CASE REPORTS AND TECHNICAL NOTES

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

WITH REPORT OF SURGICAL TREATMENT

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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.