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Radiologic-Pathologic Correlation
Congenital Choroid Plexus Papillomas

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Patient History

At 33 weeks of pregnancy, a 29-year-old woman presented with uterine contractions. A transabdominal obstetrical sonogram with Doppler revealed severe hydrocephalus secondary to a large avascular third ventricular mass (Fig 1A). Delivery by cesarean section at 34 weeks revealed marked macrocephaly, widely separated sutures, and tense fontanelles. Contrast-enhanced computed tomography (CT) of the infant's brain revealed an enhancing slightly corrugated mass in the third ventricle with dilatation of the lateral ventricles, intraventricular hemorrhage (Figs 1B, 2, and 3), and an occipital subgaleal collection, which communicated with the left lateral ventricle consistent with a pseudomeningocele (Figs 2–4A). Surgery was not performed because of the patient's poor medical condition. At 10 days of life, the occipital pseudomeningocele spontaneously ruptured, causing leakage of cerebrospinal fluid (CSF) onto the scalp, an event that ultimately led to the infant's death.

Autopsy revealed a 2.5 × 2 × 2-cm discrete spherical mass in the posterior third ventricle. The mass was compressing the midbrain and aqueduct, causing severe secondary obstructive hydrocephalus. (Figs 2, 3, and 4B). Microscopic sections revealed a typical choroid plexus papilloma consisting of papillary fronds simulating a normal choroid plexus. There were multiple segmented vascularized fibrous stroma lined by a single layer of cuboidal epithelium with scattered areas of hydropic degeneration (Fig 4C).

Discussion

Choroid plexus papillomas are primary intraventricular neoplasms, accounting for 3% of intracranial neoplasms in children and 0.6% in adults (1). Although the mean patient age is 5.2 years (2), these tumors may be present at birth, accounting for 42% of pediatric tumors that present within the first 60 days of life. In children younger than 2 months of age, choroid plexus papillomas account for 42% of neonatal brain tumors, whereas teratomas and hypothalamic gliomas are found in 25%, and gliosarcomas, medulloblastomas, and primitive neuroectodermal tumors are found in 8.9% each (3). It has been suggested that because most choroid plexus papillomas present as large tumors at a young age, they may represent a true congenital neoplasm of the brain (4–6). The following are common features of choroid plexus papillomas:

Age	Peak in sixth year
Sex	Male predilection
Location	Atrium of lateral ventricle > fourth ventricle > third ventricle

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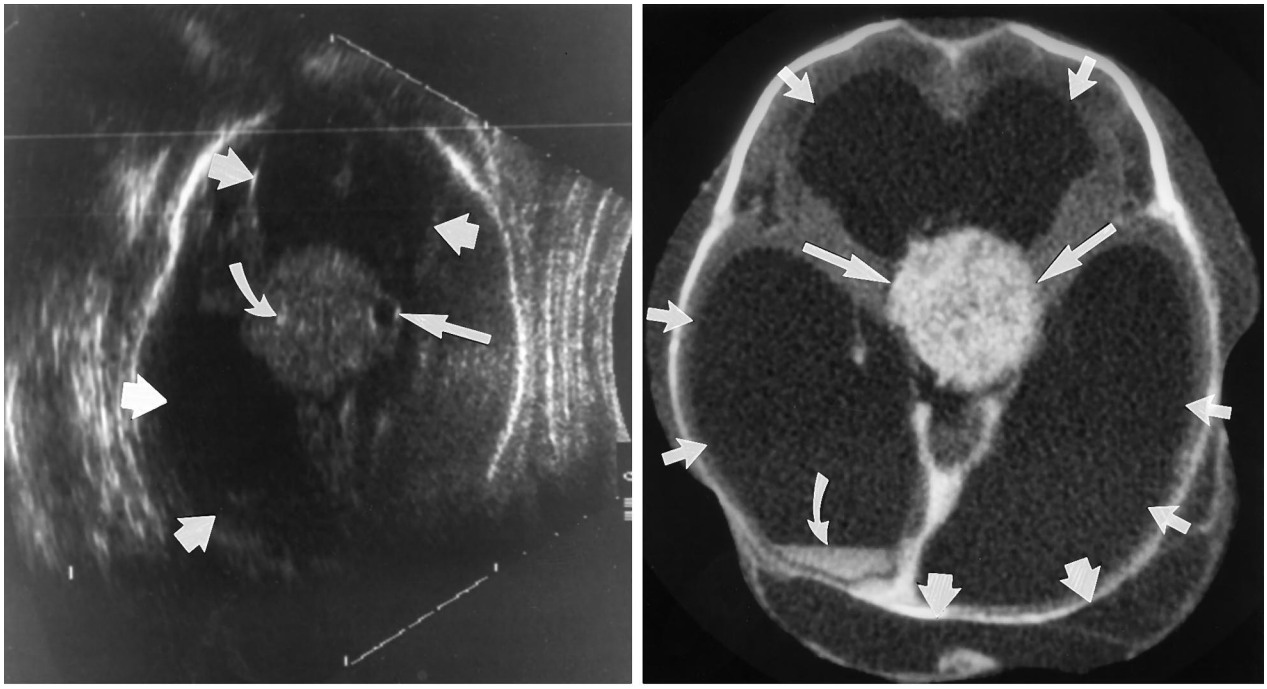


