Diffuse neurofibroma of the scalp with alopecia: A case report

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ABSTRACT:

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Diffuse neurofibroma is an uncommon type of neurofibroma, a benign tumor of the peripheral nerve sheath. It normally occurs on the head and neck areas of children or young adults. The diagnosis is based on both clinical and histopathological findings. Characteristics of histopathological findings are infiltrative spindle cell tumor locating in the entire dermis and subcutaneous tissue. We report a case of 34-year-old female who had a diffuse neurofibroma of the scalp with alopecia.

Key words: Diffuse neurofibroma, alopecia

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Introduction

Diffuse neurofibroma is an unusual type of neurofibroma. It normally presents on the head and neck areas of children or young adults. The patients usually present with variable sizes of the soft tissue mass. Only few cases of diffuse neurofibroma on the scalp with alopecia were previously reported. We report a case of 34-year-old female who had diffuse neurofibroma of the scalp with alopecia on the spots.

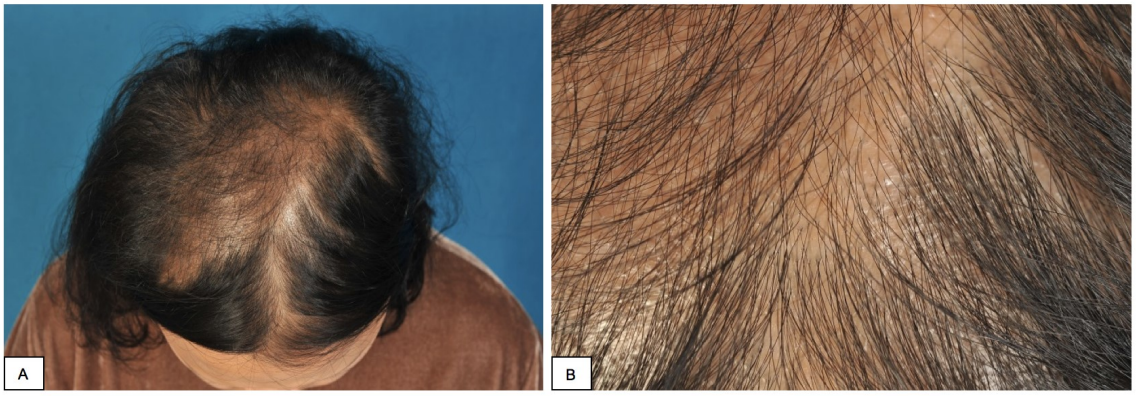

Figure 1 A) Multiple confluent nodules and plaques with ill-defined border on midscalp and crown regions B) Patches of non-scarring alopecia

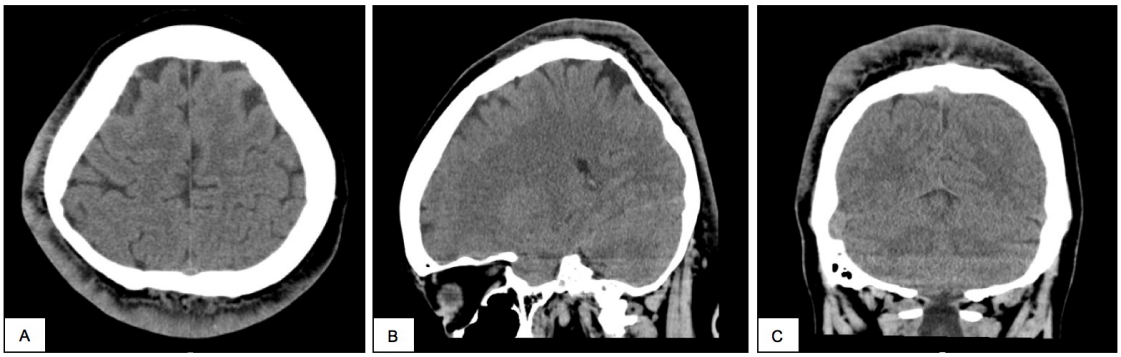

Figure 2 Computer tomography findings. Axial (A), sagittal (B) and coronal view (C) show diffuse thickening of the scalp. No abnormal enhancing intracerebral lesion.

Case report

A 34-year-old Thai female presented with asymptomatic swelling in the midscalp and crown regions for 4 years. She also noticed

concomitant loss of overlying hair. She denied previous illness before the onset of alopecia. No other systemic manifestations including constitutional symptoms were noted.

