LARGE CALCIFIED CRANIOPHARYNGIOMA AND BILATERAL SUBDURAL HEMATOMATA PRESENT AT BIRTH

SURVEY OF NEONATAL BRAIN TUMORS

H. Harvey Gass, M.D.*

Detroit, Michigan

(Received for publication April 5, 1956)

This report is made to record the occurrence in a newborn not only of a large calcified intracranial mass (craniopharyngioma) but its concomitant association with bilateral subdural hematomata developing in utero. A survey of the literature has failed to reveal another patient in whom a craniopharyngioma was present at birth. The earliest case found was one mentioned in a report by Jackson. In his paper of 1916 he referred to a case reported by Lawson in 1887. The patient was a 3-month-old male infant. Two references to craniopharyngioma occurring in children of the age of 2 years have also been found. The rarity of a large calcified tumor at birth and the presence of bilateral subdural hematomata are self-evident. A survey of other brain tumors occurring in the newborn will be made.

CASE REPORT

A neurosurgical consultation was requested at St. Joseph Mercy Hospital in Detroit, Michigan by the pediatrician in attendance who recognized an enlarged head and bulging widely patent fontanelles in a newborn full-term male delivered on April 24, 1955. He showed no other physical defects. The pregnancy had been normal, and the child had two normal siblings. The delivery was uncomplicated, but low forceps was employed. Immediately following birth the condition of the child was considered poor, and artificial respiration was instituted subsequent to which he was placed in an incubator. The birth weight was 7 pounds and 15 ounces.

Examination. When first seen by the writer the child was 2 days of age and showed no neurological deficit except for the enlarged head (circumference 43 cm.). He cried somewhat feebly, but was otherwise active and alert. There was marked palpable separation of suture lines and bulging of intracranial contents between all ununited bone plates.

Course. The initial clinical impression was of marked hydrocephalus developing in utero. However, bilateral fontanelle taps at the age of 7 days revealed that the condition apparently was caused by bilateral subdural hematomata. Sixty-three cc. of yellow fluid were easily aspirated from the right subdural space, and 58 cc. of yellowish somewhat sanguineous fluid were similarly removed from the left subdural space, resulting in marked scaphoid depression of the anterior fontanelle. The size and color of these hematomata at such a short time following birth, plus the existence of an enlarged head at birth seemed to indicate that these clots had existed in utero and had not occurred as a result of parturition. Attempts to discover a blood dyscrasia as an explanation of the development of hematomata in utero were unrewarding and became unnecessary when roentgenograms of the skull revealed the large calcified intracranial mass shown in Fig. 1. The presence of this mass and its movement with in utero movements of the fetus could well explain a tear of bridging subdural veins and consequent development of subdural hematomata.

During the next 4 weeks bilateral fontanelle taps with drainage by needle of the hema-

* 612 Kales Bldg., Detroit 26, Michigan.
tomata were done on six different occasions. After the fourth tapping the left hematoma was dry and remained so. However, the right-sided one continued to accumulate. Beginning at the fourth week of life further tapping of the right subdural hematoma failed to reduce the bulging anterior fontanelle to a scaphoid state, as had occurred previously, and the growth of the head increased more rapidly (from 43 to 46 cm. in the 2-week period) suggesting abrupt development of obstructive hydrocephalus. The mass itself was needled through the fontanelle on two occasions. The first time this was done was at 11 days of age, but only 1–2 cc. of bloody fluid could be obtained and no significant tissue was removed. At 2½ weeks of age the mass was again needled and 5 cc. of straw-colored oily fluid were obtained. The infant was discharged home on May 26, 1955.

2nd Admission. The baby was admitted to Mt. Carmel Mercy Hospital in Detroit, Michigan on June 1, 1955. He now weighed 9 pounds, but the head had grown to 49 cm. in circumference. Otherwise the baby seemed more vigorous and alert than it had previously.

The left fontanelle tap on June 1, 1955 was dry, but 45 cc. of straw-colored fluid were removed from the right subdural space without depressing the fontanelle.

On June 3, 1955, 43 cc. were removed from the right subdural space through the anterior fontanelle, and at the same time 8 cc. of black-colored fluid was aspirated from within the calcified mass and replaced with an equivalent amount of air. Roentgenograms at this time showed the air confined within the calcified mass in a scattered fashion without outlining any single loculated cyst.