A B

Fig 1. *A*, Prenatal transabdominal ultrasound in the axial plane shows a large mass in the area of the third ventricle with an eccentric cystic area (*straight arrow*) and an echogenic focus (*curved arrow*). There is marked dilatation of the frontal horns and atrium of the lateral ventricles (*short arrows*).

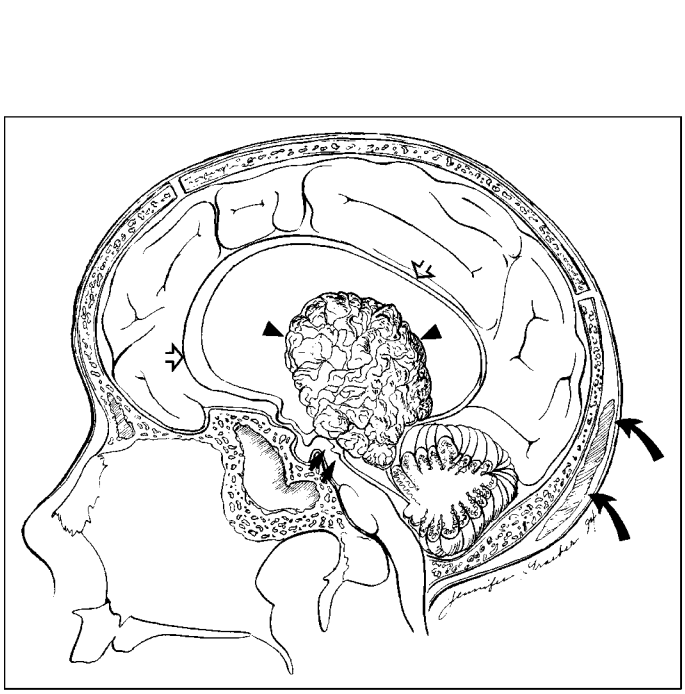
B, Contrast-enhanced axial CT reveals a large, heterogeneous mass (*arrows*) in the area of the third ventricle with dilatation of the lateral ventricles (*short thin arrows*). There is a blood-fluid level in the right lateral ventricle (*curved arrow*). There is a dorsal pseudomeningocele (*short wide arrows*).

