

CASE REPORT

A Case of Diffuse Neurofibroma of the Scalp

Kwang-Ho Yoo, M.D., Beom-Joon Kim, M.D., Yong-Kwan Rho, M.D., Jin-Woong Lee, M.D., Yeun-Jin Kim, M.D., Myeung-Nam Kim, M.D., Kye-Yong Song, M.D.¹

Departments of Dermatology, ¹Pathology, College of Medicine, Chung-Ang University, Seoul, Korea

A neurofibroma is a benign tumor of the peripheral nerve sheath characterized by proliferation of Schwann cells, perineural cells, and endoneurial fibroblasts. Different types of neurofibromas can be identified, including localized, plexiform, and diffuse types. Neurofibromas can involve any site on the body skin. The diffuse variant is rare and occurs primarily in children and young adults. It involves the skin and subcutaneous tissue in a plaque-like fashion on the head and neck regions. We present a case of a 10-year-old boy who had a diffuse neurofibroma on the scalp. (*Ann Dermatol (Seoul)* 21(1) 46~48, 2009)

-Keywords-

Diffuse neurofibroma, Scalp

INTRODUCTION

Diffuse neurofibroma is an uncommon, but distinct, variety of neurofibroma usually affecting the trunk, head, and neck regions of adolescents and young adults. It is not clear exactly how often diffuse neurofibromas are associated with neurofibromatosis, although it has been suggested that about 10% of patients with diffuse neurofibromas have neurofibromatosis 1 (NF1, von Recklinghausen's disease)^{1,2}.

There are only three previously reported cases of diffuse neurofibroma of the scalp in the Korean literature^{2,3}. We report a case of a diffuse neurofibroma on the scalp of a young boy, which was confirmed by radiologic imaging and histopathologic examination.

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Reprint request to: Myeung Nam Kim, M.D., Department of Dermatology, Chung-Ang University Yongsan Hospital, 65-207, Hangang-ro 3 ga, Yongsan-gu, Seoul 140-757, Korea. Tel: 82-2-748-9573, Fax: 82-2-6359-9573, E-mail: mnkim@cau.ac.kr

CASE REPORT

A 10-year-old boy with a 4-year-long swelling of the scalp presented to the authors. The patient's parents said that the boy had fallen head-first four years prior when he was riding a kickboard. They took their son to the Department of Neurosurgery at that time to have him evaluated for head trauma. A plain x-ray of the patient's skull showed no bony erosion, and the neurosurgeon removed the hematoma through aspiration. According to the parents, the edematous swelling of the scalp had remained since that time.

Physical examination revealed a 7×8 cm sized, swollen mass on the scalp (Fig. 1). There were no changes in the bone around the margins of the swollen mass, nor was there any bruit over the mass. Computed tomography (CT) performed four months before the patient's physical examination showed diffuse thickening and increased density. A provisional diagnosis of lipoma was made, and for further evaluation, magnetic resonance imaging (MRI) and punch biopsy were performed. The MRI showed extensive soft-tissue infiltration with superficial mass-like



Fig. 1. A 7×8 cm swollen mass on the patient's scalp.

