Congenital Ependymoma

Case Report

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The congenital occurrence of an intracranial tumor is a rarity. A review by Solitare and Krigman in 1964 uncovered only 45 intracranial tumors believed to be present prior to or at the time of birth. This paper adds one additional case which appears to be the only reported congenital ependymoma of the fourth ventricle.

Case Report

This white male infant was the product of an uncomplicated pregnancy and was delivered vaginally after 5 hours of labor. He was considered normal at birth and was discharged on the third day of life. At 2 weeks of age he was adopted and apparently remained well until he was 6 weeks old, when progressively severe vomiting developed. Two weeks later he was seen by a physician, who noted cranial enlargement.

Examination. On admission to the UCLA Hospital, the patient was an alert 9-lb baby boy whose head was abnormally large for his age. The head circumference was 43.8 cm; the cranial sutures were separated, and the anterior fontanelle was tense but pulsatile. The head did not transilluminate, and the general physical examination was otherwise normal. Further neurological examination showed no cranial nerve abnormalities, and the optic disc margins were distinct. Primitive reflexes were normal for age as was the examination of the extremities. The preliminary diagnosis was idiopathic hydrocephalus.

Laboratory studies revealed a normal hemogram, urinalysis, and EEG. Skull films showed an enlarged head with separated sutures but no abnormalities of localizing value. Pneumoencephalography demonstrated symmetrical hydrocephalus with 1.5 cm of frontal cortex, a dilated third ventricle, and a questionable mass in a dilated fourth ventricle. The cerebrospinal fluid was xanthochromic. Subsequently, a ventriculogram showed a lobulated fourth ventricular mass in clear detail.

First Operation. Posterior fossa exploration was undertaken but discontinued when vigorous venous bleeding from the dura caused a momentary loss of blood pressure. Immediate recovery was uncomplicated. However, increasing intracranial pressure soon necessitated a ventricular puncture. Because the ventricular fluid protein was 800 mg%, ventricular fluid exchange with lactated Ringer's solution was undertaken on 4 successive days, lowering this value to 100 mg%.

Second Operation. One week after the attempted exploration, a jugular shunt was placed; this functioned well, and over the next 2 weeks the patient's condition improved steadily.

Third Operation. At this time reexploration of the posterior fossa was performed. A large, vascular, beefy-red tumor was encountered, obliterating the cerebellar vermis and replacing the bulk of both cerebellar hemispheres. During partial removal the child suffered episodic circulatory collapses, and, postoperatively, artificial respiration was required to sustain life. The patient lapsed into coma and died on the first postoperative day. He was then 14 weeks old.

Autopsy Findings. The pertinent pathological findings were confined to the central nervous system, and no evidence of neoplasia was found elsewhere. The operative site was obscured by hematoma, and the spinal subarachnoid space showed no evidence of metastases but was filled with blood. There was symmetrical hydrocephalus of the lateral ventricles and a conspicuous space (cavum) within the septum pellucidum. The shunting device was well placed. The tumor arose from the floor of the fourth ventricle at the level of the lateral recesses. Its central portion, which had obliterated the vermis, had been surgically removed; but the lateral extensions replaced the medial halves of the cerebellar hemispheres. The underlying medulla and

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caudal pons were severely distorted and compressed ventrally toward the clivus. The tumor mass was approximately 5 cm in diameter and its cut surface was deep red in color and of a granular texture.

Sections of the brain stem and cerebellum demonstrated a neoplastic lesion filling the fourth ventricle. Although there was marked ventral compression of the brain stem, little tendency toward invasion of the underlying nervous parenchyma was seen. At the rostral medullary level, the right half of the medulla was well preserved except for tegmental edema. On the left side of the medulla, however, the tumor had superficially invaded the ventricular floor and replaced the left vestibular nuclei and medial restiform body. At the pontine level, the right fourth ventricular floor was intact, but tumor had replaced the medial portion of the left brachium pontis and contiguous cerebellar tissue. No evidence of tumor was found in the third or lateral ventricles, or in the spinal subarachnoid space.

The tumor was highly cellular and well vascularized (Figs. 1 and 2). The densely packed tumor cells exhibited slight-to-moderate pleomorphism and occasional mitoses. Tumor giant cells were rare. Most of the tumor cells were slightly larger than normal ependymal cells and were elongated, polyhedral, or rounded. The nuclei contained a small-to-moderate amount of fine chromatin particles and an occasional single nucleolus. The cytoplasm was generally scant and acidophilic. In many places, the tumor cells were grouped in a characteristic radial formation around capillary walls and were elongated or cylindrical. There were occasional rosettes of the Flexner type. The tumor was diagnosed as a high-grade, cellular ependymoma.

Discussion

This tumor was definitely present at 6 weeks of age and, in all likelihood, at birth. The histology is that of a cellular ependymoma arising in the fourth ventricle. Reference to the literature reveals that this is indeed a rare finding.

In several extensive studies dealing with intracranial neoplasms of infancy and childhood, ependymomas have not been found in the first months of life.6–8,11 Cuneo and Rand6 cited seven cases, the youngest being 17 months. Bailey, et al.,8 reported one case at 6 months of age, as did Stern.17 Kricheff, et al.,10 referred to a case at 4 months of age.

The review of congenital intracranial tumors by Solitare and Krigman16 cited no case
of congenital ependymoma. However, close examination of the original papers revealed four cases that may have been ependymal neoplasms. Peace reported the death of a 3-day-old infant with a right cerebral hemisphere neoplasm. Histological study demonstrated a cellular tumor interpreted as an hemangiopericytoma, but cellular ependymoma was listed as a possibility. Braune described a left hemispheric tumor in a 14-week-old infant which was labeled glioma and may have been ependymal. Alpers reported a fourth ventricular neoplasm in a 5-week-old infant, and Globus cited a “neuroepithelioma” in a 2-month-old child. Neither case is described well enough to draw definite conclusions.

Our review reveals four cases not alluded to previously. Klein cited an infant, symptomatic at 4 weeks of age, in whom a large left cerebral hemisphere tumor was found at autopsy. Histologically this was well described as a cellular ependymoma. His second case was an apple-sized tumor of the right temporal lobe in a child a few weeks of age which was also described as ependymal in origin. Saito reported an infant symptomatic at 1 month in whom a right lateral ventricle tumor was found and histologically shown to be an ependymoma. Luyendijk and Staal described a right cerebral hemisphere neoplasm, probably an ependymoma. These cases bear out Zülch’s contention that ependymal neoplasms in infancy are usually cerebral in location.

It seems very likely that our patient had a congenital ependymoma; no other case justifying that diagnosis has been reported in English language journals. The difficulty in making this diagnosis during life has previously been stressed and was emphasized by our experience.

Summary
We have reported the details of a case in which a verified ependymoma of the fourth ventricle had caused symptoms of obstructive hydrocephalus by the time the child was 6 weeks old. We have reviewed related points.

References


