Symptomatic cysts of the telencephalic choroid plexus

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SUMMARY Symptomatic cysts of the telencephalic choroid plexus are rare. This is a paediatric problem, with the oldest patient being 10 years old. Pertinent cases from the literature are reviewed. The case of a 9 year old girl with suboccipital headaches made more severe by lying on her right side or on her abdomen is discussed. Physical examination was within normal limits except for evidence of early papilloedema. The cerebrospinal fluid pressure was normal, and the protein was not elevated (32 mg/100 ml.). The brain scan showed a left frontoparietal mass near the midline, and the electroencephalogram was abnormal. The pneumoencephalogram demonstrated a mobile, pedunculated mass in the left trigone which approached the foramen of Monro when the patient assumed the head-erect position. A transcallosal approach was used and the cyst was easily removed. Post-operatively the patient has done well and is currently asymptomatic and without headaches. The significance of the presenting symptoms, the cerebrospinal fluid and brain scan findings, as well as the surgical approach, are discussed.

Headaches are a common presenting complaint, and an evaluation of this problem is made frequently by all clinicians. A child who complains of a headache which is related to assuming certain positions may be harbouring an intraventricular tumour. Walter Dandy (1934) noted that tumours of the lateral ventricle comprise only 0.75% of intracranial neoplasms. In 1934 he stated, 'I know of no instance when a cyst of the (telencephalic) choroid plexus has caused symptoms While it is by no means improbable that cysts of the choroid plexus may at a later date be found to cause obstruction to the (lateral) ventricular system, the great frequency of cysts without any evidence as yet of one causing symptoms is proof that such an outcome will, at least, be rare' (Dandy, 1934). Since that time two cases have been documented in the literature (Baker and Gottlieb, 1956; de la Torre, Davis, and Crandell, 1963). This presentation provides the third case of a telencephalic choroid plexus cyst which was symptomatic. The mechanism of intermittent obstruction of the foramen of Monro by a large, pedunculated cystic mass is graphically illustrated by pneumoencephalography. The consistent finding of normal cerebrospinal fluid and the presence of significant brain scans are described and a review of the litera-

ture is provided. A transcallosal approach to lateral ventricle masses is recommended because of its relative ease and the resultant good visualization of the lateral ventricle without requiring the sacrifice of any neurological function.

PATHOGENESIS

Cysts of the telencephalic choroid plexus have been found in from 57 to 64% of serial necropsy studies (Findlay, 1899; Dunn, 1954; Shuangshoti and Netsky, 1966a). Several theories have been presented to explain their occurrence, but the most acceptable explanation is provided by Shuangshoti and Netsky (1966a, b). There are four stages in the development of the human choroid plexus. In stage I, the choroid villi are created by the folding of the epithelium into the choroidal matrix, and the stroma into the cerebral ventricle. These tips of folded epithelium become neuroepithelial lined cysts within the choroidal matrix. Mesenchymal mucin and other materials accumulate, enlarging these spaces. With time there is an increase in collection of these substances plus a retrogressive change in the plexus. The result is a visible cyst.

Two facts must be kept in mind: first these cysts are

not simply the result of advancing age, for their prevalence is as great at age 1 year as at 90 years (Shuangshoti and Netsky, 1966a). The ages of the three patients with symptomatic cysts were 4, 9, and 10 years. Second, only a very few of these cysts become symptomatic, because of the critical relationship which must exist between the size of the cyst and its proximity to the foramen of Monro. Because most cysts arise in the glomus, they must attain sufficient size to extend rostrally to the level of the foramen.

REVIEW OF CASES

In 1876, George Brown recounted the history of a 4 month old white male who presented with a three-day siege of intractable vomiting. By the ninth day of illness, the infant 'took no notice of any person or thing; the pupils were widely dilated and there was lachrymation of the left eye; no paralysis or convulsions . . . There was no bulging of the fontanelles, but the left parietal eminence was markedly more prominent than the right' (Brown, 1876). The patient expired the next day. At postmortem examination, there was a cyst arising from the choroid plexus of the left lateral ventricle, which completely filled this cavity and protruded through the corpus callosum and between the hemispheres, touching the dura mater. The walls of the cyst were extremely thin and translucent, being filled with clear serous fluid. No histopathological description was provided. This is the classic article, but as noted by Dandy, the absence of histological data 'makes such speculation idle' (Dandy, 1934). Without this information it cannot be determined if this was a cyst of the choroid plexus.

