Case Reports

Intracerebral Schwannoma

Report of a Case*

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Solitary Schwannomas form about 8 per cent of all primary intracranial tumors but almost all occur at middle age. The tumor is extremely rare in patients under the age of 15 years and reports of these are limited mainly to a few isolated cases or those cited briefly in reviews of large series of intracranial tumors within this age group.

In children, as in adults, the acoustic nerve is the site of predilection. Craig, Dodge and Ross found 2 cases under 15 years in their series of 410 verified unilateral acoustic neuromas. Björksten described a single case and Erickson, Sorensen and McGavran referred to a 13-year-old boy in a series of 140 acoustic neuromas. The literature reviewed in these publications provided 7 other cases under 15 years. A few cases of intracranial Schwannoma on the trigeminal nerve have been described in adults but none in children. The only example of intracranial Schwannoma in the surgical and autopsy files of our hospital involved the trigeminal nerve in a 14-year-old girl with neurofibromatosis. All other intracranial nerves are extremely rare sites of solitary tumor even in the adult population. A recent case report described a facial nerve Schwanna which probably arose at 12 years of age. The only report of involvement of the hypoglossal nerve was in a child with von Recklinghausen's syndrome.

Intracranial Schwannomas originate from the distal segment of the nerve root after it has penetrated the pia mater. This report describes a large temporal lobe tumor with features indistinguishable from those of a Schwannoma, in a 6-year-old boy. A search of the literature failed to reveal any record of an intracerebral Schwann cell tumor. The interest of this case centres on the possible sites of origin of the tumor.

Case Report

History. A 6-year-old coloured boy was admitted to The Hospital for Sick Children, Toronto, for investigation of "grand mal" seizures of about one year's duration. They were described as right-sided convulsions lasting 1 to 2 minutes, preceded by an aura and followed by a period of drowsiness or sleep. The seizures occurred about 3 or 4 times daily but were reduced to once or twice per day with anticonvulsant therapy. His past history included asthmatic attacks from the age of 18 months. He had been investigated previously and found to be allergic to a wide range of antigens. Neurological abnormalities were absent on all previous general physical examinations.

Examination. He was well developed and healthy. No abnormality was detected on physical examination initially, but one week later neurological examination demonstrated slight weakness of the right arm and leg. Cranial nerve function was unimpaired. Routine blood and urine examinations were normal. Levels of blood sugar (both fasting and randomly drawn), of serum calcium, phosphorus and magnesium and blood urea nitrogen were all within normal ranges. Lumbar puncture revealed cerebrospinal fluid (CSF) to be at normal pressure, with a few red blood cells. Levels of CSF sugar and protein were 68 mg. per cent and 201.5 mg. per cent respectively. The electroencephalogram was abnormal with a discharging focus at the left posterior parieto-temporal region. Slight bony asymmetry was demonstrated in skull roentgenograms but intracranial calcification was absent. Air encephalogram (Fig. 1) demonstrated displacement of the ventricular system to the

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Fig. 1. Air encephalogram to show lateral displacement of the ventricular system without dilatation.
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right without evidence of dilatation. The left temporal horn was displaced medially and inferiorly and its posterior outline was irregular, giving the impression of a large space-occupying lesion in the left hemisphere, probably in the temporo-parietal area. A review of the plain skull films showed separation of the cranial sutures with increased size of the left temporal fossa and thinning of bone in this region.

Operation. Craniotomy was performed through a left temporo-parietal flap 14 months after the onset of symptoms. The dura was reflected and a tumor palpated in the superior portion of the mid-temporal lobe. The arachnoid was cauterized and the pia opened. At a depth of 1 cm, a firm tumor, well demarcated from surrounding brain, was encountered. The lesion was outlined by blunt dissection; the exact medial limit of the mass was uncertain, but the ventricle was not entered in the course of removal. During the operation the diagnosis of Schwannoma was suggested by Cryostat sections but after careful search no relation to cranial nerves could be demonstrated. The tumor was completely excised.