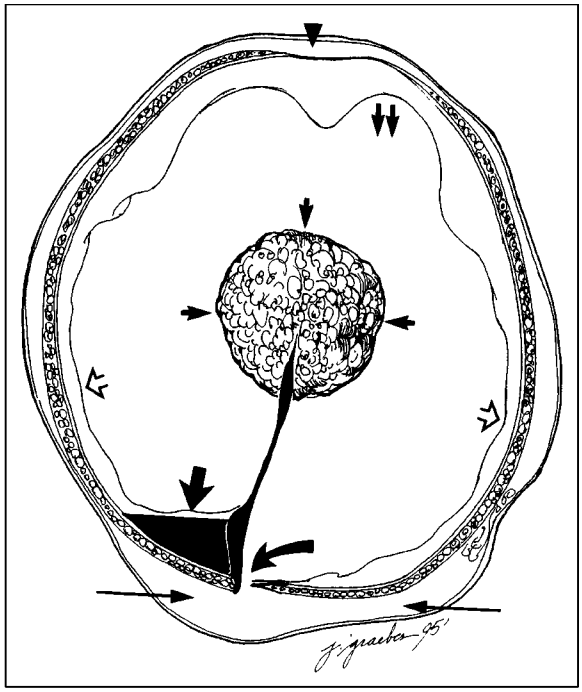
Echogenicity on ultrasound	Highly echogenic
Density on CT	Hyperdense
Enhancement on CT	Homogeneous intense enhancement
Gross pathology	Lobulated encapsulated masses within ventricular system
Histology	Epithelial lined villi with calcification

Choroid plexus neoplasms arise from the epithelial cells of the choroid plexus and may originate wherever a choroid plexus is found within the ventricular system. They are highly vascular tumors. In childhood 80% of choroid plexus papillomas arise in the lateral ventricle, 16% in the fourth ventricle, and 4% in the third (7). The incidence of third-ventricular choroid plexus papillomas is higher in the first decade (29.5%) of life, with a female predominance, although overall these tumors are more common in male subjects (8).

Hydrocephalus is a common feature of papillomas. It may be caused by mechanical obstruction of CSF pathways, adhesions because of bleeding from the tumor, overproduction of CSF by the papilloma, or a combination of these (4, 9, 10). When the choroid plexus papillomas are within the third ventricle, hydrocephalus seems to be caused by obstruction of CSF outflow at the foramina of Monro. Sudden death because of acute ventricular obstruction by choroid plexus papillomas of the third ventricle has been reported (11).

Most choroid plexus neoplasms arise from the epithelial cells inside the ventricles, as in our case. However, extraventricular growth is seen in several circumstances: direct extension of a primary intraventricular papilloma, drop metastases, and development of a primary tumor from the choroidal tuft at the foramen of Luschka (12). From the foramen of Luschka, these tumors may extend to the cerebellopontine angle cistern or to the foramen magnum, where they may compress the brain stem (6, 13). Other extraventricular





2 3

Fig 2. Midline sagittal anatomic diagram demonstrates the pathologic findings in a third-ventricular choroid plexus papilloma. The tumor occupies much of the expanded third ventricle with secondary thinning of the corpus callosum (*open arrows*). There is compression of the midbrain by the mass (*short arrows*). The frondlike appearance of the tumor is seen (*arrowheads*). The occipital pseudomeningocele is also depicted (*curved arrows*).

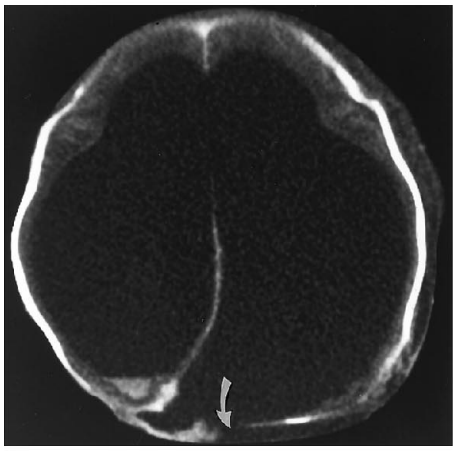
Fig 3. Axial anatomic diagram confirms the findings in a third ventricular choroid plexus papilloma. The tumor is seen in the area of the third ventricle and has a frondlike appearance (*short arrows*). The dilated lateral ventricles (*double arrows*) compress the thinned cortical mantle (*open arrows*) against the calvarium. The defect in the posterior calvarium (*curved arrow*) allows communication of the lateral ventricle with the occipital pseudomeningocele (*long arrow*). The blood-CSF level (*thick arrow*) is seen in the dependent portion of the right lateral ventricle. The anterior fontanelle is patent (*arrowhead*).

sites have been reported in the brain parenchyma, pineal area, and suprasellar area (14).

