Suprachiasmal and Intraventricular Meningioma in a Four-Year-Old Child

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The present communication concerns a 4-year-old boy from whom two unusual meningiomas have been removed successfully in a span of 2 years. The first tumor, which has been reported previously, was removed from the suprachiasmal region. The second one was found in the left lateral ventricle.

Report of Case

History. In January 1960, when the patient was 2 years old, a large meningioma, weighing 250 gm. (Fig. 1) was removed from the suprachiasmal area. The tumor was located on the dorsal surface of the optic chiasma. The flattened optic nerves, chiasm and optic tracts were markedly stretched downward and bowing, resembling a small concave hammock in which the giant neoplasm nested. The meningioma was not attached to the basilar dura mater, the tuberculum sellae, the olfactory grooves, or the falx. Its blood supply was derived from the ethmoidal arteries.

The postoperative course was uneventful. He made a satisfactory recovery. Aside from a bilateral anosmia, all neurological findings were essentially normal including fundi and vision.

Course. Sixteen months later seizures occurred, each attack lasting 2 to 5 minutes. During these seizures the child was stuporous but not unconscious, with eyes fixed, pupils dilated, pale, at times perspiring and hyper-ventilating without tonic or clonic phase. The seizures were largely controlled by Dilantin and Mysoline.

Since the operation the child was noted to be restless and hyperactive. He seemed to be everywhere at once, running and hopping, shifting from one activity to another, but short of concentration. He was constantly exploring and manipulating objects in the environment, not remaining with any one thing for any great length of time but moving about from one thing to another as if driven from within. Fits of anger were easily provoked. In tantrum, he rolled on the floor. At meals he was running in and out for a mouthful at a time. For this hyperkinetic behavior he was treated with Dexedrine 5 mg. t.i.d. The hyperactivity was reduced considerably.

Repeated electroencephalography showed diffuse bilateral delta activity, especially prominent in the left frontotemporal region. Spike potentials often were intermixed with the slow activity.

In December 1961, examination disclosed a bilateral weakness of the 6th cranial nerve, more marked on the left. There was no papilledema. His fundi and vision were normal. Measurement of the head was normal for his age. All other neurological findings were essentially negative.

A radiograph of the skull showed the cranial sutures to be normal. A globular shadow of calcification was visualized extending from the midline to the left (Fig. 2). Radiograms taken in 1960 before the first operation showed no evidence of such calcification (Fig. 3). On Jan. 8, 1962, at the age of 4, the patient was readmitted for diagnostic studies. Lumbar puncture demonstrated normal pressure and clear cerebrospinal fluid containing a total of 80 mg. protein per cent. A percutaneous left carotid arteriogram disclosed a bowing anterior cerebral artery embracing the shadow of calcification without a lateral shift. The venous phase demonstrated an enlarged internal cerebral vein draining the tumor. A pneumoencephalogram performed with 30 cc. of air revealed a dilated right lateral ventricle; the left was not filled. The brain had fully re-expanded to fill the frontotemporal cranial cavity in the region where the enormous meningioma had been evacuated previously (Fig. 2).

Operation. The left craniectomy window was reopened through the previous frontal longitudinal incision in the scalp. Cortical adhesions were noted here and there, but otherwise the cortex appeared normal. A transcortical incision was made, and the ventricle was entered. The tumor was embedded in the floor and lateral wall of the ventricle. It covered the foramen of Monro and was greyish white in color, well encapsulated, and the size of a large olive. It was avascular, hard and gritty to the scalp. Its scanty blood supply was derived from the ventral wall of the lateral ventricle and a vein was noted draining the tumor to the terminal vein which was clipped. The tumor was removed completely. It was not connected to the choroid plexus, which was left intact.

