Papilloma of the choroid plexus is a rare tumor. Its incidence according to Herren is about 0.4 per cent of all tumors of the brain and less than 100 examples have been reported. The majority of these tumors have been found at autopsy. Some have been diagnosed in life and a few have been successfully removed.

Apart from its interest as a pathological curiosity, the tumor has a special interest in its relationship to hydrocephalus. Russell in her classical monograph on the pathology of hydrocephalus states that theoretically this condition may arise in three ways: through oversecretion of cerebrospinal fluid, through obstruction at some point in the cerebrospinal fluid pathway or through impairment of absorption. Kahn and Luros cited a personal communication from Russell in which she gives it as her opinion that although the first method of production of hydrocephalus may exist, there is not as yet a well enough documented case to give absolute proof that hydrocephalus can result from the overproduction of cerebrospinal fluid alone.

In the case here reported, evidence has accrued which suggests that overproduction of cerebrospinal fluid was in fact responsible for the hydrocephalus observed.

**CASE REPORT**

G.A.R., a 15-month-old girl, was admitted to Oldchurch Hospital on Nov. 11, 1957 as a case of suspected cerebral tumor.

Her development and health had been normal until the middle of October 1957 when she started to vomit after being fed and a cough developed. She was noticed to be irritable and lethargic. At the end of October she appeared to be suffering from headache and became drowsy.

She was first admitted to hospital on Oct. 31, 1957. Her temperature was 101°F (38.3°C.) and she was lethargic and drowsy. There was no stiffness of the neck and Kernig's sign was negative. Circumference of the head was 17.5 in. (44.5 cm.). She showed no abnormal neurological signs apart from bilateral extensor plantar responses. Her throat was a little reddened and the left ear drum was slightly injected. Count of white blood cells was 20,700 per c. mm. (polymorphonuclear cells 74 per cent); haemoglobin was 91 per cent, and urine was normal. Cerebrospinal fluid pressure was 180 mm. The fluid contained 30 red blood cells per c. mm., no white blood cells, protein 10 mg. per 100 ml., and chlorides 740 mg. per 100 ml. She was treated with trisulphon and penicillin and over the next few days her temperature became normal, her vomiting ceased and she became brighter. She was discharged home much improved on November 5.

Three days later the vomiting recurred, she again became lethargic and drowsy, and developed a high-pitched cry. She was readmitted to hospital on Nov. 11, 1957. She was now irritable and photophobic. The anterior fontanelle was bulging and she had bilateral early papilloedema. Her plantar responses were still extensor but she showed no other abnormal neurological signs. Roentgenograms of the skull showed diastasis of the sutures. The cerebrospinal fluid pressure was 180 mm. The fluid contained 30 red blood cells per c. mm., no white blood cells, protein 10 mg. per 100 ml., and chlorides 740 mg. per 100 ml. She was treated with trisulphon and penicillin and over the next few days her temperature became normal, her vomiting ceased and she became brighter. She was discharged home much improved on November 5.

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*Based on a paper read at a joint meeting of the Dutch and British Neurosurgical Societies at Wassenaar, Holland, on May 9, 1958.
brospinal fluid was under a pressure of 300 mm. It contained 40 red blood cells per c.mm., 10 mg. protein per 100 ml., and 640 mg. chloride per 100 ml. She was transferred to the Neurosurgical Department at Oldchurch Hospital later the same day.

On November 12 ventriculography was carried out by passing a short lumbar puncture needle into the right lateral ventricle through the coronal suture. The cerebrospinal fluid pressure was 300 mm. and the fluid contained 35 red blood cells per c.mm., no excess of white blood cells and 10 mg. of protein per 100 ml.; 100 ml. of cerebrospinal fluid were replaced by air. Radiography demonstrated gross symmetrical dilatation of the lateral ventricles (Figs. 1–3). The right temporal horn did not fill, but the significance of this finding was not appreciated at the time. The third ventricle was moderately enlarged and in the midline. The fourth ventricle was small and appeared to be displaced to the left. There was a large amount of air in the cisterna magna.

The interpretation of these ventriculograms gave rise to considerable difficulty. The differential diagnosis at this stage seemed to be between a cerebellar tumour, otitic hydro-

Figs. 1 and 2. Ventriculograms. (Left) Anteroposterior view, showing symmetrical dilatation of lateral ventricles. The right temporal horn does not fill. (Right) Lateral brow-up view, showing distended frontal horns.

cephalus following her illness 2 weeks earlier, obstructive hydrocephalus resulting from adhesions around the brain stem and a primary failure of absorption of cerebrospinal fluid. The latter diagnosis was difficult to reconcile with her normal development up to the age of 15 months and the normal size of the head.

