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 dermis. 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 birth. 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.
cells originating from the ectoderm. Dermal melanocytosis has been suggested to occur due to the arrest of melanocyte migration, resulting in increased number of melanocytes in the dermis. 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 collagen fibers. Clinically, the types of dermal melanocytosis, such as nevus of Ota or Ito, are distinguished by their unique location—trigeminal or acromioclavicular distribution. Dermal melanocyte hamartoma is a rare dermal melanocytosis pattern that does not fit into the typical classification of dermal melanocytosis. 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 dermis. Several cases of CDM have been previously reported (Table 1). 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. 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. | 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    | Burkhardt and Gohara | 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. | 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. | 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. | 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. | 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. | 5 yr /female | Localized (left palm) | Speckled bluish patch | Scattered melanocytes in the dermis | Persistent |
| 8    | Lee et al. | 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 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 childhood\(^7\).

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 characteristics of CDM.

**CONFLICTS OF INTEREST**

The authors have nothing to disclose.

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