A Case of Pigmented Neurofibroma

Hyun Jin Jeon, M.D., Eun Joo Park, M.D., Sung Sik Kim, M.D., Kwang Ho Kim, M.D., Kwang Joong Kim, M.D.

Department of Dermatology, College of Medicine, Hallym University, Anyang, Korea

Pigmented neurofibroma, a variant of neurofibroma, has rarely been observed in patients with or without neurofibromatosis. Pigmented neurofibroma is characterized histologically by the coexistence of scattered melanin-laden cells and benign spindle cells with neural differentiation. We report a case of pigmented neurofibroma in a 22-year-old female patient.
(Ann Dermatol 15(3) 122~124, 2003).

Key Words : Pigmented Neurofibroma

Neurofibromas with melanin-laden pigmented cells are rare, accounting for less than 1% of all neurofibromas[1]. Although it is unclear whether the tumor cells are pigment-synthesizing Schwann cells or whether they originate from a coexisting melanocytic tumor, this tumor again demonstrates the close relationship between peripheral nerve sheath tumors and melanocytic malformations. We describe a case of pigmented neurofibroma in a 22-year-old female patient.

CASE REPORT

A 22-year-old female patient presented hyperpigmented patches with centrally lobulated mass on the back. The patches had been present since birth. A well-circumscribed walnut sized lobulated mass soft in consistency developed in the center of preexisting patches 10 years ago. The family history was unremarkable. Examination of the skin revealed over 20×20 cm sized, hyperpigmented patches with centrally lobulated mass on the left side of her back, brown hyperpigmented macules on the right side of her back, and a few cafe au lait spots on her trunk(Fig. 1). But axillary freckling was not noted. CBC, urinalysis, liver function test, and chest X-ray were within normal limits or negative. Skin biopsy taken from the mass revealed a poorly-delineated tumor that showed the appearance of neurofibroma, consisting of elongated cells having oval nuclei associated with abundant collagen fiber. Some foci of scattered cells containing dark-brown pigments in the cytoplasm were detected in the dermis(Fig. 2-A). Examination of the specimen taken from a hyperpigmented patch showed slightly elongate rete ridges, basal layer hyperpigmentation, and melanin pigmentation in the dermis (Fig. 2-B). Immunohistochemically, the elongated cells were positive for vimentin(Fig. 3-A) and S-100 protein(Fig. 3-B). The pigmented cells were highlighted with the Fontana-Masson stain (Fig. 4). The patient underwent excision of the mass. There was no recurrence of the tumor 6 months postoperatively.

DISCUSSION

Pigmented neurofibromas are very uncommon tumors. These tumors were observed more often during the second and third decades, occasionally later(sixth decade). Females were slightly more commonly affected[2]. The clinical features and histological appearances present diagnostic difficulties. In one review of 17 cases, new criteria were proposed for the pathological diagnosis of pigmented neurofibroma, where the tumor is described as having a strong association with neurofibromatosis[1]. The

Received February 15, 2003
Accepted for publication May 30, 2003
Reprint request to : Hyun Jin Jeon, M.D., Department of Dermatology, College of Medicine, Hallym University, 896, Pyungchon-dong, Dongan-ku, Anyang, Kyunggi-do, 431-070, Korea
Tel. (031)380-3760, Fax: (031)386-3761
E-mail. hjinjeon@hanmail.net

