

CASE REPORT

Neurocristic Cutaneous Hamartoma of the Scalp

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Neurocristic cutaneous hamartomas (NCHs) result from aberrant development of the neuromesenchyme. In addition to a dermal melanocytic component, these tumors can contain neuro sustentacular and fibrogenic components. The clinical importance of these lesions includes the potential for misdiagnosis as well as the development of malignant melanomas over a poorly described period of time. We present a rare case of NCH of the scalp in a 1-year-old female. (*Ann Dermatol* 21(4) 396~398, 2009)

-Keywords-

Hamartoma, Neurocristic, Scalp

INTRODUCTION

Neurocristic hamartomas (NCH) result from the aberrant development of cells derived from the neural crest. This abnormality was first described by Tuthill et al.¹ in 1982 and referred to as a pilar neurocristic hamartoma¹⁻³. Neural crest-derived cells show fibrogenic, melanogenic, and neurosustentacular differentiation, and contribute to the formation and development of the local mesenchyme, especially in the cephalic regions³. The importance of the recognition of a NCH is due to the possible transformation to a malignant melanoma of the pigmented lesions over an unpredictable period of time⁴. We report a patient that presented with a blue-gray alopecic plaque on the scalp with the histopathological features of a NCH.

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CASE REPORT

A 1-year-old female presented with an asymptomatic alopecic plaque that was present since birth on the scalp. Physical examination showed a 3×2 cm blue-gray plaque on the scalp with focal alopecia (Fig. 1). We initially suspected a blue nevus and performed a skin biopsy. The biopsy specimen revealed proliferation of nerve-like structures, spindle cells, and melanocytes. High power microscopic views showed fascicles of spindled cells and heavily pigmented cells with dendritic morphology (Fig. 2). Immunohistochemical staining showed diffuse S-100 protein staining within nerve-like structures and pigmented cells that extended to the deep dermis. HMB-45 also diffusely stained the pigmented cells; however, the area with schwannian differentiation did not stain with HMB-45. The stromal cells throughout the lesion were diffusely stained with CD34. NSE stained within the nerve-like structures. Magnetic resonance imaging showed a focal enhancing lesion in the deep soft tissue of the scalp. However, there was no evidence of abnormalities in the cortex of the brain. Because the patient was too young at the time of the initial evaluation surgical excision was planned for later.



Fig. 1. 3×2 cm blue-gray plaque on the scalp.

