CASE REPORTS AND TECHNICAL NOTES

HYDROCEPHALUS IN A PREMATURE INFANT CAUSED BY PAPILLOMA OF THE CHOROID PLEXUS

WITH REPORT OF SURGICAL TREATMENT

DONALD D. MATSON, M.D.

Neurosurgical Service of the Children's Medical Center, and Department of Surgery,
Harvard Medical School, Boston, Massachusetts

(Received for publication December 9, 1952)

To warrant publication, a single case report should record a clinical experience sufficiently unusual and provocative to become valuable in itself to others who may encounter a similar problem. There is a widespread clinical impression, shared certainly by this author, that the outlook for infants born with any marked degree of cranial enlargement, as well as the outlook for infants with rapidly increasing head circumference during the first few weeks of life, is uniformly poor. Perhaps it is important, therefore, to set down an exception to this rule, and thus not only present a unique case, but in so doing, suggest an occasional happier outcome to an ordinarily discouraging and futile clinical problem.

The purpose of this report is to describe a 5-weeks premature infant, born with an enlarged head, from whom a large papilloma of the choroid plexus was removed at 4 weeks of age, or 1 week before the infant's estimated birth date, with apparent surgical cure (1 year) of a severe degree of hydrocephalus and with normal mental and physical development to date.

C.M.C. #378515. C.T. was born 5 weeks prematurely on Sept. 25, 1951. The mother had profuse vaginal bleeding for 48 hours, at which time a diagnosis of premature separation of the placenta was made and the infant was delivered by Cesarian section at the Rutland, Vt., Hospital. The birth weight was 6 lbs., 7 oz. The baby seemed normal at birth except for a large head.

At 1 week of age it was noted that the head seemed to be growing rapidly. At that time it measured 15½ in. (38.7 cm.) in circumference and 1 week later it had increased to 16½ in. (41.5 cm.). At 15 days of age the infant was seen by a consultant and transferred to the neurosurgical service of the Children's Medical Center for evaluation and decision regarding the possibility or advisability of treatment.

Examination. On admission the baby was hypertonic and irritable. He weighed 6 lbs., 14 oz. The head appeared markedly enlarged with respect to the size of the trunk (Fig. 1); it measured 41.5 cm. in circumference. The anterior and posterior

Fig. 1. Appearance of 5-weeks premature infant 15 days after birth. Head circumference increased almost 3 cm. during the second week of life.
fontanelles were enlarged and tense, the scalp was thin and shiny, and the veins of the scalp were congested. The cranial sutures were palpably separated and there was an extreme "cracked-pot" percussion note. The deep tendon reflexes were diffusely hyperactive. The baby's vital signs were normal and routine blood and urine analyses were not significant.

Plain roentgenograms of the skull showed an enlarged cranial vault with extremely thin bones, separation of the sutures, and soft tissue swelling in the region of the anterior fontanelle. There were no localized abnormalities or areas of intracranial calcification.

Bilateral subdural taps were negative. Ventricular puncture through the right coronal suture revealed slightly xanthochromic clear fluid at a depth of 1 cm. with a pressure of 200 mm. water. The ventricle was not located on attempted tap on the left side. Lumbar puncture revealed clear, slightly xanthochromic spinal fluid with a pressure also of 200 mm. water. The ventricular fluid protein content was 210 mg. per cent and the lumbar spinal fluid, 374 mg. per cent.

Ventricular air studies were performed the following day. The left ventricle was encountered at a depth of 3 cm.; air was introduced separately into this space and into the more superficial fluid-containing cavity on the right (Fig. 2). This study was interpreted as indicating a large congenital porencephalic cyst of the right cerebral hemisphere and surgical exploration was planned to attempt to establish communication between this cyst and either the lateral ventricle or the basilar cisternae.

1st Operation. On Oct. 16, 1951, under endotracheal ether and oxygen anesthesia, supplemented by local procaine infiltration, a right parietal osteoplastic craniotomy was performed. The infant's general condition was extremely poor, most of the operation being conducted under artificial respiration by positive pressure oxygen insufflation through the endotracheal tube. When the cortex was exposed the expected large subcortical cyst was easily identified and opened. Exploration within revealed a multiloculated cyst, the medial wall of which consisted partially of a large, irregular, reddish-purple vascular tumor and partially of a translucent membrane which when incised proved to be the lateral wall

Fig. 2. Ventricular air study at 18 days of age. There is a huge multiloculated cyst in the right cerebral hemisphere, with displacement of the dilated ventricular system toward the left. Both the cyst and the ventricular system contained clear xanthochromic fluid; the color was deeper in the cyst fluid.