Postoperative Course. The postoperative course was characterized by recurrent pyrexia, headache and some nuchal rigidity. CSF contained up to 250 white blood cells per cu. mm., predominantly neutrophils, but was sterile. This aseptic meningitis suggested that the tumor had bordered on the lateral ventricle. The patient was continued on anticonvulsant therapy (Dilantin and phenobarbital) and only one small seizure was observed up to the time of his discharge 4 weeks after operation. He was readmitted 10 days later with a slightly tender, fluctuant swelling beneath the scalp at the posterior aspect of the craniotomy flap. The incision was soundly healed. The swelling was aspirated on 3 occasions; the first yielded about 20 ml. of purulent blood-stained fluid from which nonpyrogenic Staphylococci were cultured. Bacitracin was injected. Subsequent aspirates contained only blood and were sterile on culture. He was discharged 9 days after re-admission.

Follow-up. Six months after operation the patient had had no seizures or symptoms of any type. The scar had healed and the boy was normal neurologically. He was still on sodium dilantin 30 mg. t.i.d.

Pathology. The tumor was a well circumscribed mass (8×6×4.5 cm., 105 gm.). Although brain thinly covered a small area of its surface, most of it was completely free from adjacent normal tissues. Its outer surface was nodular (Fig. 2a). The mass was firm throughout and the cut surface was composed of strikingly convoluted broad bands of grey-white tumor separated by thin bands of softer red-grey tissue (Fig. 2b). Hemorrhage, necrosis or cystic change were not grossly evident.

Microscopically, interwoven bundles of long bipolar spindle cells were arranged uniformly in whorled masses (Fig. 3a). Nuclei were centrally situated, and either oval, or elongated with homogeneous chromatin content and inconspicuous nucleoli. Mitoses were absent. In many areas the nuclei were aligned in palisade arrangement. Reticulin and collagen fibrils were abundant. Elastic fibres could not be demonstrated using Verhoeff’s elastic van Gieson or Weigert’s resorcin-fuchsin methods. Blood vessels were thin-walled and normal. This pattern of the tumor was characteristic of type A tissue of Antoni described in Schwannomas. Similarly, the pattern of linear areas between the main bands of tumor was an open one of widely separated cells with indistinct cell boundaries separated by intervening eosinophilic matrix associated with Antoni type B tissue (Fig. 3b). Secondary xanthomatous change, necrosis or hemorrhage were all absent.

Electron Microscopy. Fragments of tumor were fixed in 1 per cent osmic acid immediately after surgical removal and embedded in Epon 812. Sections cut at 0.5 micron thickness, stained with 1 per cent toluidine blue,
were screened by light microscopy for identification of suitable portions for electron microscopy. In an area containing palisaded nuclei (Fig. 4 and inset) cells with abundant cytoplasm contained multiple complex interdigitated processes. A well-defined basement membrane covered most cell surfaces (Fig. 5). This basement membrane was fibrillar and microfibrils were numerous. Groups of cells were widely separated by abundant collagen but no fibroblasts were identified. Fig. 5 also illustrates an appearance resembling elastic fibers but none could definitely be identified by electron microscope using phosphotungstic acid stain. Collagen fibrils

Fig. 2(b). Gross appearance of the tumor showing cut surface.

Fig. 3(a). Photomicrograph of tumor showing Antoni type A tissue. Hematoxylin and cosin; X 250.
Fig. 3(b). Photomicrograph of tumor showing Antoni type B tissue. Hematoxylin and cosin; X 250.
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were numerous and occasional non-medullated nerve fibers surrounded by collagen were identified. The peculiar extracellular fibrils or collagen bodies first described in Schwannoma by Luse were absent despite an exhaustive search for them. Occasional non-medullated nerve fibers surrounded by collagen were identified. Myelinated nerve fibers were absent.

Discussion

In spite of the highly characteristic light microscopic picture of Schwannoma, the site of the tumor induced natural caution in accepting the diagnosis originally based on the Cryostat sections. A reasonable alternate diagnosis was a meningioma of intraventricular type or of deep pial origin in which the microscopic pattern of whorled fibroblasts and nuclear palisading are frequent. The medial extent of this tumor was not defined but the lateral ventricle had not been opened during surgical dissection and removal of the mass. In addition, the electron microscopy demonstration of a definitive basement membrane covering the predominant cell-type appeared to exclude a diagnosis of meningioma since meningocytes normally have no basement membranes. Tissue cultures were not made from this tumor although they have been reported as a diagnostic aid and both Schwannoma and meningioma have distinctive growth patterns and morphology in culture.