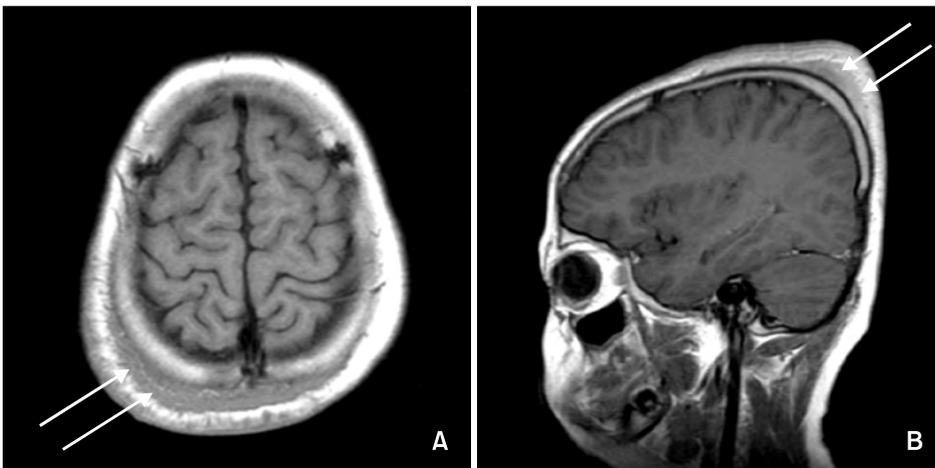


Fig. 2. Radiologic findings. (A) The MRI scan. The axial T1-weighted spin-echo image (TR/TE, 483/12) shows thickening with intermediate signal intensity in the right high frontoparietal scalp. (B) The MRI scan. The post-contrast sagittal T1-weighted spin-echo image shows marked enhancement of the lesion in the right high frontoparietal scalp.

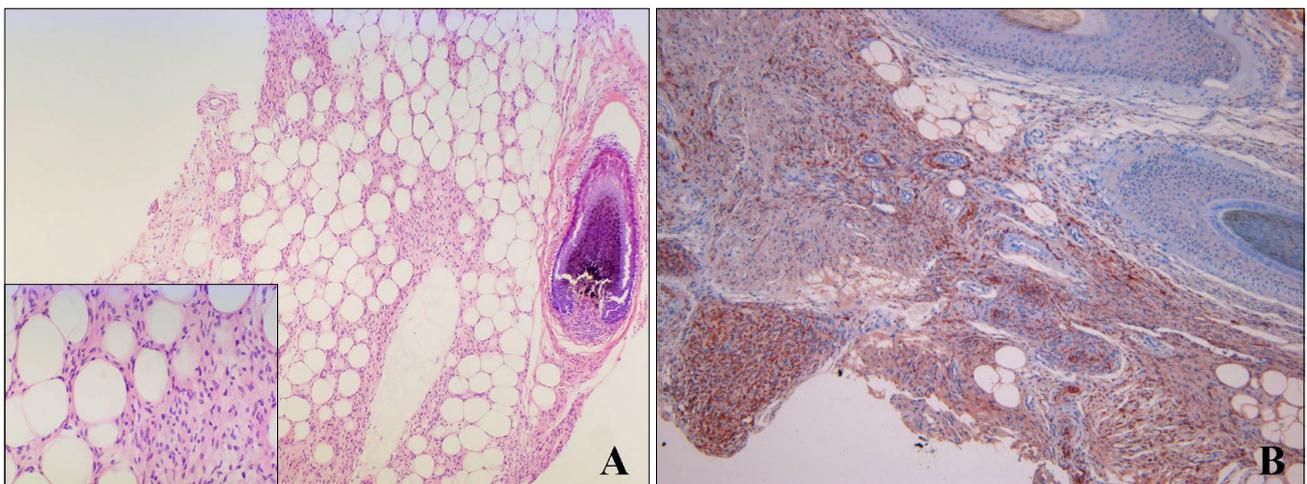


Fig. 3. (A) Histopathologic examination showed fusiform cells with elongated nuclei in a myxoid matrix with wire-like collagen fibers (H&E, $\times 100$, inset, $\times 400$). (B) Immunoperoxidase staining for S-100 protein was positive (S-100, $\times 100$).

lesions and deeper ill-defined infiltration. The lesion exhibited thickening with intermediate signal intensity on axial T1-weighted (T1-W) spin-echo images (Fig. 2A). Post-contrast sagittal T1-W spin-echo images showed marked enhancement of the lesion in the right high frontoparietal scalp (Fig. 2B).