Baker and Gottlieb (1956) described the case of a 10 year old white male who presented with a five-month history of intermittent headaches. It was noted by the child that changes in the position of his head produced severe bi-frontal headaches. The patient stated that upon assuming the supine position his headache subsided, but while in an upright position with certain head placements, the headache became intense. This headache was associated with nausea and vomiting, and on one occasion, with the loss of consciousness. Examination of the cerebrospinal fluid revealed a normal pressure with a protein of 30 mg/100 ml., and 8 lymphocytes/cu. mm. A ventriculogram demonstrated an intraventricular mass in the left lateral ventricle in the region of the trigone.

A left parieto-occipital craniotomy was performed with an approach through the superior parietal lobe entering the posterior horn of the lateral ventricle. A bluish-red cystic tumour measuring approximately three-fourths of an inch in diameter was found situ-

ated in the choroid plexus. The base of the cyst was exposed, coagulated, and completely removed. During the procedure the structure ruptured and clear fluid with an appearance similar to cerebrospinal fluid was released.

Microscopically, the cyst was unilocular and the walls were friable, very thin, and composed entirely of fibrous tissue elements. The patient followed an essentially uncomplicated postoperative course and remained free of headaches. The authors emphasized the significance of the paroxysmal nature of the headaches, associated with an influence of positional change. They believed the mechanism was a ball-valve type of obstruction with intermittent blockage of the occipital and temporal horns of the left lateral ventricle.

De la Torre *et al.* (1963) presented the case of a 4 year old female with generalized seizures and mild left hemiparesis. The child had an ataxic gait and an equivocal left extensor plantar response. Plain skull films were within normal limits. The cerebrospinal fluid pressure and protein were both normal. A ventriculogram demonstrated moderate dilatation of both lateral ventricles with the right slightly larger than the left. There was a filling defect in the right lateral ventricle in the region of the trigone. A posterior temporoparietal craniotomy was performed with a posteroparietal cortical incision. The cyst was removed and the child did well postoperatively. A description of the microscopic findings was not provided.

CASE REPORT

This patient was a 9 year old white female who presented with a two-year history of bilateral occipital headaches. The pain was of mild intensity and appeared periodically. These headaches progressed in frequency, intensity, and duration. The patient was seen by the family physician who obtained radiographs of the skull and an electroencephalogram, both of which were within normal limits. One month later the patient developed a severe suboccipital headache followed by a grand mal seizure. A repeat electroencephalogram demonstrated only diffuse slow wave activity. Skull radiographs were again negative, and a brain scan was normal. The patient was placed on 32 mg of phenobarbital, three times daily. Over the course of the next three months the headaches persisted and continued to increase in severity. It was then noted by her parents that the headaches were most severe upon arising in the morning, and tended to subside once the child began to walk. If she slept on her right side, or on her abdomen, the headaches were of greater intensity. No other head or body position produced a change of symptomatology. Occasionally the child experienced nausea without vomiting. The patient also had 'bad thoughts' consisting of visual and auditory hallucinations. For example, she once tried to show her mother a trailer-house which she 'saw' on a vacant lot next to their home.

Similar episodes occurred on three or four occasions.

On physical examination there was a 2-dioptre papilloedema OS present, and 3-dioptre papilloedema OD. The remainder of the neurological and physical examination was within normal limits.

Routine laboratory estimations were within normal limits. On spinal puncture, which was mildly traumatic, the opening pressure was 150 mm of water. There were 2,750 red blood cells c.mm and the protein was 32 mg/100 ml. of CSF. A second cerebrospinal fluid specimen was obtained at the time of the pneumoencephalogram and the

protein was 33 mg./100 ml., and no cells were present. A brain scan using technetium⁹⁹ revealed a left frontoparietal mass near the midline (Fig. 1). The electroencephalogram demonstrated diffuse slow wave activity. On pneumoencephalography a 17×14 mm, pedunculated, mobile choroid plexus mass was identified at the medial aspect of the floor of the anterior body of the left lateral ventricle. In the brow-up position, the mass lay several millimetres dorsal to the left foramen of Monro, but in the head-erect position the mass moved forward into a position capable of producing at least partial obstruction

FIG. 1. Frontal and lateral brain scan demonstrate increased uptake in the left frontoparietal region.

