Giant Epicranial Neurofibroma

—Case Report—

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Abstract

The authors describe a case of giant epicranial neurofibroma unassociated with von Recklinghausen's neurofibromatosis. The tumor, which was situated in the occipital region, gradually enlarged over a 14-year period, while the two skull defects beneath it increased in size over a period of 3 years. On angiography the tumor exhibited marked hypervascularity. It was fed mainly by the bilateral occipital arteries and drained into the transverse sinus. After its embolization and total excision the pathological diagnosis was plexiform neurofibroma. This and two similar reported cases have features in common with those of von Recklinghausen's neurofibromatosis.

Key words: neurofibroma, skull defect, von Recklinghausen's neurofibromatosis

Introduction

An epicranial plexiform neurofibroma in a patient with no signs of von Recklinghausen's neurofibromatosis was reported by Scott¹²⁾ in 1966, and Ohaegbulam¹⁰⁾ presented a second such case in 1977. There were some clinical similarities between those two cases and seven cases of von Recklinghausen's neurofibromatosis have been reported since 1906.^{3,5,9,14,18)} It has not been established, however, whether or not the two disorders should be classified together. We treated a patient with an epicranial plexiform neurofibroma but no clinical signs of von Recklinghausen's neurofibromatosis. Of particular interest were the progression of the bone defects and the tumor's hypervascularity, as evidenced by angiography and computed tomography (CT).

Case Report

The mother of the patient had first noticed a small swelling in the occipital region when the patient was 5 or 6 years old. It gradually increased in size, but was not painful. He first visited our clinic in March of 1975 with a large, elastic-hard tumor in the right occipital region but no neurological deficit. Plain

skull x-ray revealed thinning of the occipital bone on the right side. Right retrograde brachial angiography displayed no tumor staining but demonstrated displacement of the external carotid artery. By this time the tumor had grown very large, but according to his family there were no signs of von Recklinghausen's disease.

In July, 1978, at the age of 19 years, he was hospitalized for resection of the tumor. On examination, the tumor (26×19 cm) was found to extend from the right occipital to the upper right cervical region, and was elastic and hard but not tender (Fig. 1). Its border was fairly definite. A bruit synchronous with the pulse was audible in the right mastoid region. The skin was somewhat discolored and telangiectatic, but its appearance did not suggest hemangioma. Blood tests were all normal.

Plain skull x-ray and tomography showed two skull defects in the right side of the occipital bone. One had an osteolytic, irregular margin and the other, which had manifested as bone thinning 3 years previously, had an osteosclerotic margin (Fig. 2). A CT scan showed a large extracranial mass of soft tissue density, markedly enhanced except at its center. There were no abnormal findings in the posterior fossa (Fig. 3). Transfemoral selective angiog-

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Fig. 1 Photograph taken on admission, showing a large mass extending from the right occipital to the high cervical region. There was some telangiectatic discoloration.

Fig. 2 A plain skull x-ray demonstrating the bone thinning and defect. The thinned portion (*arrows*), which had been noted 3 years previously, has a hyperostotic margin. This skull x-ray reveals a new bone defect (*arrowheads*) at the transverse sinus with an osteolytic, irregular margin.

Fig. 3 CT scan revealing an isodense mass in the occipital region. The tumor is markedly enhanced except at its center. No abnormalities are seen in the posterior fossa.

raphy performed 4 days before surgery showed the tumor to be fed mainly by bilateral occipital arteries and partially by the ascending cervical artery and the posterior meningeal branches of both vertebral arteries. There was intense tumor staining, but it was heterogeneous, with early venous filling in the capillary phase. Drainage was into the dural vein through the skull defect and into the transverse sinus in the venous phase (Fig. 4). Selective embolization was performed with gelfoam inserted into the occipital and ascending cervical arteries. The tumor stain subsequently disappeared (Fig. 5).

On July 19 he underwent surgery. The tumor, approached through a midline skin incision, lay beneath the muscles and was beef red and well encapsulated. Fibrous tissue connected the tumor to the dura at the site of the skull defect, and the tumor was easily freed from the dura and the transverse sinus. The margin of the bone defect was covered by connective tissue. The tumor was situated between the atlas and axis. The tumor was totally removed, with blood loss of about 300 ml. The histological diagnosis was plexiform neurofibroma (Fig. 6). The only postoperative symptom was hypoesthesia in the right occipital region. He was discharged on August 8 and has had no recurrence of the tumor in 1985.

Discussion

Epicranial neurofibroma is rare; only two other cases not associated with von Recklinghausen's neurofibromatosis,^{10,12)} and only seven cases associated with von Recklinghausen's disease,^{3,5,9,14,18)} have been reported (Table 1). In all cases, the tumors were

Fig. 4 Selective angiography demonstrating the hypervascular nature of the tumor, which was fed primarily by the bilateral occipital arteries. Tumor staining was heterogeneous, showing a mixture of fine and dense areas, early filling veins, and venous pooling. The feeding arteries were hypertrophic and the venous return was to the transverse sinus. *upper*: Right occipital angiograms, *lower*: left occipital angiograms. 1: occipital artery, 2: tumor staining, 3: dural vein within bone defect, 4: transverse sinus, 5: internal jugular vein.

Fig. 5 Embolization of the bilateral occipital arteries resulted in complete disappearance of tumor staining. *left*: Left external carotid angiogram, *right*: right external carotid angiogram.

