Intracerebral schwannoma

Case report

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A rare case of schwannoma occurring in the parietal lobe of an 8-year-old boy has been described. While the precise origin of such an unusually situated schwannoma remains uncertain, it appears most likely that the origin was from ectopic or perivascular nerve plexus Schwann cells within the parietal lobe.

Key Words - brain tumor - schwannoma - cerebrum - perivascular neuroplexus

Solitary schwannomas comprise approximately 8% of primary intracranial tumors. The tumor is rare in patients under the age of 15 years. In children, as in adults, the acoustic nerve is the site of predilection. Reports in the literature consist mainly of a few isolated cases. Only an occasional case of schwannoma of other cranial nerves can be found in the literature. These are almost invariably in adults and are believed to occur virtually exclusively in neurofibromatosis. Intracranial schwannoma involving cranial nerves other than the acoustic are unusual in childhood. Intramedullary schwannomas of the spinal cord have been occasionally reported.

In 1966 Gibson, et al., 7 published what appears to be the first recorded case of an intracerebral Schwann cell tumor. This developed in the temporal lobe of a 6-year-old boy.

The present report records a parietal intracerebral Schwann cell tumor occurring in an 8-year-old white boy. A search of the literature has failed to reveal any further records of intracerebral Schwann cell tumors.

Case Report

This 8-year-old boy had had episodic frontal headache and vomiting for 2 years, and a grand mal seizure a few days before admission.

Examination. Neurological examination was normal except for slight congestion of the optic discs. The electroencephalogram (EEG) was markedly abnormal over the right cerebral hemisphere. Radiological examination of the skull revealed suture diastasis and small foci of calcification in the central portion of the right parietal area. Isotope brain scan revealed increased activity in the right parietal region, and right common carotid angiography a 5 mm dislocation of midline vessels to the left, with more marked displacement posteriorly than anteriorly. There was diminished filling of small vessels throughout the parietal lobe. No tumor vessels were identified on standard films or subtraction studies. The findings suggested the diagnosis of a parietal lobe glioma.

Operation. A parietal craniotomy revealed tense brain, with swollen flattened gyri in the parietal region. A firm mass was encountered by ventricular needle at a depth of 1 cm.
cm in the posterior parietal region, above the area of the angular gyrus. Through a small cortical incision, a circumscribed tough tumor was found at a depth of 1 cm. Tumor was dissected out around a clearly defined periphery, with suction used to decompress the softer nonvascular central portions of the mass. The lobular mass extended into the depths of the parietooccipital white matter. Two small blood vessels which entered the tumor through the medial parietal gyri were clipped and coagulated. There was no penetration of the medial parietal and occipital cortex by the tumor and no attachment to, or vascular supply from, the falx. There was no connection with the adjacent vascular sinuses or other major blood vessels in the area.

**Pathological Examination.** The tumor measured approximately $4.5 \times 5.0$ cm, with a tan and white cut surface, and contained soft and firm areas with tissue arranged in nodules separated by shiny membrane. The outer surface was nodular. There were no gross areas of necrosis, cystic change, or hemorrhage.

Microscopically, there was a generally uniform arrangement of compactly arranged bipolar spindle cells with centrally situated long oval nuclei, largely oriented with long axes parallel to each other and with extensive regular palisade arrangement of cells (Figs. 1, 2). There were abundant reticulin and collagen fibrils and scattered thin-walled blood vessels. Mitoses were absent. The cell type and arrangement were typical of Schwannoma, predominantly of Antoni type A tissue.

**Postoperative Course.** The patient made an uneventful recovery. Follow-up over the next 3 years indicated that he was back at school and doing well, with no symptoms or neurological deficit.

**Discussion**

This case shows considerable similarity with the case reported by Gibson, *et al.*\(^7\) including age and mode of presentation. In each case, the tumor was large and lay entirely within the cerebrum, in the earlier case within the temporal lobe and in our case within the parietal lobe. In each case, the tumor was relatively large and circumscribed, with typical gross and microscopic appearances of schwannoma. Gibson, *et al.*\(^7\) offered further confirmation of the nature of the tumor by electron microscopy, which demonstrated a definitive basement membrane covering the predominant cell type. This served to exclude meningioma of deep
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pial origin, piloid astrocytoma complicating occult neurofibromatosis, and leiomyoma of blood vessel origin.

The site of origin of intracerebral schwannoma remains uncertain. Theories of origin of intramedullary schwannomas of the spinal cord have been discussed by McCormick. It has been quite widely accepted that spinal cord tumors have originated from proliferation of Schwann cells in perivascular nerve plexuses of the spinal cord. Clark examined 36 randomly selected spinal cords and identified well-developed nerve plexuses, sparsely distributed on the intrinsic ramifications of the anterior spinal arterial branches, in 14 cases. The architecture was identical with that of peripheral nerves, and a perineurial sheath was continuous with the adventitia of the arterial branches. Even in the larger plexuses there were relatively few nerve fibers, and much of the bulk resulted from apparent hyperplasia of these sheath elements. It was questioned whether the prominence of these interstitial elements represented a reactive hyperplasia, developmental dysplasia of hamartomatous type, or ectopia.

Similar foci of Schwann cells within the substance of the spinal cord have been noted, and, together with the perivascular variety, have been termed "Schwannosis." A perivascular lesion of Schwannosis has been observed in the pons, close to the floor of the fourth ventricle.

An alternative source of intracerebral schwannoma is suggested by Russell and Rubinstein, who noted the resemblance of mesodermal pial cells to neuroectodermal Schwann cells and suggested that pial cells can sometimes undergo conversion to Schwann cells.

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References


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