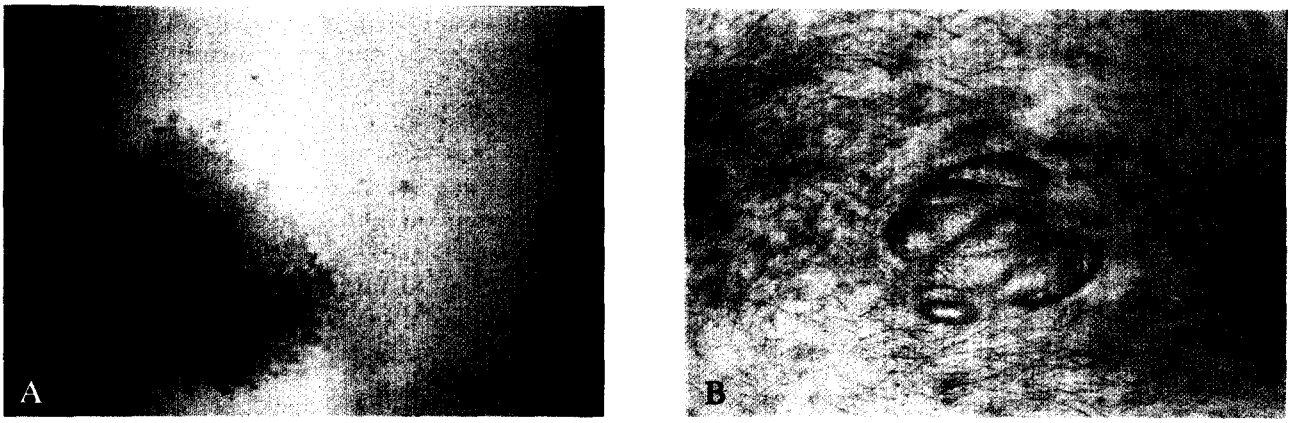

Fig. 1. A) Over 20×20 cm sized, hyperpigmented patches with a centrally lobulated mass on her left back. In addition, there were brown hyperpigmented macules on her right back. B) Close up view.

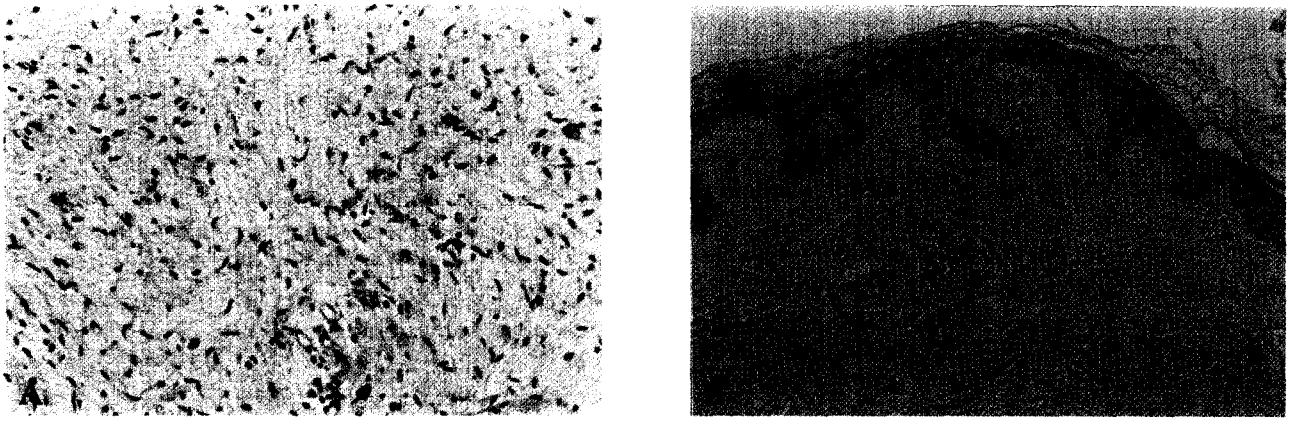

Fig. 2. A) Histopathologic findings of the mass in the back shows elongated cells having oval nuclei associated with collagen fiber. Some foci of scattered cells containing granular dark-brown pigments in the cytoplasm were detected in the dermis(H&E stain, ×200). B) Histopathologic findings of the patch shows slightly elongate rete ridges, basal layer hyperpigmentation, and melanin pigmentation in the dermis(H&E stain, ×100).