On June 7, 1955 a right subdural tap allowed removal of 20 cc. of straw-colored fluid. At the same time the right ventricle was tapped, and 5 cc. of clear colorless fluid were removed and replaced with air. Roentgenograms showed a dilated right ventricle displaced far to the right and not communicating with the left lateral or third ventricles (Fig. 2). It was of inter-

Fig. 1. Large suprasellar calcified tumor in infant at age of 9 days.
est incidentally that the mass had now shifted to the right of the midline whereas it was located primarily to the left of the midline in the first films taken when the infant was 10 days old. The tumor also appeared to have enlarged by about 25 per cent of its original size.

The likelihood of accomplishing anything worth while surgically in this infant was obviously remote. Yet there being nothing else to offer the patient or parents, the decision to undertake surgical removal of the tumor regardless of the consequences was made. Pre-

operative preparation with cortisone was thought unnecessary by the pediatrician now in attendance. The infant was now 7 weeks of age, weighed approximately 9 pounds, and the circumference of the head was 49 cm.

Operation. On June 11, 1955, under endotracheal anesthesia, through a small right frontal scalp flap an osteoperiosteal bone flap was turned down. A thin subdural hematoma and membranes were present beneath the dura mater and these were gently wiped away. The brain bulged moderately, but the convolutions were not flattened. A 1-inch cortical incision was made in the superior right frontal gyrus and at a depth of 1 inch the displaced right ventricle was entered. In apposition to its medial wall the hard surface of an encapsulated tumor was encountered. A frozen section biopsy revealed it to be a craniopharyngioma (Fig. 3). It was not cystic, but it shelled easily from surrounding brain tissue and was relatively avascular. Staying within the tumor it was relatively easy to remove large fragments of it without more than minimal hemorrhage. The infant tolerated the procedure well until removal of the last fragment of tumor. This fragment was the posterior shell of the tumor lying in apposition to the third ventricle and hypothalamic structures, all of which had been displaced far posteriorly to a position overlying the tentorium. The optic nerves and internal carotid arteries were markedly elongated, running posteriorly from their point of origin in the region of the sella turcica to an extent of about 1½ inches. The tumor had pressed into the sella turcica, but even so the optic nerves appeared to be more elongated than flattened out by pressure.
During removal of the tumor the left lateral ventricle was also entered and, as with the right ventricle, it was easily separated from the surgical field with cottonoid strips. The tumor had grown up between the lateral ventricles and separated them. At no place was the tumor adherent to surrounding brain or basal structures. When the final fragment of the posterior shell of the tumor was gently teased away from the third ventricle and hypothalamus, the infant immediately stopped breathing and could not be revived. There was no large hemor-

![Image](https://via.placeholder.com/150)

**Fig. 3.** Low-power magnification of tumor showing calcific areas and stratified squamous epithelium.

rhage at this or at any other time during the operation. The infant received about 300 cc. of blood during the 3-hour procedure as replacement for its estimated loss of blood.

**DISCUSSION**

The interest in this case centers around the occurrence at birth of an intracranial tumor. The presence of calcification in a tumor in a newborn, the incidence of this histological type at birth, and the concomitancy with bilateral subdural hematomata are by themselves unique features. Neonatal intracranial neoplasms are so uncommon, however, that the peculiarities of this case should not overshadow completely a consideration of this entire group. Arnstein et al.\(^1\) reviewed this subject in 1951 while reporting an intraventricular cavernous hemangioma in a 3-day-old infant. In a survey of the literature they were able to find 13 other cases of brain tumors occurring during the neonatal period (birth to 60 days). An additional search has disclosed 16 other cases of tumors manifesting themselves clinically before the age of
60 days, not included in their study or reported since that publication appeared. Together with this case, therefore, a total of 31 cases of neonatal intracranial neoplasms have now been found on record. In 10 of the 17 added cases the tumors were teratomas and most of these were reported in obstetrical literature as a cause of dystocia. Four of the 13 tumors in the review by Arnstein et al. were also teratomas. Thus, 14 of the 31 neonatal brain tumors reported were teratomas. Of this group, it is known that 9 produced dystocia and delivery was accomplished only after opening the head in utero. Five of the infants were born alive, 1 surviving, however, for only one-half hour. In the remaining 16 cases, exclusive of the 1 herein reported, 3 tumors were classified as probable medulloblastoma, 2 as sarcoma, and 1 each as dermoid, cystic blastoma, pineoblastoma, ependymoma, atypical polar spongioblastoma, glioblastoma multiforme, neuroepithelioma, choroid plexus papilloma, meningioma, cavernous hemangioma, and one simply as glioma.

