, has not been described. Clinically, CDM usually appears as uniform or mottled gray-blue patches with or without macules. Histopathologic findings revealed scattered melanocytes without nest formation throughout the dermis,

Table 1. Clinical features of previously reported congenital dermal melanocytosis including the present case

Case	Author	Age/sex	Distribution	Morphology	Histology	Prognosis
1	Bashiti et al. ⁶	4 d /female	Generalized distribution (scalp, nose, ears, neck, chest, abdomen, thighs, legs, back and upper extremities)	Blue-gray pigmentation	Numerous melanocytes in the dermis, more numerous in the middle and lower dermis	Decreased substantially at 15 months of age
2	Burkhart and Gohara ²	18 mo /male	Dermatomal distribution (bilateral buttocks and right leg)	Gray-blue patch with several conspicuous macules	Many melanocytes in the upper two thirds of the dermis	Persistent
3	Vélez et al. ⁷	28 yr /female	Segmental distribution (right aspect of the trunk with several thoracic dermatomes affected)	Mottled gray-blue pigmentation with numerous maculae of darker hue	Scattered melanocytes in the dermis (two biopsy specimens: numerous in upper dermis or lower two thirds of the reticular dermis)	Persistent
4	Grézard et al. ⁸	45 yr /female	Dermatomal distribution (two sides of the back from the fourth to the eighth dorsal dermatomes)	Gray-blue pigmentation	Scattered melanocytes within the reticular dermis	Slowly spreading
5	Krishnan et al. ⁹	31 yr /male	Localized (lower left back at the level of the L1~L2)	Speckled blue-brown patch	Not reported	Persistent
6	Kim et al. ¹⁰	21 yr /female	Localized (right arm)	Relatively well-demarcated bluish round patch encircled by larger brownish patch	Scattered melanocytes in the dermis of the central bluish patch and increased basal pigmentation in the epidermis of the surrounding brownish patch	Persistent (removed by surgical excision and 532-nm Q-switched Nd-YAG)
7	Kim et al. ³	5 yr /female	Localized (left palm)	Speckled bluish patch	Scattered melanocytes in the dermis	Persistent
8	Lee et al. ¹	21 mo /female	Segmental distribution (left unilateral aspect of face, neck, chest, shoulder and back)	Uniform deep blue patches with a few dark blue-brown macules	Scattered melanocytes in the dermis, numerous in the upper and middle dermis	Persistent at 21 months of age
9	Pessach et al. ⁴	4 mo /female	Dermatomal distribution (trunk and upper extremities)	Uniform gray-blue patches with several darker blue macules	Numerous melanocytes in the upper dermis	Persistent at 4 months of age
10	Present case	22 yr /female	Localized (right dorsum of the foot)	Mottled and pigmented brownish macules surrounding darker patches	Scattered melanocytes in the dermis	Persistent

which were subtly different from the nevus of Ota or Ito (melanocytes detected mainly in the upper dermis), and Mongolian spot (mostly in the lower dermis)¹. In most cases, the pigmentation did not disappear with age. However, in one case, a 4-day-old neonate had shown generalized blue-gray pigmentation that gradually faded until 15 months of age. Although the case was reported as a generalized dermal melanocytosis, we speculate that the case might be difficult to be differentiated from extrasacral Mongolian spot because it tends to disappear or fade during childhood⁷.

In conclusion, we report a rare case of CDM on the foot with relevant literature review. Further studies are necessary to elucidate the clinical and pathogenetic character-

istics of CDM.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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