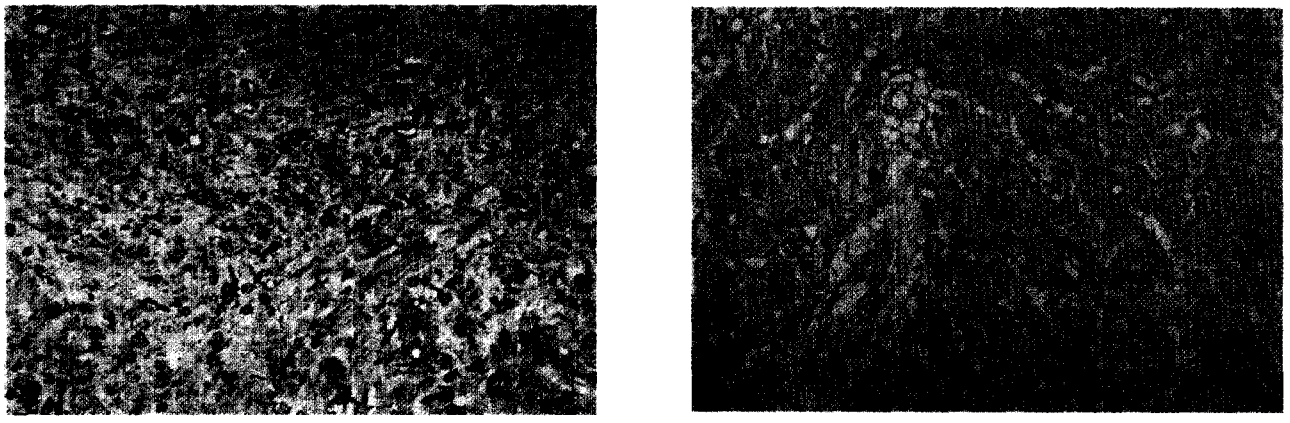

Fig. 3. Immunohistochemical stains show positive reaction to vimentin(A) and S-100 protein(B).

criteria include: (i) the low-power histological appearance typical of diffuse neurofibromas or neurofibromas with combined diffuse and plexiform features; and (ii) immunoreactivity to S-100 protein,

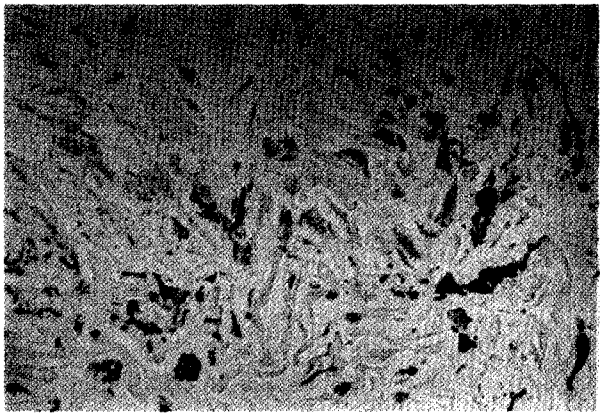

Fig. 4. The pigmented cells were highlighted with the Fontana-Masson stain.

HMB-45 antigen, and CD34. In our case, the diagnosis of pigmented neurofibroma was established by the presence of melanin-laden cells and positive immunoreactivity of the spindle cells to S-100 protein. Storiform neurofibroma(also known as Bednar tumor) has been recognized as a distinct entity that differs from neurofibroma, a variant of dermatofibrosarcoma protuberans(DFSP) with pigmentation[3]. Pigmented DFSP is characterized by spindle cells arranged in a tight storiform pattern and admixed with dendritic cells containing melanin; the S-100 protein is not identified in non-pigmented spindle cells of pigmented DFSP, whereas neurofibromas almost always contain this antigen. Furthermore, pigmented DFSP has an intermediate malignant potential with a tendency to recur locally[4]. Melanotic schwannoma is usually located deeply, such as in the paravertebral region and in the upper alimentary tract. Pigmented cells in melanotic schwannomas are larger and more epithelioid than those seen in pigmented neurofibromas[5]. Cellular blue nevus is an acquired melanocytic tumor that presents typically in young adults in the lower lumbosacral and buttock regions. This entity exhibits a more solid, organoid, or nested growth than a pigmented neurofibroma[6].

In the peripheral nervous system, melanin has also been observed in some schwannomas[7]. In both pigmented neurofibromas and schwannomas, melanosomes in all stages of development suggested the possibility of a melanin synthesis by Schwann cell[8]. The derivation of pigment cells and nerve cells from a similar neural crest stem cell offers an explanation of melanin production by the Schwann cell[9]. Furthermore, Spence et al. in their study of experimental malignant nerve sheath tumors maintained in tissue and organ culture systems demonstrate an apparent melanin producing capability in the neoplastic cells which compose the tumors[10].

The lifetime risk of a malignancy developing in a pigmented neurofibroma is unknown1. All pigmented neurofibromas should be sampled thoroughly to avoid overlooking areas with malignant transformation, and all patients carrying a diagnosis of pigmented neurofibroma should have periodic, long-term follow-up.

We report a case of pigmented neurofibroma developed in the center of preexisting patches.

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