Histopathological examination demonstrated an ill-defined, infiltrative lesion occupying the entire dermis and some of the subcutaneous tissue without destroying the skin appendages. The cells were fusiform, with elongated nuclei, and they were surrounded by a myxoid matrix with wire-like collagen fibers (Fig. 3A). No Meissner bodies were seen. S-100 protein antibody staining was positive (Fig. 3B). Based on these findings, a diagnosis of diffuse neurofibroma was made.

Examination in the Departments of Neurology and Pediatrics revealed no Lisch nodules or other findings sug-

gestive of a diagnosis of neurofibromatosis. The patient's physical and mental development showed no abnormalities, and a diagnosis of Von Recklinghausen's neurofibromatosis could not be established.

Excisional removal of the neurofibroma was recommended, but the patient refused. Follow-up after three years revealed no increase in tumor size.

DISCUSSION

Neurofibromas are benign tumors of the peripheral nerves that have a neuroectodermal origin. The essential cells in neurofibromas are of Schwann-cell origin⁴. Diffuse neurofibroma is an uncommon, but distinctive, form of neurofibroma. It is variably sized, though often large. It is characterized by marked dermal and subcutaneous thickening that most often appears in the trunk or head

and neck regions of adolescents or young adults^{4,5}. This lesion has also been termed "paraneurofibroma" to indicate the extension of the tumor beyond the confines of the perineurium⁴. Intracranial extension of the extracranial variety of this tumor has rarely been reported⁶. At least 10% of these tumors are associated with NF-1, although Megahed⁷ reported that 61% of his 13 patients with diffuse neurofibromas had NF-1. The lower estimate of the incidence of NF-1 is probably due to the fact that the patients with diffuse neurofibromas were young, and young age often precludes a reliable diagnosis of NF-1.

MRI is the investigative tool of choice for this diagnosis because it better defines the anatomic relationships between the neurofibroma and the adjacent tissues, including muscular, vascular, and neural structures⁵. Most MR images of a diffuse neurofibroma reveal a focal, discrete lesion, which may have a typical "target" pattern¹. The neurofibroma may resemble an intramuscular hemangioma, with entrapped fat simulating the lace-like pattern of fat interdigitating between the vascular elements of the hemangioma. Previous neurofibroma studies concluded that these tumors may mimic fat-containing lesions due to the presence of adipocytes intermingling with neurofibroma cells, and due to the entrapment of perineural adipose tissue by diffuse neurofibromas⁸. The MRI findings of our patient, who had a diffuse neurofibroma, supported all these previous observations.

On histopathological examination, the tumor is ill-defined, and it diffusely infiltrates the dermis and subcutaneous tissue. Despite its infiltrative nature, the tumor envelops the normal structures rather than destroying them. It is composed of elongated, spindle-shaped cells with round or fusiform nuclei and eosinophilic cytoplasm within a loose matrix of fine fibrillary collagen. Meissner bodies are characteristic, but are not always present. Neurofibromatous tissues show immunoreactivity with S-100 protein, a sensitive, but non-specific, marker of benign nerve sheath tumors⁹.

Although malignant transformation of neurofibromas has been reported in patients with NF-1, it rarely occurs in solitary diffuse neurofibromas¹⁰. Pain or enlargement of a neurofibroma may herald malignant transformation.

The treatment of large neurofibromas consists of partial or complete surgical excision. Even after complete excision, clinical recurrence may develop because of the infiltrative

growth pattern of the tumor. Because of possible recurrence and the potential development of neurofibromatosis, yearly follow-up is recommended^{4,6}. In the present case, the imaging studies and clinical findings demonstrated an infiltrative pattern of growth, and complete excision of the diffuse neurofibroma was unlikely to achieve success without extensive and potentially mutilating surgery.

In summary, we reported a case of a diffuse neurofibroma occurring on the scalp of a young boy. This is a rare nerve tumor with a characteristic radiological appearance that correlates well with histopathological studies. The reticular appearance is due to a network of abnormal nerves set in a collagenous matrix. It is known to be best demonstrated on MRI, particularly with contrast-enhanced T1-W sequences.

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