A posterior fossa exploration was carried out on Nov. 12, 1957. The cisterna magna was greatly enlarged and under tension. After the arachnoid had been opened, large quantities of cerebrospinal fluid were seen to be coming from a distended foramen of Magendie. The cerebellar hemispheres had been pushed upwards and outwards by the dilated cisterna magna. There was no evidence of a cerebellar tumour.

The child’s immediate postoperative recovery appeared to be very satisfactory and within a week she was alert and apparently free from headache. Two weeks after operation, however, the posterior fossa decompression began to bulge and to become tense. It was aspirated daily for several days, about 50 ml. of clear cerebrospinal fluid being removed on each occasion. The protein content of the fluid was estimated on three separate occasions and found to be 10 mg. per 100 ml. each time. After each aspiration it was noted that there was a profuse leakage of cerebrospinal fluid for several hours from the site of the puncture. The dressings and the
pillow on which the child's head was resting would become soaked with cerebrospinal fluid. The leak would cease after a few hours and by the following morning the decompression would be very tense again. In spite of these punctures the child's condition steadily deteriorated. She became drowsy and lethargic again, and although large quantities of fluids and electrolytes were being given rectally and intravenously, signs of dehydration with peripheral circulatory failure appeared.

By December 3 the intracranial hypertension was still uncontrolled and the child was thought to be blind. A right frontal burr-hole was made and a fine rubber catheter was inserted into the right lateral ventricle to establish continuous ventricular drainage. The ventricular pressure at this time was 300 mm. It was lowered to 50 mm. over a period of 24 hours and kept at that level until the time of the child's death just over 3 days later. During these 3 days the quantities of cerebrospinal fluid drained through the catheter were 500 ml., 400 ml., and 960 ml. respectively. Clinical evidence of dehydration was present throughout this period. In spite of every effort to keep pace with this great loss of fluid and electrolytes the child became steadily worse and died in coma on Dec. 6, 1937.

Postmortem Examination (Dr. C. Raeburn). Apart from terminal bronchopneumonia, the only abnormality was in the brain. In the posterior part of the right lateral ventricle there was a rounded red tumour weighing 13 gm. It had a finely lobulated surface, measured $1\frac{3}{4} \times 2 \frac{3}{4} \times 1\frac{1}{2}$ (2.5 cm. x 3.75 cm. x 1.25 cm.), and was attached to the free posterior extremity of the choroid plexus (Fig. 4). The tumour was freely mobile on its pedicle within the ventricle (Fig. 5). All the intracranial venous sinuses were patent and normal. There were no adhesions around the brain stem or at the tentorial opening.

Histology (Dr. I. M. Larkin). The tumour was a well-differentiated papillomatous growth, very similar in appearance to normal choroid plexus and showing a little granular calcification (Fig. 6).

DISCUSSION

According to Van Wagenen, the favourite site of papillomas of the choroid plexus is the fourth ventricle, 50 per cent being so situated; 17.3 per cent occur in the third
CHOROID PLEXUS PAPILLOMA

Figs. 4 and 5. (Left) The tumour *in situ* in the right lateral ventricle. (Right) Showing free mobility of tumour on its pedicle.

Fig. 6. Photomicrograph of tumour showing typical appearance of benign choroid plexus papilloma (×115).
ventricle and 34.7 per cent in the lateral ventricles. When the tumour is present in
the third or fourth ventricle hydrocephalus presumably results from obstruction to
the flow of cerebrospinal fluid. However, hydrocephalus is not confined to this type
of case. Herren,² in a careful analysis of over 80 published cases, stated that “internal
hydrocephalus is an almost universal accompaniment of this kind of tumor . . . and
this condition obtains whether or not there is mechanical obstruction to the circu-
lation of cerebrospinal fluid.”

What evidence is there as to the mode of production of hydrocephalus in those
cases in which a papilloma of the choroid plexus causes no obstruction to the flow
of cerebrospinal fluid?

In the first place, these tumours consist of a benign overgrowth of the choroid
plexus. They have a large secretory surface and might therefore be expected to se-
crete an excessive quantity of cerebrospinal fluid.

