Choroid Plexus Papilloma Five Years after Shunting for Hydrocephalus

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CHOROID PLEXUS PAPILLOMAS are rare tumors. They constitute 0.5% of all intracranial neoplasms.[3,5,6,11] Although usually located in the fourth ventricle in the adult, the lateral ventricle would appear to be the more common site in infants and children. Other locations include the third ventricle and cerebellopontine angles. They show a predilection for the younger age group with greatest incidence in the first and second decades of of life. Males seem to have a slightly greater incidence.

Raised intracranial pressure with hydrocephalus is the common presentation. Associated neurological findings will be dependent on the location and rate of growth of these tumors. Rarely malignant change may occur, when they can be multiple and show dissemination along the cerebrospinal fluid pathways. Diagnosis is established by contrast studies of which ventriculography is the more definitive.

We would like to report a case of choroid plexus papilloma that arose in a patient who had 5 years earlier undergone a ventriculo-atrial shunt for hydrocephalus. The bubble study done then documented the hydrocephalus but unfortunately the reason for it was not apparent. This particular case exemplifies the need for adequate contrast neuroradiological studies prior to shunting procedures.

Case Report

Y.M., a Malay male infant, was initially admitted to our Department at the General Hospital, Kuala Lumpur in January 1970 at the age of 5 months. He was a full term baby and had a normal delivery. The parents stated that the child had begun to have an increasing head size. He was unable to hold his head up. A week prior to admission, he had become apathetic and drowsy. There was no history of meningitis or trauma prior to his admission. On examination, the head circumference was 45 cm. The fontanelles were full and bulging. Head lag was present. Routine laboratory tests and chest x'rays were normal. Skull x'rays revealed a large head with wide separation of sutures. Bubble ventriculography confirmed the hydrocephalus. There was no evidence of tumor at that time. A ventriculo-peritoneal shunt was then concluded and the child discharged well.

Subsequently, three shunt revisions were necessary (all for blocked ventricular ends) over the next 5 years. In between episodes of re-operations for malfunctioning shunt, the patient was alert and well. No contrast studies were performed prior to any of these revisions.

On 17.4.75, the patient was readmitted for vomiting of some 5 days duration. The ventricular end of the shunt was again found to be blocked. A roentgenogram of the skull now revealed a large calcified mass near the midline in the (R) parietal area. This led to further investigation. A (R) brachial angiogram showed a shift (R) to (L) of the anterior cerebral artery with evidence of hydrocephalus. Air studies showed that the ventricular system was indeed generously dilated and the calcified mass was found protruding into the floor and body of the (R) lateral ventricle. CSF examination at this time showed protein of 48 mg%,

sugar 60 mg% with 55 polymorphs and 45 lymphocytes and few RBCs. Some few days prior to definitive craniotomy, the patient developed a (L) hemiparesis.

On 13.5.75, some 4 years and 4 months after his initial operation, a (R) parieto-occipital cranitomy was performed. The tumor was approached interhemispherically. By way of a transcallosal approach the (R) lateral ventricle was opened revealing the tumor. At this time, it was evident that the ventricle was the seat of recent hemorrhage. The tumor was attached to the lateral wall of the ventricle by a vascular pedicle. It appeared papilliferous, soft to gritty in consistency and was relatively avascular. It was totally removed.

The tumor weighed 28 grams and histopathological examination revealed this to be a choroid plexus papilloma with calcification. Villous fronds were numerous, lined by a single layer of cunoidal epithelium. Psammoma bodies and plaques of calcium were numerous.

Post operative recovery was satisfactory. He developed one generalised fit and was placed on Dilantin and Phenobarbitone. On discharge on 28.5.75, he was afebrile and generally well except for a residual (L) hemiparesis. There were no further fits.

A follow-up examination a month later (at the age of 6 years), revealed him to be in good general health with average mental functions and following a continued pattern of normal growth and development. The hemiparesis was hardly present and he was up and about to the satisfaction of all concerned.

Discussion

Progressively enlarging heads in children is often presumed to be due to congenital hydrocephalus. In this patient, it is highly probably that the tumor was present from the very beginning although it was not evident radiologically or felt during ventricular punctures for shunt placements. This may account for the ventricular end of the shunt being blocked on three separate occasions prior to definitive cranitomy to remove the tumor. The need for contrast radiological diagnostic studies prior to shunting cannot, therefore, be overemphasized.

Morbidity and mortality and neurological deficits depend on the size, situation and nature of a tumor. This patient, although had a significantly large tumor, was relatively asymptomatic following shunting as the tumor lay mainly within the lateral ventricle with little involvement of the surrounding areas. This would explain the insidious onset of lateralizing sign and the good post operative recovery. Following its removal, hydrocephalus can be expected to regress. Although rate of growth of these tumors is not known, there is evidence that at least in our case it took 5 years to become clinically manifest and significant. We are inclined to think that shunting may be all that is necessary in small tumors of the choroid plexus. However, in such small tumors, patients should be followed closely. Surgery should be carried out when focal neurological deficits develop, or when hydrocephalus is significant.

