Neurofibroma Associated with Alopecia

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Neurofibromas are benign tumors composed of a complex proliferation of neuromesenchymal tissue with residual nerve fibers. There are several distinct types of neurofibromas: cutaneous, subcutaneous, nodular plexiform, and diffuse plexiform. To our knowledge, none of these have previously been described in association with alopecia in the literature. We present a case of neurofibroma of the scalp which is associated with alopecia.
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Key Words: Alopecia, Neurofibroma

INTRODUCTION

Alopecia has been reported in association with many diseases such as thyroid dysfunction, nutritional deficiency, anemia, lupus erythematosus, and various infections[1]. However, hair loss due to the invasion of tumor cells rarely happens[2]. In this report, we present a case of neurofibroma of the scalp which is associated with alopecia. To our knowledge, alopecia in a lesion of neurofibroma has not previously been described in the Korean or English literature.

CASE REPORT

A 28-year-old male presented with a 20-year history of asymptomatic, subcutaneous mass on his scalp. He discovered hair loss over the mass several years before and complained of its progressive enlargement as the mass had slowly increased. He denied any history of trauma or chronic irritation. Physical examination demonstrated a 6 × 3 cm sized, nontender soft mass on the scalp and a 3 × 2 cm sized alopecic patch over the mass (Fig. 1). Vellus hair was not even observed. There were no other cutaneous lesions or regional lymphadenopathy. Routine laboratory evaluations, including a complete blood count, serum chemistry, and urine analysis showed no abnormalities. Histopathological examination of the lesion showed hair follicles in the upper dermis and a nonencapsulated mass composed of spindle cells with wavy nuclei in the deep dermis. The mass in the dermis displaced hair follicles into the upper dermis rather than surrounded them and the adnexal structure was not seen in the mass (Fig. 2A, 2B). A T2 weighted MRI of the brain demonstrated a hyperintense subcutaneous mass on the right frontoparietal area. (Fig. 2C). A roentgenogram of the scalp showed no gross bony abnormality.

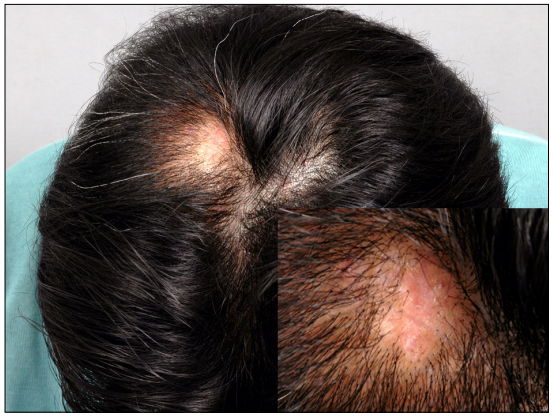

Fig. 1. Asymptomatic soft mass on the scalp and alopecic patch over the mass.

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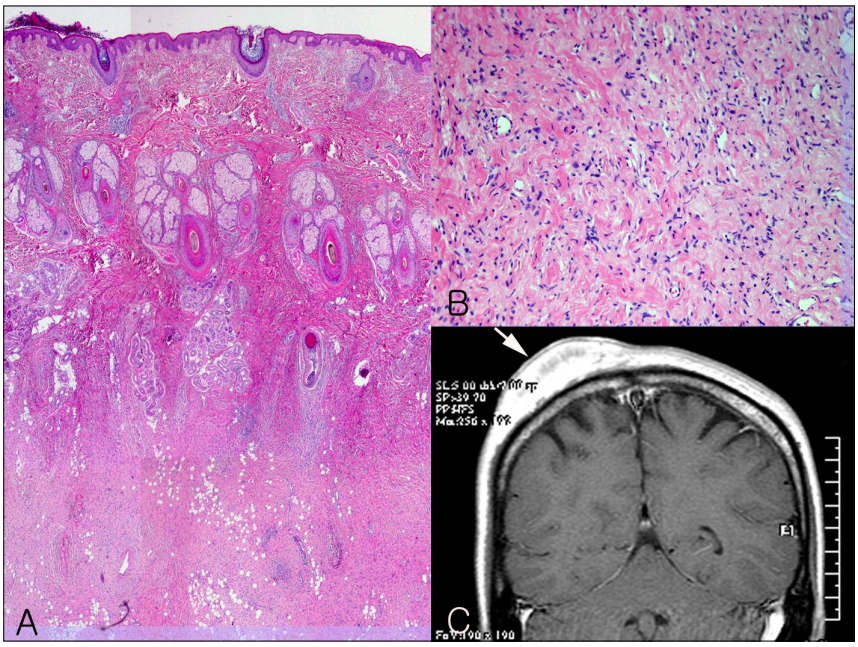

Fig. 2. (A) A nonencapsulated mass in the deep dermis displaced hair follicles into the upper dermis rather than surrounding them (H&E, × 40). (B) The mass in the dermis was composed of spindle cells with wavy nuclei (H&E, × 200). (C) A T2 weighted MRI of the brain demonstrated a hyperintense subcutaneous mass (arrow) on the right frontoparietal area.

DISCUSSION

Neurofibromas may occur as a solitary tumor or as multiple lesions in a segmental or widespread distribution, referred to as neurofibromatosis. Neurofibromas are complex proliferations of the various components of the neuromesenchyme, including Schwann cells, endoneurial fibroblasts, perineural cells, and mast cells[3]. In this complex proliferation, however, the proportion of each cell type varies, providing the broad histologic manifestation of neurofibromas. There are several distinct types of neurofibromas: cutaneous type, subcutaneous type, nodular plexiform type, and diffuse plexiform type[4]. To our knowledge, none of these have previously been described in association with alopecia in the literature to date.

Several tumors can manifest on the scalp with a mass partially or completely devoid of hair. They include basal cell carcinoma, adnexal tumors like syringoma, and cutaneous metastasis of carcinomas that are primary in the mammary glands or internal organs[2]. In these tumors, hair loss happens because of the destruction of the hair follicles by the proliferating neoplastic cells. In a report of occult syringoma associated with alopecia, authors have suggested that stromal changes may inhibit the development of hair follicles[5]. Recently described lipedematous alopecia should also be considered in the differential diagnosis. In cases of lipedematous alopecia, the scalp has a tumid, spongy texture and irregular surface accompanied by a diffuse loss and sparseness of hair[6]. Diffuse hair loss in this rare disease is thought to be due to the thickening of the layer of adipose tissue covering the scalp and to the disturbance of the hair growth cycle.

The mechanism by which neurofibroma may cause hair loss in our case is unclear. Neurofibromas usually surround, but spare rather than displace preexisting structures like skin appendages[7,8]. But in our patient, a skin biopsy showed the neurofibroma in the deep dermis dislodged hair follicles into the upper dermis rather than surrounded them, so this might be the course of alopecia in this case. Vellus hair was not seen in the lesional area.

In our case, both the longstanding mass on the scalp for 20 years and the slow progression of the alopecic process for several years without signs of systemic disease were evidence enough to say that this was a benign process. In the presence of an enlarging alopecic patch, which does not completely fulfill the diagnostic criteria for other forms of alopecia, a neoplascic process including neurofibromas must be suspected and a surgical biopsy must be performed to confirm this.

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