MENINGEAL SARCOMA IN A TWO-WEEK-OLD INFANT SIMULATING HYDROCEPHALUS

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Brain tumors in the newborn infant are rare and usually of such size that the condition is hope- less, should the diagnosis be made before death.

Arnstein et al.1 (1951), in a survey of brain tumors during the neonatal period, found 13 recorded cases of intracranial tumors occurring during the first 2 months of life.

In a more recent article, De Saussure et al.2 reported a case of astrocytoma in a 3-day-old infant found at autopsy, and recorded 24 cases from the literature in which the tumor was found in the neonatal period (first 60 days of life). They also included an additional 13 cases in which the tumor probably was present at birth but found beyond the neonatal period. In only 1 of the 37 cases was the tumor a sarcoma.

The purpose of our report is to add another case of an unusual type of tumor of extremely large size in a newborn infant, presenting as hydrocephalus, with a description of the clinical, diagnostic, operative and autopsy findings, and discussion of case reports in the literature.

CASE REPORT

History. A 2-week-old female was referred to WBGH because of rapidly progressive hydrocephalus and vomiting. The circumference of the head had increased 2 in. in the first 2 weeks of life. The baby appeared to be normal at birth and the pregnancy and delivery were uncomplicated.

Examination. Weight was 6.5 lbs. Temperature was 99.2°F, pulse rate 180, and respiratory rate 24/min. She was thin and slightly dehydrated. Activity was sluggish and the cry was weak. Circumference of the head measured 16.5 in. Veins of the scalp were prominent and fontanelles bulging. Sutures were separated on palpation. No evidence of papilledema was noted.

Hematocrit was 53 per cent, and hemoglobin 16.6 gm. There were many white blood cells in the urine. Cerebrospinal fluid was xanthochromic with a protein of 220 mg. per cent; it showed 29 white blood cells, 13 polymorphonuclear leucocytes, and 16 lymphocytes.

Course. On the day after admission, an attempt was made to perform a dye test to establish communicating or noncommunicating hydrocephalus. Subdural taps were done first and were negative. The needle was advanced through the region of the right coronal suture but the suspected dilated ventricle was not encountered. Instead there was moderate brain resistance to the sharp 18-gauge spinal needle. The needle was re-directed laterally, posteriorly, anteriorly, and medially and inserted to various depths but no ventricular fluid was found and the resistance of the brain tissue was similar throughout. One additional puncture was made through the left coronal suture but no ventricle was encountered in its usual location.

The patient then was placed in the sitting position and pneumoecephalography with 15 cc. air was carried out (Fig. 1). No apparent ill effects resulted from the air study. Therefore the proposed craniotomy was delayed 24 hours.

Operation. Right parietofrontal craniotomy was performed under endotracheal anesthesia. When the scalp flap was turned, a caput medusae of the parietal bone was quite striking. The bone in this area was slightly more elevated and very vascular. The dura mater was reflected in the parietal area and when the extensiveness of the tumor disclosed the problem to be hopeless, only a biopsy was taken, which was reported as meningioma, and the wound was closed.

The baby remained in a semicomatose state and died within 24 hours.

Autopsy. The body was that of a 17-day-old Caucasian female infant, measuring 52 cm. in length, and weighing 2935 gm. The abnormal findings were confined to the head and the brain.

The head measured 39.5 cm. in circumference, with an anteroposterior diameter of 19.5 cm. and a transverse diameter of 10 cm. A recent surgical incision was present in the right parietal region. The cranial sutures showed marked spreading.

On opening the cranium, the dura mater in the right parietal region was found to be adherent to the underlying structures. The major portions of the right parietal and temporal lobes, as well as the anterior portion of the right occipital lobe and the posterior portion of the right frontal lobe, were replaced by a tumor, which measured 10 cm. in anteroposterior diameter, 9 cm. in vertical diameter, and 6 cm. in transverse diameter. It extended to within about 2 cm. of the tips of the right frontal and occipital lobes, to the tip of the right temporal lobe, and to within 1.3 cm. of the superior longitudinal fissure.

On cut section, the tumor was a homogeneous light gray, with a dense, fibrous consistency. Several areas of hemorrhage and softening were present, with the portion of the tumor which protruded into the posterior portion of the right lateral ventricle being quite necrotic and hemorrhagic. The right lateral ventricle was markedly compressed, and contained a small amount of

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clotted blood. The left cerebral hemisphere and the cerebellum were compressed and displaced to the left. The left lateral ventricle was moderately dilated (Fig. 2).

Microscopic sections (Figs. 3 and 4) showed considerable variation in the histologic pattern of the tumor. The greater portion was composed of thin, elongated cells, with indistinct cell borders and fibrillar, eosinophilic cytoplasm. The nuclei of these cells ranged from broadly oval to spindle-shaped, with distinct nuclear membranes, indistinct nucleoli, and finely granular chromatin. Marked variation in nuclear size was present, but mitoses were infrequent. There appeared to be some edema present, with empty clefts separating adjacent cells. The cells showed a distinct tendency to be aligned parallel with the cells adjacent to them, forming poorly defined bundles of cells, with the bundles interlacing with one another in a swirled pattern.

DISCUSSION

In 1933, Russell and Ellis\(^5\) reported 3 circumscribed cerebral tumors in young infants and discussed the clinical and autopsy findings. Two of the tumors were sarcomas: 1 in a 5-month-old infant and 1 in a 23-month-old infant. They pointed out the rarity, difficulty in histologic diagnosis, and problems of early diagnosis of cerebral tumors in young infants because of the readiness with which the expansile skull of the infant compensates for raised intracranial pressure. From the macroscopic features of such tumors, they believed early recognition of the condition followed by radical surgical treatment might be of benefit to the patient.

In the survey of brain tumors during the neonatal period by Arnstein et al.,\(^4\) they found that most of the tumors occurring in this period are supratentorial. It also was noted that hydrocephalus was the most frequently encountered feature and in 4 of the 18 patients was unusually rapid. They also believed that the presence of these lesions during this very early period of life