Summary

This is a case report of a patient now 6 years old with a calcified choroid plexus papilloma of the (R) lateral ventricle. He was first seen as an infant with the diagnosis of congenital hydrocephalus, aetiology unknown. He was then given a ventriculoperitoneal shunt. Subsequently, he came to require three revisions for blocked ventricular ends of the shunt. An intraventricular tumor was never suspected until radiological evidence was available some 5 years later. That ventricular hemorrhage can occur with these tumors is documented in our case as per our operative findings. The hemiparesis, which developed some 3 days previous to the craniotomy, was presumably the result of this intraventricular bleed.

This paper emphasizes the importance of investigating all forms of hydrocephalus before a decision not to shunt a patient is undertaken. Further, it shows the importance of regular follow-up care in these patients.

References

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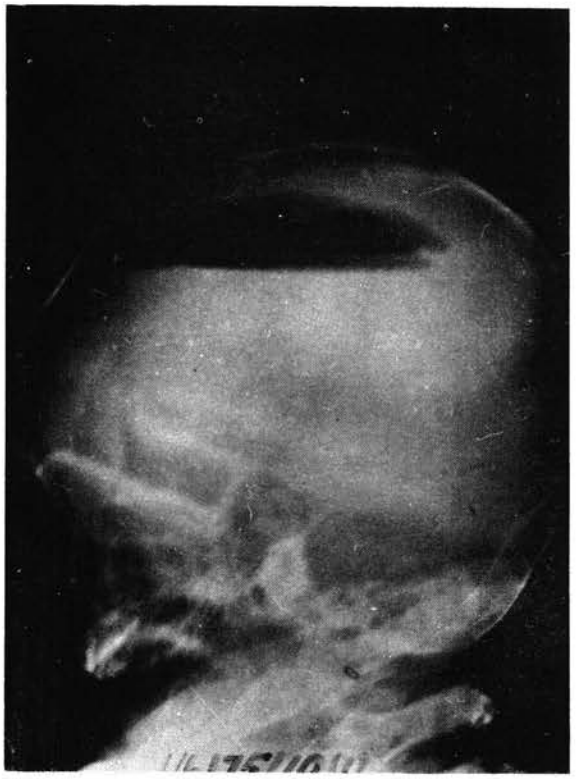

Diagram I
Bubble ventriculography at the age of 5 months documenting hydrocephalus.

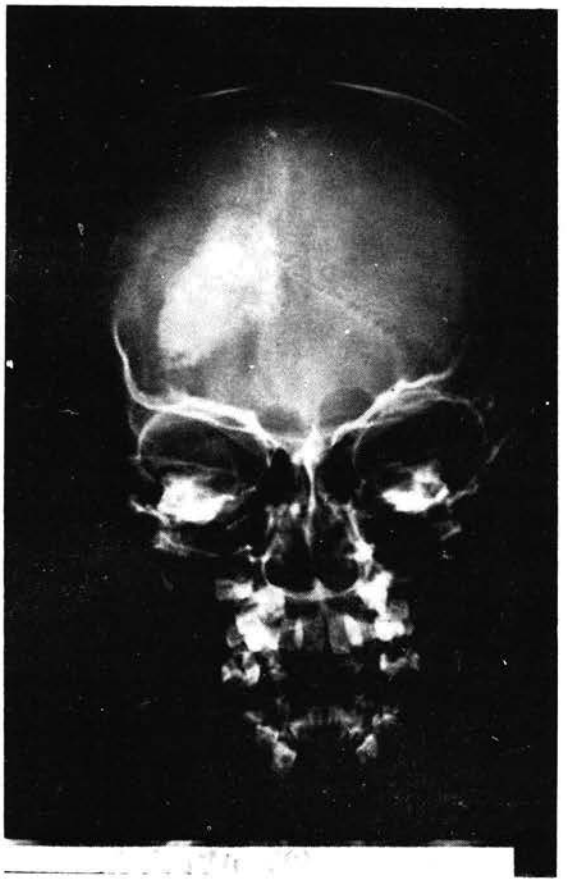

Diagram II
Skull radiograph at the age of about 5 years showed a large calcified tumor in the shape of the (R) ventricle.

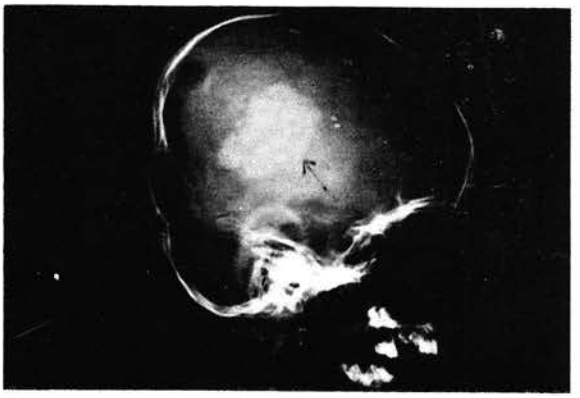

Diagram III
Roentgenogram lateral view illustrating the calcified tumor.