FIG. 2. (a) Brow-up pneumoencephalogram shows a 17×14 mm pedunculated cyst of the left telencephalic choroid plexus which, in this position, lies several mm posterior to the foramen of Monro. (b) the erect head position demonstrates how this pedunculated cyst falls forward to obstruct the passage of cerebrospinal fluid through the foramen of Monro.

of the cerebrospinal fluid passageway (Fig. 2). There was a moderate enlargement of the left lateral ventricle in comparison with the normal size of the right lateral ventricle (Fig. 3).

A frontal flap was turned and a transcallosal approach was used. The left lateral ventricle was opened and a greyish mass measuring approximately 20 mm in diameter was noted extending from the glomus. At this point in the procedure, the mass was ruptured and a clear fluid similar to cerebrospinal fluid extruded from this cystic mass which collapsed. In the trigone a mass of tissue compatible with choroid plexus was noted. This was excised at the base, haemostasis being assured by application of tantalum clips to its vascular base. There was no blood loss. After this, the closure was secured.

On examination of the tissue specimen, it was found to balloon out into a large cystic mass when placed in a fluid medium. On microscopic examination it was a thin-walled, cystic structure with delicate vascular spaces and loose connective tissue. This appearance was compatible with distended choroid plexus (Fig. 4).

The patient followed an uncomplicated, headache-free, postoperative course for eight months. Then a series of seizures prompted her readmission to the hospital. All

FIG. 4. Collapsed and convoluted cyst wall which is almost devoid of lining epithelium.

aspects of the work-up were within normal limits. A repeat pneumoencephalogram demonstrated less asymmetry between the lateral ventricles and no evidence of the obstructing cystic mass. Since that time the patient has been asymptomatic.

DISCUSSION

There are several aspects of this problem worthy of discussion. Baker and Gottlieb (1956) stressed the paroxysmal nature of the symptoms, as well as the influence of head position upon the intensity of the headache. Our case also displays these characteristics, and the pneumoencephalogram confirmed the mechanism. The pedunculated, mobile mass periodically obstructed the left foramen of Monro when the head was in a head-erect or brow-down position (Fig. 2). This graphically explains the significance of head position and the reason for the intermittent nature of these symptoms. When performing the air contrast study, it is recommended that a sufficient quantity of air be used to provide adequate filling of the lateral ventricles so that small masses will not go unvisualized.

Examination of the cerebrospinal fluid provides interesting findings. Measurements of the pressure are within normal ranges. The protein values were normal in the two cases from the literature as well as in the case presented here. This is important from a diagnostic standpoint, because in the face of an air contrast study which demonstrates a choroid plexus mass, it is generally expected that there will be found an increase in protein in association with a papilloma of the choroid plexus.

The brain scan using technetium⁹⁹ was positive in this case for a left frontoparietal mass in the position of the telencephalic choroid plexus. It is recom-

FIG. 3. Frontal pneumoencephalogram shows the cyst of the choroid plexus on the medial aspect of the body of the left lateral ventricle. Note that the left lateral ventricle is larger than the right, thus fulfilling one of the true criteria as specified by Dandy.

mended that repeat scans be obtained after the patient is given Lugol's solution or potassium perchlorate, so that the active uptake of technetium⁹⁹ can be inhibited and confirmation of the presence of a choroid plexus mass can be obtained (Witcofski, Janeway, Maynard, Bearden, and Schultz, 1967).

The surgical approach is also important. De la Torre *et al.* (1963) discuss a vertical incision and transection of the cortex in the posterior parietal region, noting that it avoids damaging the functioning cortex. However, this leaves the surgeon lateral, posterior, and superior to the tumour, making exposure of the main vascular pedicle of the tumour difficult, as the initial step. Therefore, they proposed the use of a horizontal transcortical incision along the posterior portion of the middle temporal gyrus which would permit the surgeon to elevate the tumour and to clip the feeding vessels, the choroidal arteries, which usually enter the lateral ventricle through the choroidal fissure. Severe, and sometimes permanent, dysphasia has been reported as a consequence of this approach when the cortical incision is made in the dominant hemisphere.

The transcallosal approach is highly recommended for telencephalic intraventricular tumours. A standard frontal flap is fashioned and the dura mater incised laterally and reflected medially to the superior sagittal sinus. The sagittal portion of the frontal lobe is retracted laterally and the pericallosal arteries are identified and separated. The anterior corpus callosum is then incised and the lateral ventricle entered. From this vantage point masses can be visualized and removed with relative ease. A transcallosal approach is advocated because it can be accomplished with relative ease, good visualization of the lateral ventricular system is provided, and this procedure does not inherently produce any neurological deficit.

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