Development of Halo Nevus Around Nevus Spilus as a Central Nevus, and the Concurrent Vitiligo

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Halo nevus is a benign melanocytic nevus that is surrounded by a hypopigmented zone. The most frequent association with halo nevus is vitiligo, and this also appears in nearby regions, as well as at other remote sites. Although the mechanism for developing the depigmentation around nevus spilus is uncertain an immunologic process may be responsible for the finding of inflammatory infiltrates of the upper dermis in the depigmented lesions. We report here on a 13-year-old boy who showed a depigmented zone around a nevus spilus on the right side of his neck with simultaneous vitiligo lesions on the face.

Key Words: Halo nevus, Nevus spilus, Vitiligo

INTRODUCTION

Halo nevus has been termed as leukoderma acquisitum centrifugum or Sutton's nevus; this is a benign melanocytic nevus that's surrounded by a hypopigmented zone, and this type of lesion is found in about 1% of the normal population. Depigmented zones appear mostly around several types of acquired pigmented lesions, such as dermal, junctional and compound nevi, Spitz's nevus and malignant melanomas. In contrast, it is rare that depigmented zones develop around a congenital nevus. Sometimes, vitiligo also appears concurrently in the nearby regions, as in our current case. Vitiligo is frequently associated with a halo nevus (18～26%), and appears in nearby regions as well as at other remote sites. Herein we report on a 13-year-old boy who developed a depigmented zone around a congenital nevus spilus on the right side of his neck at the age of 6, there were simultaneous vitiligo lesions on his face.

CASE REPORT

A 13-year-old boy presented with a 3×4 cm, well demarcated, pale brown colored patch with multiple scattered, dark brown colored speckles, which were 2～3 mm in diameter, on the right side of neck, and these lesions were present since birth. At the age of 6, a white hypopigmented zone had developed around the nevus and it had gradually enlarged to 7 mm in diameter (Fig. 1A). In addition, vitiligo lesions were found in the periorbital, perinasal and perioral regions, simultaneously with the halo nevus (Fig. 1B). There was no personal or family history of autoimmune disorders. The histopathological findings of the hypopigmented lesion revealed a finding of decreased melanin in the basal layer of the epidermis, and the pale brown colored patch lesion revealed elongation of the rete ridges and increased melanin pigmentation in the basal layer of the epidermis (Fig. 2A). A biopsy of the dark brown colored speckles showed increased melanin in the dermoeidermal junction and the upper dermis (Fig. 2B). A finding of mononuclear inflammatory cell infiltrations in the upper part of the
Fig. 1. (A) A well demarcated, pale brown colored patch with multiple, scattered, dark brown speckles on the right side of the neck with a hypopigmented zone around the nevus. (B) In addition, lesions of vitiligo were found in the perioral, perinasal and periorbital regions.

Fig. 2. (A) The histopathology of the pale brown colored patch lesion revealed elongation of the rete ridges and increased melanin pigmentation in the basal layer of the epidermis. (B) Biopsy from the dark brown colored speckles showed increased melanin in the dermoeidermal junction and upper dermis. Mononuclear cells and lymphohistiocytes were infiltrated into the upper part of the dermis in both lesions (A, B) (S-100, ×100).

dermis was seen in the pale brown patch and the dark brown speckles. At present, the nevus persists, with no obvious sign of regression. The depigmented areas are unchanged, and no more vitiligo lesion has appeared.

**DISCUSSION**

Halo nevus rarely occurs in a congenital nevus. The mechanisms leading to the development of halo nevus are still a matter of debate, but the main hypothesis involves a cellular immune response targeting the pigmented cells, and this is perhaps elicited in some cases by the presence of abnormal,
probably premalignant melanocytes either within or at some distance from the halo nevus. Some researchers have postulated that the halo nevus is related to the atypical nuclear changes "commonly" found in the scattered, dark brown speckles in the brown patch of nevus spilus. These atypical nuclear changes act as the initiating factor for an immune response. Once the immune response is initiated, it recognizes common antigens in both the atypical cells and the normal cells. It is the immunologic response that causes the halo phenomenon to cross-react with normal nevus; this would explain the occurrence of multiple halo nevi in some patients.

As for the association between halo nevus and vitiligo, both phenomena are often seen in the same patient at the same time or at the different times. There is controversy about the nature of both conditions as to whether they are merely parts of the same phenomenon or they are caused by processes that are unrelated to each other. Halo nevus and vitiligo share some histopathologic features and probably some pathophysiologic features, but a common entity has not yet been confirmed. In both conditions, a mononuclear cell infiltration is found in close contact with melanocytes, and these melanocytes show ultrastructural signs of degeneration and cellular damage, resulting in depigmentation. About 80% of the mononuclear infiltrate of halo nevi are T cells with a relatively high percentage of CD8+ suppressor/cytotoxic T-cells compared to CD4+ cells. Autoantibodies directed against melanocytes have been found in the serum of patients with halo nevus and vitiligo. Because of these observations, autoimmune mechanisms are believed to be essential for the destruction of the melanocytes in vitiligo as well as those in halo nevus. Nevertheless, halo nevus and vitiligo are distinct entities and they exhibit different pathogenic mechanisms, as has been recently shown. The differences of HLA association within the clinical subtypes of vitiligo support that vitiligo and vitiligo-associated halo nevi may have distinct pathogenic mechanisms. Moreover, even though both conditions clinically present with white appearance, there is a distinct difference under a Wood's light examination: vitiligo demonstrates a characteristic bluish/yellowish fluorescence, whereas the depigmented area of halo nevus does not. We speculate that halo nevus and vitiligo may be the end points of different immunological processes despite that the immune responses to the pigment cell antigens are shared by normal melanocytes and the nevus cells.

REFERENCES