Fehrl described a patient with a papilloma of the choroid plexus in the fourth
ventricle who was at first thought to be suffering from serous post-traumatic menin-
gitis. Although 200–300 ml. of cerebrospinal fluid were removed daily by lumbar
puncture for several days there was only transient diminution in the increased
cerebrospinal fluid pressure.

Kahn and Luros³ reported a case of papilloma of the choroid plexus of the lateral
ventricle in a 20-year-old woman associated with papilloedema and marked hydro-
cephalus. Removal of the tumour produced clinical cure over a 5-year period with
disappearance of the papilloedema. Matson⁴ successfully removed a papilloma of the
choroid plexus from the lateral ventricle of a 4-week-old baby with apparent cure of
a severe degree of hydrocephalus. A similar result followed excision of bilateral
angiomatous choroid plexus in a 3-month-old infant described by McGuire et al.⁵
The histology of these angiomas resembled that of papilloma.

Some very interesting observations were made by Ray and Peck⁶ in the course
of their treatment of a 2½-month-old infant with communicating hydrocephalus who
was eventually found to have bilateral papillomas of the choroid plexus. In the
first place a shunt was carried out between the lumbar subarachnoid space and the
right ureter. The baby tolerated the procedure well but on the 3rd postoperative
day she rapidly became dehydrated and went into shock. She responded to infusion
of saline but over the next few days it became apparent that she was losing an ex-
cessive amount of water and electrolytes via the ureteral shunt. In order to keep up
with this loss, daily infusions of 500–1,000 ml. of half-normal saline had to be given,
as well as extra salt in the diet. Any attempt to reduce these replacements resulted
in serious dehydration often bordering on shock. After 8 weeks it was clear that no
adjustment to the continued excessive loss of water and electrolytes was occurring.
The tube was therefore removed from the ureter and transplanted into the peri-
toneal cavity in the hope that the fluid would be reabsorbed. The next day there
was great abdominal distension and signs of dehydration reappeared. Thereafter the
baby improved and equilibrium was temporarily established by oral feeding alone.
About 6 months later, however, the baby was readmitted because of gross ascites.
Aspiration of 3,250 ml. of fluid from the peritoneal cavity was followed by a further
episode of acute dehydration. Subsequent operations revealed the presence of a large
papilloma in each lateral ventricle.

In the case which I have described, a papilloma of the choroid plexus of the right
lateral ventricle was associated with marked hydrocephalus in the absence of any
obstruction to the circulation of cerebrospinal fluid either inside the ventricular
CHOROID PLEXUS PAPILLOMA

system or elsewhere. The child suffered from an acute rise of intracranial pressure. A posterior fossa exploration gave very transient relief. Within 2 weeks the intracranial pressure had risen again and could not be controlled by daily punctures of the posterior fossa decompression. Each of these punctures was followed by profuse leakage of cerebrospinal fluid along the track of the needle for several hours. Finally continuous ventricular drainage was instituted through an indwelling catheter in the right lateral ventricle. In the 3 days during which this catheter was in situ, the quantities of cerebrospinal fluid drained were 500 ml., 400 ml., and 960 ml. respectively. During these 3 days, moreover, there was clinical evidence of marked dehydration. These quantities are far in excess of those obtained when continuous ventricular drainage is established for the relief of raised intracranial pressure as a preliminary to operation on cerebellar tumors.

This case, therefore, provides direct evidence of the very large quantities of cerebrospinal fluid that can be secreted by a papilloma of the choroid plexus. I have found no record of a case in which it has been possible to make similar direct observations on the volume of cerebrospinal fluid secreted in this disease. Together with the other evidence mentioned above, the facts observed in this case lend strong support to the view that oversecretion of cerebrospinal fluid is the cause of the hydrocephalus in such cases.

This tumour could readily have been removed had it been detected in life. More attention should have been given to the nonfilling of the right temporal horn in the ventriculogram. The probable reason for our failure to demonstrate the tumour in routine views was the free mobility of the tumour on its pedicle within the dilated ventricle. A larger replacement of air might have prevented this failure and should be considered if a suspicion of choroid plexus papilloma arises.

SUMMARY

1. A case of papilloma of the choroid plexus of the lateral ventricle is reported.
2. Evidence based on this and similar cases is presented to support the view that the associated hydrocephalus is caused by oversecretion of cerebrospinal fluid.

REFERENCES