Postoperatively an aseptic ventriculitis developed which gradually subsided in 8 weeks. He was last seen 7 months after the second operation. He remained hyperactive. The convergence squint had disappeared. He showed no evidence of such calcification (Fig. 3).
FIG. 2. (Left) Radiogram of skull. Note midline spherical calcification. (Right) Pneumoencephalogram showing calcified tumor in anterior portion of opposite lateral ventricle. Note full expansion of frontal lobes 2 years after removal of giant suprachiasmal meningioma; silver clips in ethmoidal area left during first operation; moderately dilated right lateral ventricle.

had occasional seizures of the diencephalic type. A complete psychometric study was done and he scored 120 in an I.Q. test* (Stanford-Binet).

The intraventricular tumor weighed 24 gm. and was a psammomatous meningioma (Fig. 4).

Discussion

There are three clinical features of this case that warrant discussion: 1) The multiplicity and lack of dural attachment of the meningiomas, 2) the absence of visual disturbance in spite of pressure upon the optic chiasm, and 3) the hyperkinetic behavior.

A review of the literature revealed 88 cases of intracranial meningiomas in children and adolescents. One of the particular features of meningiomas in these age groups is the frequent lack of dural attachment. This case probably constitutes the largest meningioma ever found and removed from the suprachiasmal region. It had no dural attachment.

Meningiomas of the lateral ventricles are rare. There have been less than 100 cases recorded in the literature. Among these, fewer than half a score were found in children and adolescents.


* Psychological evaluation done by A. M. Vasquez and Noreen Papatheodorou at The Exceptional Children's Foundation, Los Angeles, California.
Their occurrence in children under 5 years of age to our knowledge has not been recorded previously. The intraventricular meningioma of the present case was largely asymptomatic. Its small size merited a clear demonstration of its independence from the choroid plexus and its origin in the velum interpositum. Cushing and Eisenhardt differentiated choroidal meningiomas from those arising from the velum interpositum, yet they pointed out that the distinction of plexus tumors from those of velum origin is debatable because the usual large size of the tumors obscures precise definition of their origin. They observed that the former are spherical and lie in the ventricles, while the latter often are partially embedded in the cerebral substance. Our case may provide one of the few instances demonstrating the correctness of their observation.

The giant suprachiasmal meningioma of our patient, surprisingly, had not caused any visual disturbance. It seems that the stretch of the optic chiasm by pressure from its dorsal aspect produces less harm than pressure exerted upon its under surface as caused by pituitary adenomas. In the latter, visual defect is often an early symptom. Türck, Fay and Grant, Lillie, and Rucker and Kernohan explained that the visual change in cases of pituitary adenomas is the result of jacking-up of the optic nerves and chiasm against the pulsating anterior cerebral arteries crossing their dorsal surface. These authors have demonstrated the deep groove in the chiasm or optic nerves cut by the arteries. This change does not occur in a suprachiasmal lesion.

Lastly, the hyperkinetic behavior observed in our patient is an unusual clinical feature of intracranial meningiomas. It not infrequently has been noted in children after severe head injury, encephalitis and encephalopathies. The frequent finding of abnormal electroencephalographic changes in these children indicates an organic basis. Nevertheless, the anatomical correlation of this clinical syndrome is obscure and often conjectural because of the lack of pathologico-anatomic proof and the extensive nature of the cerebral pathology. In experimental animals, bilateral ablation of the orbital gyri produced hyperactivity associated with vasomotor changes. Nevertheless, this by no means resembles the hyperkinetic disorder observed in affected patients, and yet it may indicate that a large lesion involving the orbital frontal and the anterior limbic structures is necessary to produce the clinical syndrome. Our case may constitute such an instance in which the orbital cortices, paraolfactory region, and possibly the amygdaloid have been damaged both by the giant meningioma and surgery. The repeated electroencephalographic findings and the diencephalic seizures are supplementary evidence pointing to the involvement of these areas.

**Summary**

A case of an unusually large suprachiasmal, and small left lateral ventricular meningioma successfully removed from a 4-year-old boy is described with a notation on hyperkinetic behavioral disorder.

**References**