Rapid Expansion of Benign Scalp Neurofibroma Caused by Massive Intratumoral Hemorrhage
—Case Report—

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Abstract
A 62-year-old man with neurofibromatosis type 1 presented with rapid growth of a scalp mass. Head computed tomography demonstrated a large extracranial tumor of soft tissue density with massive intratumoral hematoma. Cerebral angiography demonstrated remarkable hypervascularity of the tumor. Preoperative embolization and total removal of the tumor was performed. The tumor contained a large amount of intratumoral hematoma (500 ml). The histological diagnosis was neurofibroma without malignant transformation. Benign scalp neurofibroma showing massive intratumoral hemorrhage is rare. Rapid growth or intratumoral hemorrhage in neurofibroma may be an indicator of malignant transformation.

Key words: intratumoral hemorrhage, malignant transformation, rapid growth, scalp neurofibroma

Introduction
Neurofibromatosis type 1 (NF-1) is an autosomal dominant disorder, characterized by multiple areas of cutaneous hyperpigmentation (café-au-lait spots) and neurofibromas of the peripheral nervous systems. However, scalp neurofibroma is a relatively rare manifestation in patients with NF-1. We report a case of scalp neurofibroma associated with NF-1 which underwent rapid growth caused by massive intratumoral hemorrhage.

Case Report
A 62-year-old man with a history of NF-1 since childhood suddenly heard a small plosive noise from the right occipital region at night. Next morning, he noticed a large scalp mass over the right occipital region. He was referred to our hospital on the same day. Physical examination revealed multiple areas of hyperpigmentation and multiple neurofibromas on his torso and extremities. A large scalp tumor, measuring $10 \times 15 \times 5$ cm, was found on the right occipital region (Fig. 1). The tumor was soft, non-pulsating, multilobar, and had a well-defined margin. Head computed tomography (CT) disclosed a large extracranial tumor of soft tissue density with massive intratumoral hematoma in the right occipital region. Bone window CT revealed thinning of the skull beneath the tumor (Fig. 2). Magnetic resonance imaging showed a huge tumor with the dilated tumor vessels appearing as signal voids. The tumor was markedly enhanced after gadolinium adminis-

Fig. 1 Photographs showing an elastic soft tumor extending from the right occipital to the high cervical region. There was some discoloration on the surface of the tumor.
Scalp Neurofibroma Revealing Massive Intratumoral Hemorrhage

Fig. 2 Computed tomography scans showing a large isodense scalp mass and thinning of the skull beneath the tumor. The tumor contained some high-density spots.

Fig. 3 T₁-weighted magnetic resonance image showing marked enhancement of the tumor capsule only after gadolinium administration (left). T₂-weighted image showing the dilated tumor vessels as signal voids (right).

Fig. 4 Right vertebral angiograms demonstrating a hypervascular mass mainly fed by the dilated muscle branch of the right vertebral artery and the right posterior meningeal artery.

Fig. 5 Photomicrograph of the surgical specimen demonstrating proliferation of spindle cells with collagen fiber, and Wagner-Meissner bodies in some areas. HE stain, ×150.

Discussion

Scalp neurofibroma is relatively rare, although association with evidence of NF-1 is strongly indicative of the diagnosis. Scalp neurofibroma is characterized by very slow progression, occipital location, association with skull defect or thinning, and salient hypervascularity. However, scalp neurofibroma with massive intratumoral hemorrhage is extremely rare, despite the tumor hypervascularity, with only four reported cases of scalp neurofibroma including the present case.

The etiologies of intratumoral hemorrhage are considered to be endothelial proliferation with vascular obliteration, vessel compression and distortion.
due to rapid tumor growth, vessel necrosis, and invasion of vessel walls by the tumor.\textsuperscript{2,3,6} However, these mechanisms occur in malignant tumors, and do not apply to benign neurofibroma. Scalp neurofibromas are located in the superficial layer overlaying the skull, and so are exposed to external forces such as slight trauma, compression, and distortion. The etiology of intratumoral hemorrhage in our case of neurofibroma may have involved external force.

Malignant transformation of neurofibroma to malignant schwannoma occurs in 2–29\% of patients with NF-1.\textsuperscript{1,7} Furthermore, two of the four reported cases of scalp neurofibroma with massive intratumoral hemorrhage demonstrated malignant transformation.\textsuperscript{4} Rapid growth or massive intratumoral hemorrhage in neurofibroma may indicate malignant transformation.

References


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