"Congenital" plexiform neurofibroma of the occipital scalp

Case report

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An occipital scalp tumor first noted in a Nigerian girl during the first month of her life gradually increased in size and 13 years later measured 20 x 16 x 17 cm. A skull film revealed a 2 x 4-cm skull defect underneath the tumor over the lambdoid suture. Complete excision of the tumor was achieved although it was adherent to the dura of the transverse sinus. It was histologically confirmed to be plexiform neurofibroma. There were no other signs of von Recklinghausen's neurofibromatosis.

KEY WORDS - plexiform neurofibroma - scalp - congenital skull defect - transverse sinus

PLEXIFORM neurofibromas have been reported to occur at different sites in the body, including the limbs, face, trunk, stomach, urinary bladder, and the scalp. The case reported here is of a congenital plexiform neurofibroma of the occipital scalp that was adherent to the dura of the transverse sinus through a skull defect.

Case Report

This 13-year-old girl presented with a swelling on the right side of the back of the head. The mother had noticed a small lump in the right occipital scalp during the first month of life. The lump did not upset the baby in any obvious way and normal milestones of development were maintained. The lump had slowly increased in size over 13 years and in the 12 months before admission she had become very conscious of its embarrassing size. She had also complained of dull intermittent right-sided headaches. There were no signs suggestive of raised intracranial pressure, and no other complaints.

Examination. On examination, we found a healthy looking girl, who had normal mental functions. A large oblong swelling of the scalp measuring 20 x 16 x 17 cm occupied the right parietooccipital area. The margins of the lump were ill-defined in some areas. The surface was irregular with nodular areas that were tender and firm. The overlying skin was normal and was attached to the nodular areas of the lump (Fig. 1). There were no pulsations or bruits. No enlarged lymph nodes were noted in the head and neck. Cranial nerves, motor, sensory, and reflex functions were intact and there were no abnormal cerebellar signs. The patient had no other evidence of von Recklinghausen's disease.

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FIG. 1. Lateral view of the patient showing the occipital mass.

Tuberculin test and Wassermann reaction were negative. Hemoglobin was 12.3 gm%. The white blood cell count was 6900/cu mm with the differential count as follows: neutrophils 24%, eosinophils 40%, lymphocytes 30%, monocytes 6%. Sedimentation rate was 30 mm/hr. Genotype was AS. Serum calcium was 10 mg%, and serum alkaline phosphatase was 46 IU. A skull film revealed a 2 × 4-cm bone defect in the right occipital bone with smooth margins (Fig. 2).

FIG. 2. Lateral view of the skull film showing the oblong bone defect in the right occipital bone near the lambdoid suture.

Operation. A biopsy specimen was diagnosed as plexiform neurofibroma, and 1 week later, the tumor was excised. During the operation, the quasi-encapsulated tumor was mobilized toward the periphery, where it behaved like a carpet of grass; its outermost margins were ill-defined. In the occipital area, a skull defect was defined measuring about 2 × 4 cm. Here the capsule of the tumor was adherent to the right transverse venous sinus. Intracapsular removal was performed very cautiously to safeguard the sinus. No other areas of the skull were eroded by the tumor.

Postoperative Course. The postoperative course was uneventful. Histological examination confirmed that it was a plexiform neurofibroma. There was no evidence of malignancy (Fig. 3). The patient was followed up for 18 months and there was no palpable tumor.

Discussion

Only a few cases of plexiform neurofibroma of the scalp have been reported in the neurosurgical literature. In 1906, Helmholtz and Cushing6 reported a case of what they called elephantiasis nervorum of the scalp, a manifestation of von Recklinghausen's disease. Rakshit, et al.,8 in reviewing 265 nerve-sheath tumors, noted that 22 of them were plexiform neurofibromas. Only two of these lesions were located in the scalp.

The etiology of plexiform neurofibroma is obscure. Waggener18 has suggested that plexiform neurofibroma is a tumor of nerve-sheath origin. Gruszkiewicz, et al.,4 reported a case of midline plexiform neurofibroma of the lumbar region in a 5-year-old boy. On the basis of the hirsutism of the overlying skin and because of its midline position, they presumed that it was a congenital malformation. Of the 22 patients reported by Rakshit, et al.,8 three were seen before the age of 9 years, and 11 cases occurred between the ages of 10 and 19 years. The case reported here was noticed during the first month of life, strongly suggesting a congenital lesion.

The presence of a skull defect near the lambdoid suture through which the tumor was adherent to the dura of the transverse sinus was another interesting finding in our case. Several authors3,6,7,9-12 have described the same phenomenon, although in most cases, unlike ours, the patients showed other
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Fig. 3. Photomicrograph of the plexiform neurofibroma. H & E, × 40.

features of neurofibromatosis. Selby and Pereira,\textsuperscript{12} in reviewing intracranial neoplasms in Malaysia, mentioned two cases with large plexiform neuromas of the occipital nerve with erosion of the skull overlying the transverse sinus and associated with a bruit from the impingement of the sinus against the bone edges. Scott\textsuperscript{11} described a massive occipital scalp plexiform neurofibroma in a 13-year-old girl in whom the skull film showed thinning of the bone. The significance of these skull defects has been interpreted differently by several authors. Hunt and Pugh,\textsuperscript{9} Joffe,\textsuperscript{7} Davidson,\textsuperscript{8} and Saha, et al.,\textsuperscript{10} reported cases in which skull defects occurred in the absence of neurofibroma masses. Hunt and Pugh\textsuperscript{8} argued that these defects may be due to an inherent mesodermal dysplasia of the skull.

Although the lesion we are reporting was not associated with clinical neurofibromatosis, its early onset suggested that it was congenital. Like most of the other cases already quoted, the skull defect was related to the lambdoid suture but unlike them the tumor was adherent to the dura of the transverse sinus. It would appear that the theory of inherent mesodermal dysplasia rather than bone erosion could explain the skull defect in this case. It is, however, not clear why the skull defects show a predilection for the lambdoid suture area.

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References


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