Fig. 3. Gross appearance of the tumor after excision. Actually, the size of the tumor diminished considerably as soon as its blood supply was completely secured.
of the right lateral ventricle. Grossly, the tumor had the appearance of a large ependymoma, though choroid plexus tumor was considered. At this stage the baby was moribund and hypothermic, and resection seemed ill-advised. Biopsies were therefore taken from two different parts of the tumor and closure was carried out. After an anxious few hours, the baby responded well and improved steadily.

Histological diagnosis of the biopsies was papilloma of the choroid plexus.

2nd Operation. Therefore, 7 days later, on Oct. 23, 1951, again under positive pressure endotracheal ether and oxygen insufflation plus local procaine infiltration, the osteoplastic flap was re-elevated. The cyst was re-entered and the tumor exposed and completely removed (Fig. 3). It was attached to the choroid plexus in the body of the right lateral ventricle and measured 4.0×3.5×2.5 cm. Since the entire lateral wall of the lateral ventricle was removed there was ample communication between the cyst and the ventricular lumen. All of the choroid plexus in this ventricle was excised. Again, after a rather uneasy first postoperative day, the infant improved steadily. His anterior fontanelle remained depressed. He was discharged home on the 10th postoperative day, at which time he weighed 8 lbs. 6 oz. and his head circumference was 40.5 cm., 1 cm. less than at the time of admission.

Pathological diagnosis on all samples of this large tumor was papilloma of the choroid plexus with no evidence of malignancy (Fig. 4).

Course. The patient has been followed for more than 1 year since operation and has remained well. There has been no evidence of increased intracranial pressure. One year after

![Fig. 4. Low power photomicrograph of the tumor. The histological appearance is that of normal choroid plexus, which is a core of vascularized connective tissue lined by a single layer of columnar epithelial cells. No evidence of malignancy was noted.](image-url)
HYDROCEPHALUS IN INFANT CAUSED BY PAPILLOMA

operation the head appears normal in size and shape (Fig. 5). There have been no convulsive seizures. Neurological examination is negative; there is no suggestion of hemiplegia or hemianopia on the left. The infant is alert and happy, sits up well, stands with help, says a few words and in general appears to be developing at a satisfactory rate.

COMMENT

That papillomas of the choroid plexus of the lateral ventricle may be accompanied by diffuse enlargement of the entire ventricular system in spite of the absence of any obstruction to the outflow of cerebrospinal fluid from the ventricles seems well established. Eight such patients have been encountered by the neurosurgical service of the Children's Medical Center, of whom the present patient is the youngest. Numerous others have been reported,1,4-14 including at least 2 others in newborn infants.2,3 In the latter 2 cases the diagnosis was made post mortem.

If hydrocephalus in these patients is caused solely by increased rate of formation of cerebrospinal fluid in the papilloma of the choroid plexus or simply by dilatation of the ventricular system resulting from the presence of the large pulsating mass within its confines, then the hydrocephalus should be arrested promptly by successful removal of the tumor. This appears to be true in the young patient reported here, and in others in this clinic in whom satisfactory removal of the lesion has been accomplished.

It is recognized that these tumors sometimes exhibit histological features of an adenocarcinoma and may occasionally seed to other ependymal surfaces. It is possible, therefore, that the tumor in the present patient may still recur, since the follow-up period is little more than 1 year. However, resection seemed grossly complete and the microscopic appearance throughout the tumor was uniformly benign.

Marked enlargement of the head at birth or rapid expansion during the first weeks of life is usually associated with gross malformation of structures within the posterior fossa, with extreme stenosis of the aqueduct of Sylvius, or with widespread failure of development of the surface subarachnoid pathways at the base of the brain. All of these conditions are unfavorable to surgical treatment in the first few weeks of life and severe hydrocephalus with destruction of cerebral substance usually ensues rapidly.

However, it is never safe to assume that a treatable lesion may not be present until certain basic tests have been performed. The presence of a markedly elevated total protein content in the cerebrospinal fluid or of xanthochromia in the absence of red blood cells should suggest the possibility of intraventricular tumor. In this clinic it is standard procedure to carry out bilateral subdural taps, combined ventricular and lumbar puncture and ventricular air studies on every infant with hydro-
Cephalus who does not already exhibit gross evidence of irreversible neurological damage, no matter what the age. Routine studies of this character, supplemented by additional diagnostic and surgical pursuit of unusual clinical situations, should prevent any possibility of overlooking the occasional favorable lesion of the newborn period.

SUMMARY

A case of hydrocephalus caused by a large choroid plexus papilloma of the lateral ventricle is reported in a newborn premature infant.

Following excision of the tumor at 4 weeks of age, the infant has developed normally with no evidence of convulsive seizures, neurological deficit, or increased intracranial pressure for 1 year to date.

This patient appears to represent an exception to the extremely poor prognosis ordinarily entertained for newborn infants with rapidly advancing cranial enlargement.

REFERENCES