Fig. 6 Photomicrograph of the plexiform neurofibroma. HE stain, $\times 200$.

noticed in infancy or childhood and grew very slowly. Most were located in the occipital region. A bruit was audible and the overlying skin appeared hyperemic. It is well known that these clinical features are also seen in some cases of von Recklinghausen's neurofibromatosis.^{3,9,14,18)}

A defect or thinning of the skull bone was described in seven of the 10 cases of epicranial neurofibroma. The tumor had not invaded the bone or the dura. In Scott's patient¹²⁾ and in our own, the transverse sinus under the bone defect was covered with fibrous connective tissue with a smooth margin, and the tumor was encapsulated by the fibrous tissue and was easily separated from it. On the other hand, in Ohaegbulam's case¹⁰⁾ the tumor adhered tightly to the transverse sinus, necessitating intracapsular removal. Nakasu's case⁹⁾ was of 30 years' duration, and the tumor had an osteosclerotic margin. In our case, thinning of the occipital bone was noted when the patient was 16 years old, and during the next 3 years the bone defect extended and the margin became sclerotic. There was another large osteolytic bone defect with an irregular margin on the right side of the lambdoid suture.

Progressive enlargement of multiple skull defects cannot be explained simply in terms of compression by the tumor. Such lesions may grow in synchrony with the tumor.⁴⁾ In other words, their course strongly suggests mesodermal dysplasia of the type seen in von Recklinghausen's neurofibromatosis; such bone defects have, in fact, been reported in many cases,^{2,6,7,17)} and exhibit a tendency to arise at the lambdoid suture, regardless of whether or not a neurofibroma is present.^{2,3,11)} The reported changes have been destructive, atrophic, or hyperostotic.¹⁾ Usually there was a clear osteolytic margin,^{4,11,13)} although in some cases the margin was osteoscle-

Table 1 Reported cases of epicranial plexiform neurofibroma with and without evidence of von Recklinghausen's neurofibromatosis

Case No.	Author (Year)	Patient's age, sex	Age at onset	Location of tumor	Skull defect	Hypervascularity	Bruit	Von Recklinghausen's disease	
								Family history	Patient
1	Scott (1966) ¹²⁾	27, F	13 yrs	O	thinning	?	?	—	—
2	Ohaegbulam (1977) ¹⁰⁾	13, F	1 mo	rt. O	+	?	—	—	—
3	Present case	19, M	6 yrs	rt. O	+	+	+	—	—
4	Helmholz and Cushing (1906) ⁵⁾	19, M	4 yrs	lt. P	?	?	?	—	+
5	Selby and Pereira (1973) ¹⁴⁾	?	?	O	thinning	?	+	?	+
6	"	?	?	O	thinning	?	+	?	+
7	Garcia-Uria <i>et al.</i> (1979) ³⁾	19, M	2 mos	O	—	+	+	+	+
8	"	14, F	3 mos	lt. O	+	?	?	?*	+
9	Nakasu <i>et al.</i> (1981) ⁹⁾	42, M	4–5 yrs	rt. O	+	+	—	—*	+
10	Uegaki <i>et al.</i> (1983) ¹⁸⁾	17, F	8 yrs	rt. P	—	+	?	—	+

*The family had only cafe-au-lait spots. ?: not reported. O indicates occipital; P, parietal.

rotic.⁹⁾

CT of our patient showed a large, isodense mass in the occipital epicranium. It was strongly and homogeneously enhanced, except for an oval, smoothly margined area in the center. An intratumoral cyst containing degenerated tissue was found in the removed tumor specimen. Nearly identical CT findings were described by Nakasu *et al.*⁹⁾ and Uegaki *et al.*¹⁸⁾

The angiographic findings of extracranial neurinoma have been reported in detail. Generally, neurinoma displays neither hypervascularity nor hypertrophy of the feeding arteries.¹⁵⁾ However, Moscow and Newton⁸⁾ reported four cases of hypervascular neurinoma of the neck. These tumors were encircled by tortuous, irregular feeding arteries and tumor staining was nonhomogeneous, showing mixed hypo- and hypervascular areas that were indistinguishable from those of meningioma. Sterner *et al.*¹⁶⁾ suggested that hypervascular neurinomas of the peripheral nerves are malignant. However, Moscow found no correlation between the degree of vascularity and malignancy of intracranial neurinomas.⁸⁾ Among the 10 reported cases of epicranial neurofibroma, some were hypervascular but none showed histological evidence of malignancy.

Our case had many hypertrophic feeding arteries, and this finding was also reported by both Garcia-Uria *et al.*³⁾ and Nakasu *et al.*⁹⁾ From the capillary to the venous phase, tumor staining was hetero-

geneous, demonstrating mixed fine and dense areas and early venous filling. These patterns have not been seen in cases of meningioma. In our case, the less vascular area at the center of the tumor may have corresponded to the low-density area on the postcontrast CT scan. The venous return from the tumor was a very interesting finding. Drainage was mainly into the dural vein *via* the dural attachment of the tumor just within the boundary of the skull defect, and into the transverse sinus. This type of venous return may also be observed in meningiomas that are attached to the dura. In most reported cases, separation of the tumor from the dura was difficult, especially in the region of the transverse sinus, and in one case¹⁰⁾ it was impossible. However, in Nakasu's⁹⁾ and our cases the tumor was easily removed because it had not invaded the dura. For such large tumors preoperative angiography is necessary to know the tumor circulation. We embolized our patient's tumor, which also served to minimize bleeding during surgery.

According to the available data, solitary epicranial neurofibromas have many clinical and radiological similarities to von Recklinghausen's neurofibromatosis. Therefore, we feel that the two may be classified together.

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