Cytologic detail provided by electron microscopy was consistent with Schwann cell proliferation. The large amounts of collagen in this tumor were presumably produced by Schwann cells since no fibroblasts were identified. The type of extracellular fibril described by Luse, and later...
shown by her to be probably fibrous long-spacing collagen\textsuperscript{10} was confirmed by Raimondi in 8 other cases of acoustic Schwannoma.\textsuperscript{14} These collagen bodies were described in a peripheral nerve lesion in von Recklinghausen's disease and 4 brain tumours in a review by Ramsey\textsuperscript{15} who commented that the mucopoly-saccharide abnormalities that have been found in some brain tumours may produce the collagen variants. These bodies have not been identified so far in any other tissue, normal or neoplastic, and their absence in the present case might be related to the unusual site of the tumor rather than its nature and is therefore of particular interest.

The possibility that this growth might represent an unusual case of von Recklinghausen's disease in the central nervous system was considered. There was no family history or physical stigmata of this condition in the boy but "formes frustes" are known to exist. In cases where the central nervous system is involved, the peripheral nervous system is little involved or totally spared. Even in classic neurofibromatosis, peripheral manifestations are not obvious within the first decade of life. Some glial tumors in this condition were originally described as "central neurinomas" or "diffuse Schwannomas"\textsuperscript{16} but are now usually interpreted as piloid astrocytomas. The microscopic features of the tumor reported here do not favour the latter diagnosis because intracytoplasmic glial fibrils were absent. A further point against central neurofibromatosis is the absence of multiple lesions (although the boy has been followed only a short time).

A more remote alternative diagnosis was leiomyoma arising from smooth muscle of a blood vessel. This diagnosis was easily excluded by electron microscopy since characteristic myofilaments and fusiform densities were absent.

Accepting the diagnosis of Schwannoma, there remains the problem of the site of origin. A nerve root is normally ensheathed by Schwann cells only as it perforates the pia mater and all the evidence in the present case points to the lesion being situated deeply within the left temporal lobe.

A few cases of intramedullary Schwann cell tumor of the spinal cord have been reported.\textsuperscript{11} Nerve plexuses accompanying intrinsic arteries of the cord have been suggested as a possible source of such tumors.\textsuperscript{14} Such collections of Schwann cells closely related to, and often sheathing, spinal blood vessels have been observed in a number of chronic diseases of the cord including neoplasms and syringomyelia.\textsuperscript{18} Similar foci of Schwann cells

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**Fig. 5.** Electron micrograph showing the prominent basement membrane (arrows) between the cell membrane (CM) and collagen fibers (C). Uranyl acetate, $\times39,000$. 

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have been found within the substance of the spinal cord and together with the perivascular variety have been termed Schwannosis, possibly hamartomatous in nature. A perivascular lesion of Schwannosis has been observed in the pons close to the floor of the 4th ventricle. Abnormal foci of Schwannosis occurring in the cerebrum have not been reported. There are no theoretical grounds why it should not occur.

Pia mater accompanies blood vessels into the brain. Russell and Rubinstein noted the resemblance of mesodermal pial cells to neuroectodermal Schwann cells. They believed that pial cells can sometimes undergo conversion to Schwann cells. Schwann cells have been described within the cerebrospinal nervous tissue in relation to lipomatous of the leptomeninges. They were almost always superficial but in our patient adipose tissue or other connective tissue elements were not related to the tumor.

The most likely source of origin in this case was a focus of Schwannosis, whatever its nature might be, in which neoplastic transformation had occurred.

**Summary**

We have described a Schwannoma in the temporal lobe of a 6-year-old boy and presented evidence to support the nature of the tumor, based on light and electron microscopy. We have discussed the possible cytogenesis causing this tumor to arise in such an unusual site, and have suggested a focus of Schwannosis as the most likely possibility.

**References**

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