It is known that 13 of the entire group of 31 patients were dead at birth or immediately thereafter. There were only 2, other than the 1 reported here, who underwent intracranial surgery. In 1 of these an enormous calcified parieto-occipital meningioma was only biopsied, and the patient expired 6 days later. In the other, the patient expired following the second choroid plexectomy for what was thought to be a communicating hydrocephalus. At autopsy a pineoblastoma was found. Only in the present case was an effort made for cure by total removal of the tumor. The specimens in all other patients were obtained at autopsy.

Although certain of the living patients presented focal neurological signs and symptoms, such manifestations were exceptional. The most common feature was an enlarging head which in several instances occurred with great rapidity. Many of the tumors, particularly the teratomas, were so large that intra-uterine perforation of the head was necessary to enable delivery. In these cases and in certain of the others the tumors had grown to such size as to nearly entirely replace brain. Tumors that were smaller showed no predilection for location. They were found within, embedding, and between hemispheres, within the mid-brain or the cerebellum, and several were intraventricular.

Besides the present case, there were 2 others in which the tumor showed calcification by roentgen ray. In 1 of these, a case of Cushing's, the patient was a 2-month-old infant who appeared normal at birth but a rapidly enlarging head developed. Roentgenograms revealed irregular calcified areas scattered throughout the cranial chamber. Aside from needling the lesion and draining off cystic fluid, no surgical procedure was done. The lesion at autopsy proved to be a huge teratoma. The other infant was one reported by Cuneo and Rand who was operated upon at the age of 3 months after a 2-month history of vomiting and opisthotonic spells. Roentgenograms showed a lacy calcification in the left parieto-occipital area with a superimposed, circumscribed thickened skull. The tumor proved to be a meningioma and after biopsy was considered too large for attempted removal. In neither of these cases, apparently, was the tumor and its extent as sharply delineated by the calcium as in the case reported here. The calcification and its apparent growth in this patient raise a provocative speculation concerning the fetal age when this tumor began. If during the 7 weeks of life the tumor enlarged in size by 25 per cent, one might presume by retrograde interpolation that the tumor at birth was at least 6 months old. That would fix the onset of tumor at less than 3 months' fetal age. Rathke's pouch first begins to develop in the fourth week of fetal life. From another point of view it is now evident that it is possible, at least in an infant, for a calcified tumor to grow
to the size of a baseball in less than 9 months and that the long duration usually attributed to most calcified intracranial tumors is not necessarily indicated. This is perhaps more valid for children in whom increased calcium metabolism in conjunction with growth processes is more apt to occur.

It appears, therefore, that of the tumors presenting at birth or shortly thereafter about half prove to be teratomas and often cause dystocia sufficient enough to force perforation of the head for delivery. Judging by this case and the experience of others the prognosis even with surgical intervention seems to be hopeless. Many of these cases, however, appeared before the availability of modern neurosurgical and anesthetic techniques.

Since several of the tumor types that occurred were enucleable lesions, with prompt recognition of the etiology surgical cure of certain neonatal brain tumors may yet be possible if they are accessible. Identification of the lesion may be made more easily if, as in some of these cases, calcification is present. A brain tumor, therefore, must be considered in the differential diagnosis in the presence of intracranial calcification in a newborn. Hydrocephalus that develops with great rapidity may be another clue indicating a tumor.

SUMMARY

A case report is made of a large calcified craniopharyngioma present at birth in conjunction with bilateral subdural hematomata. Its relationship to other neonatal brain tumors reported in the literature is discussed.

I wish to acknowledge with gratitude assistance given in the preparation of this report by Mrs. Clara Cziske.

REFERENCES