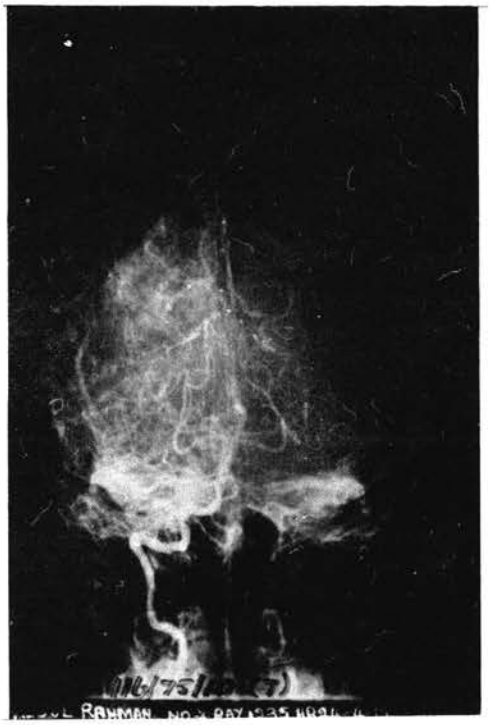

Diagram IV
(R) brachial angiogram showed a (R) to (L) shift of the anterior cerebral artery and lateral bowing of the middle cerebral artery.

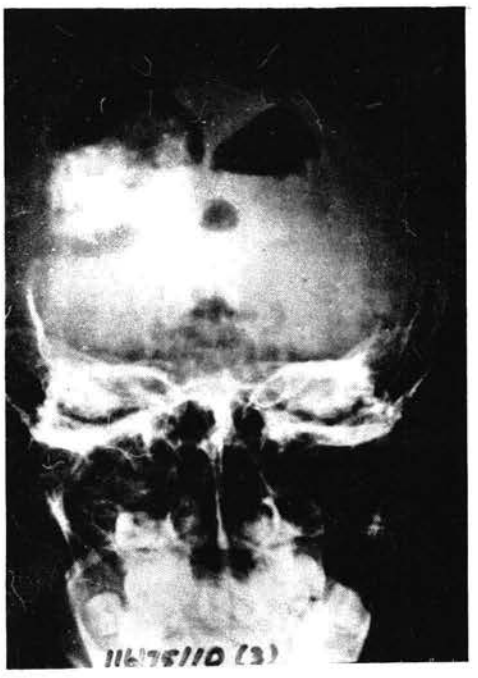

Diagram V
Air study revealed dilated (R) ventricle containing the calcified tumor and also an enlarged 3rd ventricle that was slightly shifted to the (L).

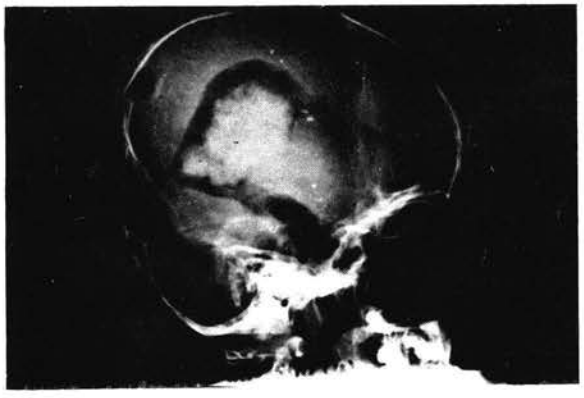

Diagram VI
Air study lateral view further delineated the extent of the tumor and hydrocephalus.

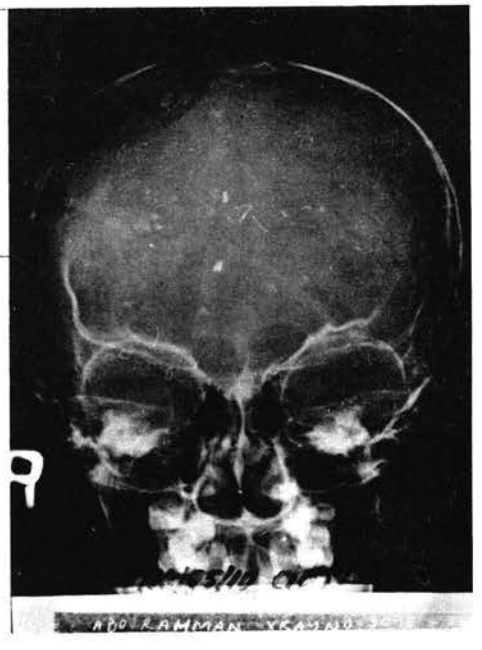

Diagram VII
Radiograph 5 months after operation showed the presence of shunt tip only.