Plexiform Neurofibroma of Scalp in a Child: A Case Report
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Summary
Epicranial neurofibromatosis is a rare diagnosis in Neurofibromatosis patients, frequently seen in the occipital region, which can be the cause of bone destruction. Although plexiform neurofibromas are congenital lesions, they have a risk of malignant transformation. Total excision is recommended at the time of diagnosis. They can be found out just after birth and in some cases none of neurofibromatosis signs exist. Physician should differentiate this high vascular tumor from other benign lesions of scalp. In this case report, a 10-year-old boy having progressively enlarging and painful right occipital mass was presented. The patient was operated successfully and the pathology was plexiform neurofibroma.

Key words: Plexiform neurofibroma, neurofibromatosis, scalp

Pediatrisk Hastada Saçlı Deri Yerleşimli Pleksiform Nörofibroma: Olgu Sunumu

Özet

Anahtar Kelimeler: Pleksiform Nörofibroma, nörofibromatosis, saçlı deri

INTRODUCTION
Neurofibromatosis type 1 (NF1) is an autosomal dominant hereditary disorder, characterized by cutaneous hyperpigmentation (Cafe- au- lait) in some specific regions of the body and neurofibromas of the peripheral nervous systems(2,8). Plexiform neurofibroma of the scalp was first described as a manifestation of Von Recklinghausen's Syndrome in a case report in 1906(4,6). Plexiform neurofibromas are among the most common and debilitating complications of NF1. A plexiform neurofibroma consists of a proliferation of cells in the nerve sheath extending across the length of a nerve and involving multiple nerve fascicles(4).
Although plexiform variety of neurofibromas is relatively common in children, they are rarely seen in the scalp\(^6\).

**CASE PRESENTATION**

A 10-year-old child with a right occipital painful mass (progressively enlarging within past 2 or 3 years), causing headache referred to our hospital. Physical examination revealed a soft, non-pulsating, sensitive scalp tumor measuring 6x5x4 cm and multiple areas of hyperpigmentation on his torso. Computed tomography revealed an extracranial lesion and a large temporal arachnoid cyst at the same side. Magnetic resonance imaging (MRI) showed the multilobar lesion with dilated tumor vessels, and incidental Pons hamartoma (Figure 1). During the operation under local anesthesia, a subgaleal necklace shaped mass was started to dissect through to a little bony defect of 0.5 cm diameter and a thin vessel of each bead of the necklace was extending to sigmoid sinus just beneath the bony defect (Figure 2). The tumor was extremely vascular. Due to massive bleeding, we decided to pass through general anesthesia urgently. After gross total excision of the tumour, the bony defect was covered by bone wax to stop bleeding. Patient was taken to the ICU for a few hours for erythrocyte replacement and discharged from the hospital next day. The pathology was neurofibroma without the evidence of malignancy (Figure 3).

**Figure 1:** A: Axial plan computed tomography image revealed subgaleal hypodense solid lesion at the right occipital region (small arrow). B-C: Lesion had low signal on T1 weighted images, and high signal on T2 weighted images. D-F: Post contrast T1 weighed images demonstrated non-homogenous enhancement of the tumour. Axial T2 weighted image (C) also showed hyperintense pontin lesion (hamartoma) that extends to middle cerebellar peduncle (thick arrow). Additionally, there was an arachnoid cyst at the right cranial middle fossa (curved arrow).
Figure 2: A subgaleal necklace-shaped mass which is extremely vascular was started to dissect through to a little bony defect and a thin vessel of each bead of the mass was extending to sigmoid sinus just beneath the bony defect.

Figure 3: The pathology of the mass without the evidence of malignancy (Hematoxylene and Eosin staining, x200)
DISCUSSION

Neurofibromatosis type 1 is an autosomal dominant disease affecting one in 3000 to one in 4000 people, with a great variability of clinical expression. NF1 patients have an increased risk of developing both benign and malignant tumors. The most common tumor is the neurofibroma, a heterogeneous benign nerve sheath tumor, which represents the primary clinical characteristic of neurofibromatosis\(^4,6,7\).

Plexiform neurofibromas commonly occur in persons with NF1, and comprise one of the most common sources of morbidity and cosmetic impairment. Epicranial plexiform neurofibroma is a rare presentation of neurofibromatosis\(^8\). It is a slow growing hypervascular tumor of the occipital region, which can cause skull defect. In 2-29% of the cases, neurofibroma can transform to malignant schwannoma, which can be difficult to diagnose due to the surrounding mass of nonmalignant soft tissue overgrowth\(^5\). Rapid growth, intratumoral hemorrhage are indicators of malign transformation\(^6\).

Plexiform neurofibroma treatment is currently surgical. Extensive growth and the surrounding invasion of the tumor make the complete resection difficult, so recurrence after surgery is common. Rapid growing and invasion to surrounding tissues are often seen during childhood, so early surgery may prevent development of cosmetic and functional problems\(^3\). Benefits of chemotherapy or radiation therapy have not been extensively tested yet, but there is concern of such treatments for slow-growing tumors in patients who are at risk for the development of additional tumors\(^7\).

Growth of plexiform neurofibromas can occur at any time in life. They tend to grow mostly in the early childhood or during the times of hormonal changes like puberty or pregnancy\(^7\).

Epicranial plexiform neurofibroma is a rare presentation of neurofibromatosis. On preoperative imaging and physical examination, they may mimic lipoma of the scalp. Although lipoma is benign and poor bleeding lesions, surgeon should be careful and plexiform neurofibromas should be kept on mind in NF1 patients to make the discrimination of scalp lipoma. In this way we can avoid local anesthesia surgery in this type of a high vascular tumor.

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