On physical examination, there were multiple confluent nodules and plaques with ill-defined border measuring 1-8 cm in diameter located in the midscalp and crown regions, irregular patches of non-scarring alopecia on the overlying and negative hair pull test. No other pigmentary lesions were observed (Figure 1). Neurological examination was also unremarkable.

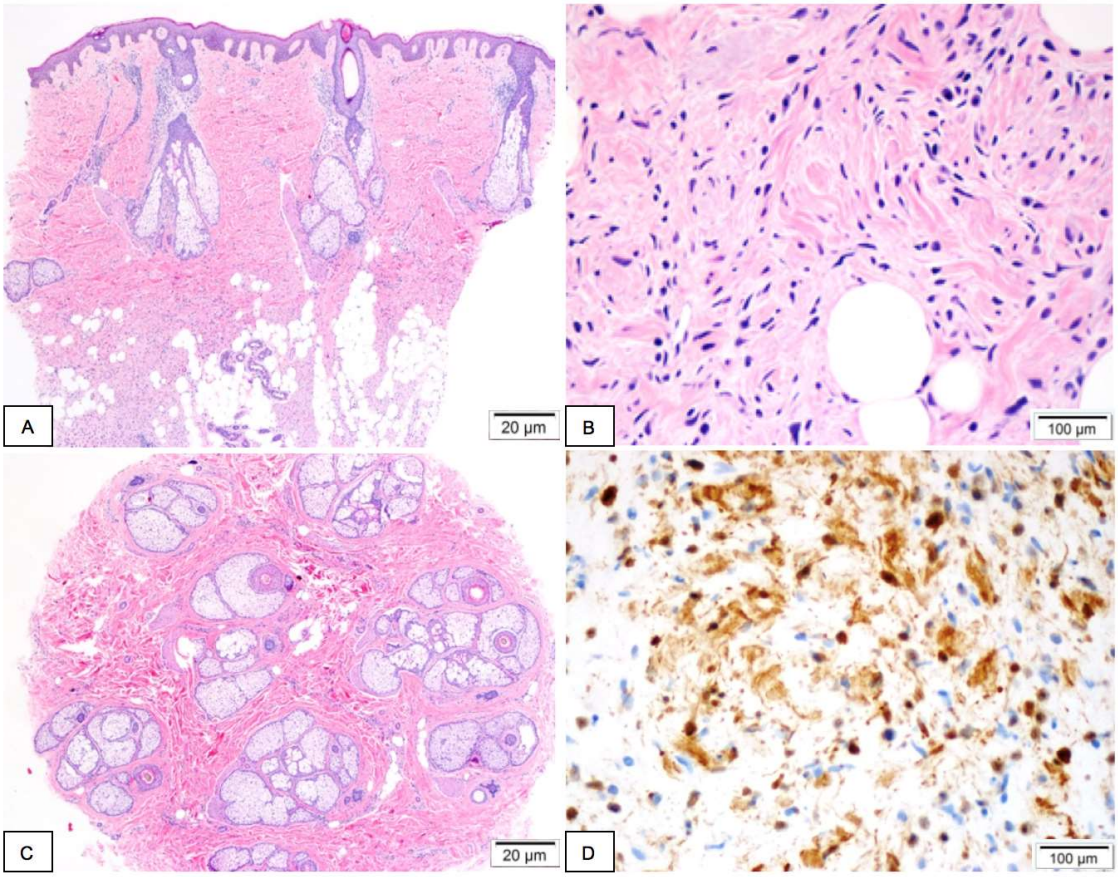

Figure 3 Histopathological examination. A) The vertical section revealed infiltrative lesions in the deep dermis and subcutaneous fat. B) The tumor was composed of spindle cells with elongated wavy nuclei and numerous mast cells. C) The horizontal section revealed non-scarring alopecia associated with increased numbers of telogen and miniaturized follicles. D) S100 immunostaining was positive

Computed tomography (CT) of the brain showed a rather well-defined enhancing hypodensity nodule without calcification or adjacent bony destruction in subcutaneous tissue of vertex. Diffuse thickening of scalp with several small nodules and a few small enhancing nodules scattered in subcutaneous tissue layer of scalps. There is no detectable intracerebral abnormality (Figure 2).

