Massive Plexiform Neurofibroma of the Occipital Scalp
Case Report

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We are reporting this case of a scalp tumor because of its unusual size, shape, location, and histological structure.

Case Report

M. B., age 27 years, was referred to the Neurosurgical Service of Temple University Hospital on September 18, 1949, by Dr. W. E. Burnett. The patient had first noticed a lump in the back of her scalp in 1935. Removal was advised but she feared surgery. As the tumor enlarged, she groomed her hair over it. The lesion eventually assumed the shape of a “large firm loaf of Vienna bread” and she finally agreed to operation. There was no family history of a similar condition or lesions suggesting neurofibromatosis.

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Peared to be attempts at Wagner-Meissner tactile corpuscle formation (Fig. 3). Diagnosis: plexiform neurofibroma of the occipital scalp (Dr. A. Peale). The slides were reviewed by Dr. Bernard J. Alpers of Jefferson Medical College and Dr. Arthur P. Stout of the College of Physicians and Surgeons, Columbia University, who concurred.

Postoperative Course. The patient made an uneventful recovery. Fig. 4 shows the appearance of the incision at discharge. She was seen June 3, 1965, 16 years following surgery. There was no recurrence of the tumor. She felt well and her head and neck movements were normal. The neurological examination was negative. There was no evidence of any skin lesions or generalized neurofibromatosis.

Discussion

Although an identical case could not be found in the literature, the observations of Helmholtz and Cushing concerning lesions of similar pathology involving the forehead and temporal scalp are of interest. In 1906, these authors reported a case of a 19-year-old youth, who had generalized neurofibromatosis. He had a large, soft scalp mass over the left side of the face and head. They dissected from this mass a small neurofibroma involving the auriculo-temporal nerve and named the lesion "Elephantiasis Nervorum of the scalp." They also briefly described similar cases from the literature occurring mainly in young males, some of whom had generalized neurofibromatosis. They noted that these lesions had a predilection for the "trigeminal field and temporal region in particular." None of the lesions were located in the occipital area. They stated that although these large isolated growths are almost invariably accompanied by other manifestations of von Recklinghausen's disease, the tumors themselves have come to be designated by a variable terminology which they list.

Since my patient did not have evidence of generalized neurofibromatosis nor a family history of it, and since her tumor did not occur in the scalp area reported by Helmholtz and Cushing's case or in their review, I have used the term "massive plexiform neurofibroma of the occipital scalp" to describe her lesion.

Summary

We have reported a case of massive plexiform neurofibroma of the occipital scalp, successfully removed. There has been no recurrence 15 years after surgery, nor has there been any evidence of generalized neurofibromatosis.

Reference


Fig. 3. Plexiform neurofibroma (see description in text). H&E: X255.

Fig. 4. Incision at discharge.