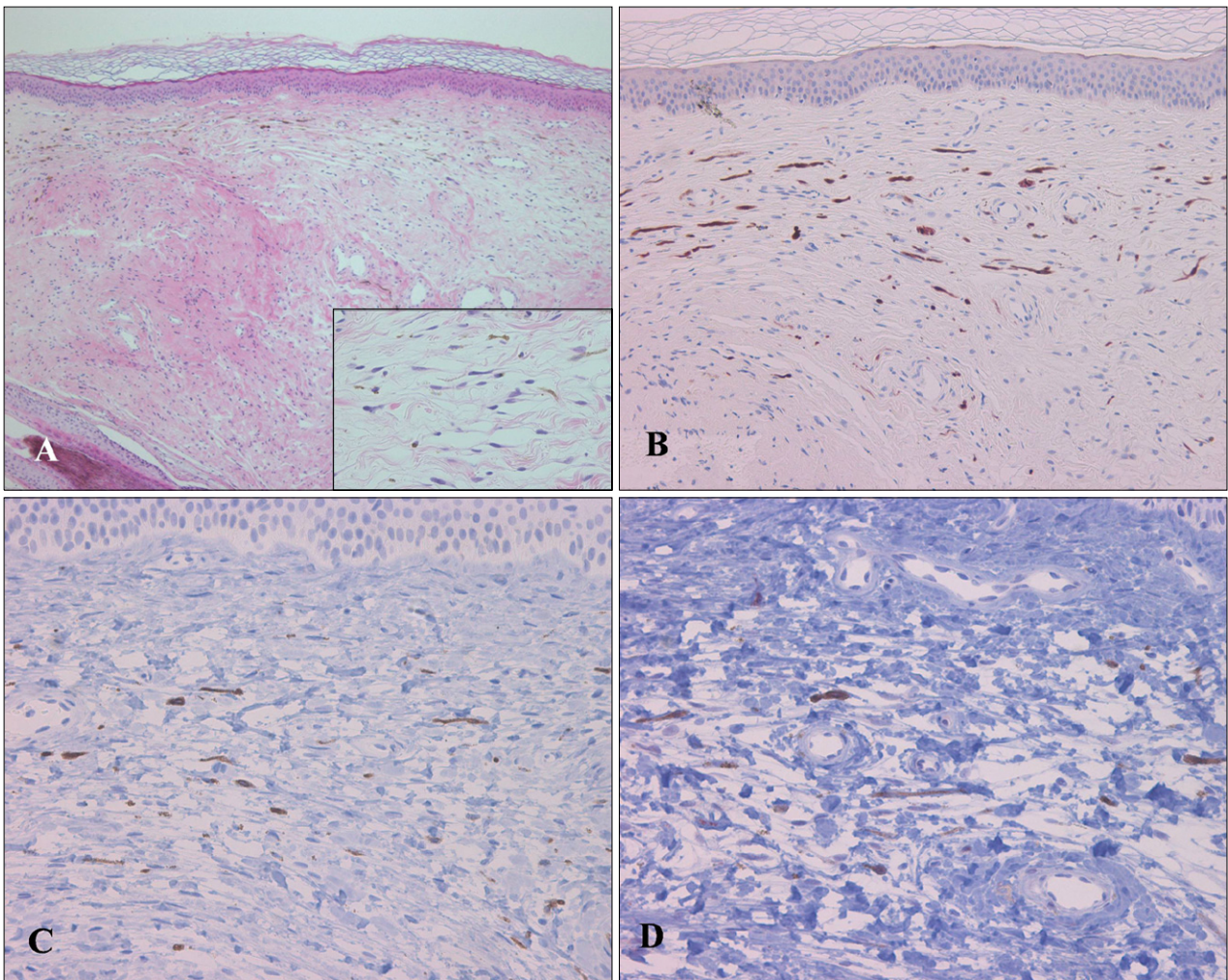


Fig. 2. (A) Proliferation of nerve-like structures, spindle cells, and melanocytes (H&E, $\times 100$) (inset, wavy spindle cells, $\times 400$). (B) Diffuse S-100 protein staining within the nerve-like structures and pigmented cells (Immunohistochemical staining using the ABC method, $\times 200$). (C) HMB-45 diffusely stained the pigmented cells, however, the area with schwannian differentiation did not stain with HMB-45 (Immunohistochemical staining using the ABC method, $\times 400$). (D) NSE staining within the nerve-like structures (Immunohistochemical staining using the ABC method, $\times 400$).

DISCUSSION

Several cases of NCHs have been reported previously^{1,2,4,11}. In the Korean dermatologic literature, a case of dermal melanocytosis on the trunk with features of neurocristic cutaneous hamartoma has been reported¹². The dermal melanocytoses include a variety of pigmented lesions that are formed from the aberrant development of neural crest-derived melanocytes as they migrate through the dermis during embryogenesis⁵. NCH is one type of dermal melanocytosis. NCH is composed of melanocytes that are confined to the dermis and sometimes the subcutaneous tissue with a neural crest-derived Schwann cell component⁵. Clinically and histopathologically, the features of NCHs overlap with other dermal melanocytoses,

especially the blue nevus. The clinical features in this case resembled the plaque-type blue nevus. However, NCHs have a predilection for the scalp with focal alopecia and generally are not found on the trunk. Histologically, NCH lesions are composed of dermal melanocytes and neurocristic structures with schwannian differentiation. NCH lesions usually stain with CD34 and have a decreased number of hair follicles⁵. However, blue nevus lesions usually are not stained with CD34 and the number of hair follicles with a blue nevus are not decreased⁹. Other reported features of NCH include melanocytes distributed around hair follicles, eccrine glands, vessels and nerves, involvement of the subcutaneous tissue, and the presence of tactoid bodies^{1,2,9-11}. In our case, we also noted proliferation of dermal melanocytes and neurocristic structures with sch-

wannian differentiation. The immunohistochemical staining with S-100 protein, HMB-45, CD34, and NSE confirmed the diagnosis.

Although only a few cases have been reported, a significant number of these cases have shown development of a malignant melanocytic component^{2,10}. Melanomas have developed within 1~6 years from the initial diagnosis of NCH. However, melanomas developing from congenital NCH lesions have been identified 15~67 years after birth². Because of the unpredictable development of a malignancy, long-term follow up with cancer surveillance is generally recommended in patients with NCH^{2,13}. Complete surgical resection of the lesion likely prevents the development of a melanoma.

In summary, we describe a rare case of NCH of the scalp with alopecia. Dermatologists should consider NCH in the differential diagnosis of a blue-gray alopecic plaque on the scalp. Early diagnosis is useful for the proper management of NCH including surveillance for the development of a malignant melanoma.

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