The histopathological finding in the vertical section revealed mild perifollicular lymphocytic infiltrate in the upper dermis. There was a cellular infiltration composed of spindle cells with elongated wavy nuclei in the deep dermis and subcutaneous fat. Numerous mast cells were also observed. The horizontal section demonstrated non-scarring alopecia associated with decreased total number of hair follicles. From approximately 14 intact follicles identified, four were terminal hairs (3 terminal anagen, 1 terminal telogen) and the remaining were miniaturized telogen hairs. S100 immunostaining highlighted many spindle cells (Figure 3). These findings confirmed the diagnosis of diffuse neurofibroma.

The patient did not receive any surgical treatment due to large tumor infiltration. She was advised to observe the progression of tumor and follow up at least annually due to potential malignant transformation. She was treated with topical 2% minoxidil apply at scalp twice daily. At 6-month follow up, she had progressive hair loss but no increase in size of the tumor.

Discussion

Neurofibroma is the most common benign tumors of peripheral nerve sheath which derived from Schwann cell origin. It was classified into 3 subtypes including localized, plexiform and diffuse types. Diffuse neurofibroma is a rare subtype which typically occurs on the head and neck regions.[1-3] However, other anatomical sites such as trunk and extremities have been reported. This variant predominantly manifests in children or young adults. It is related with neurofibromatosis type 1 (NF1, Von Recklinghausen's disease) about 10% of patients.[3]

The patients usually present with variable sizes of the soft tissue mass which grows infiltratively in the dermis and subcutaneous fat but rarely to has intracranial extension and malignant transformation.[4,5] The histopathologic findings are ill-defined infiltrative tumor, composed of spindle cells diffusely in the dermis and subcutaneous tissue. It often spreads along connective tissue septa but preserves the normal structures.[6] Immunostaining for S100 protein is a sensitive marker of benign nerve sheath tumor but non-specific.[7]

Alopecia can be a consequence from tumor on the scalp such as basal cell carcinoma, adnexal tumors and cutaneous metastasis of

carcinoma. The proliferation of neoplastic cells leads to localized destruction of hair follicle.[8] Diffuse neurofibroma of the scalp with alopecia is a rare presentation of neurofibroma and a few cases have been reported. The pathogenesis of alopecia is currently not known.[6] Findings from histopathology revealed tumor occupying the whole dermis resulting in hair follicles moved towards the upper dermis might be the cause of alopecia.[6]

Clinical assessment and histopathological are needed to obtain the diagnosis. One of the differential diagnoses of diffuse neurofibroma is dermatofibrosarcoma protuberans which shows negative of S-100 protein.[9] Imaging studies, including ultrasound, computed tomography scan and magnetic resonance imaging, are helpful to determine the extension of tumor needed to determine the treatment plan.[10]

To date, only few cases of diffuse neurofibroma of the scalp with alopecia have been previously reported in the literature.[2,6,8] All cases showed asymptomatic swelling accompanied by loss of overlying hair on the scalp. In addition, imaging studies reported only local extension in the soft tissue of the scalp without any invasion to the structure underneath. In general, malignant transformation in neurofibroma to malignant peripheral nerve sheath tumor (MPNST) or neurofibrosarcoma occurs in 5-10% of patient with Neurofibromatosis type 1, especially in plexiform or localized intraneural variant.[11] As in prior reports, at least three NF1 patients had MPNST arising from diffuse neurofibroma of the scalp but alopecia was not mentioned.[11-13] Therefore, annual follow-up is recommended due to potential malignant transformation.

The treatment of diffuse neurofibroma can be achieved via radical excision. However, it is challenging to be removed completely by surgical approach. From three cases of diffuse neurofibroma of the scalp with alopecia, there were two cases treated by surgical excision.[2,6,8] However, only one case was followed-up until 6 months after procedure and no recurrence of the tumor was noted.[6] There was another case that did not receive any surgical treatment, but regular examination was performed throughout three years and revealed no enlargement of tumor size.[1]

Summary

We report the patient who presented with asymptomatic swelling at the scalp with alopecia. The diagnosis is diffuse neurofibroma of the scalp with alopecia by clinicopathological correlation. The patient did not receive any surgical treatment due to large tumor infiltration. She was advised to observe the progression of tumor and follow up at least annually due to potential malignant transformation.

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