Treatment for choroid plexus papillomas involves shunting of the hydrocephalus and surgical resection of the tumor (7). Although papillomas are histologically benign, postoperative recurrence of tumors has been seen (12). Complications such as seeding of benign papillomas within the subarachnoid space can occur, and secondary implants may be detected far from the original tumor (2). Overall, the long-term prognosis has improved, with high survival rates, although in some patients seizures or mental retardation may develop. Radiation therapy after surgical intervention is generally reserved for those patients with choroid plexus carcinomas (7, 15, 16).

Radiologic-Pathologic Correlation

Ultrasound. On ultrasound, large choroid plexus papillomas appear as highly echogenic masses with irregular borders related to the glomus of the choroid plexus and are associated with hydrocephalus (Figs 1A, 2, and 3). When they are located in a lateral ventricle and cause unilateral hydrocephalus, they are easier to diagnose. However, when located in the third ventricle, as in our case, choroid plexus papillomas can mimic aqueductal stenosis, and diagnosis may prove difficult, particularly in the prenatal period, unless the echogenic mass is visible (1). Occasionally, intratumoral cysts may also be seen on ultrasound in choroid plexus papillomas (3), as was seen in our case. These intratumoral cysts appear on ultrasound as hypoechoic areas (Fig 1A) and represent ar-



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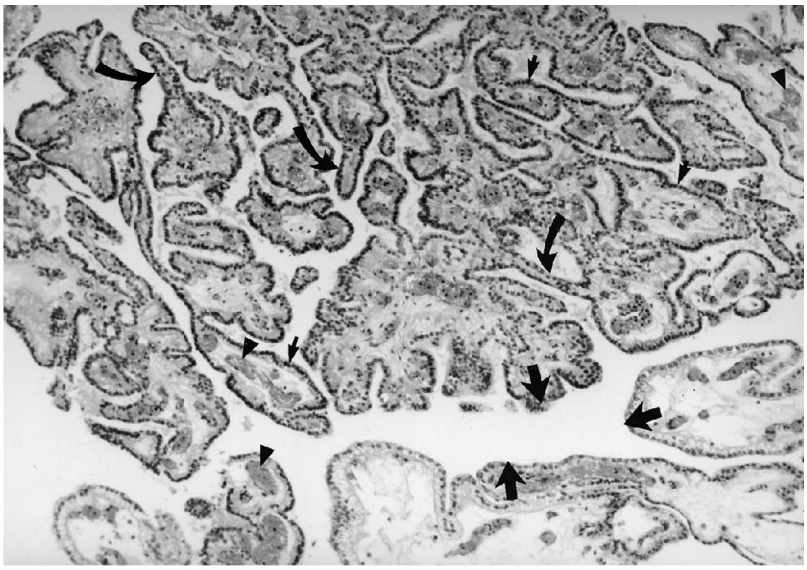

C

B

Fig 4. *A*, CT through a higher level reveals a communication (*arrow*) between the left lateral ventricle and the pseudomeningocele through the posterior fontanelle.

B, Grossly, a horizontal cut through the brain stem (removed) at the level of the mamillary bodies reveals a discrete spherical mass (*short arrows*) primarily occupying the third ventricle with marked dilatation of the lateral ventricles (*long arrows*). There is a central slitlike cavity (*curved arrow*), which may correspond with hydropic degeneration. The frondlike appearance of the mass is seen (*white arrows*). The septum pellucidum is noted (*thin arrow*).

C, Microscopic section of the tumor demonstrates papillary fronds (*curved arrows*) consisting of branches of highly vascularized fibrous stroma (*arrowheads*) lined by a single layer of cuboidal epithelium (*small arrows*). Hydropic changes are noted (*straight arrows*) (hematoxylin and eosin, magnification × 200).

eas of hydropic degeneration (Figs 4A and B). If calcifications are present in the tumor, echogenic foci with shadowing may be seen. The nonvisualization of microscopic calcifications in our case may be attributable to sampling.

Typically, Doppler examination will show a biphasic flow on choroid plexus papillomas because of their highly vascular nature. This finding has been thought to be specific for choroid plexus papillomas (6). However, in our case Doppler ultrasound was hypovascular. This difference may be explained by the fact that pathologic examination revealed no large, discrete feeding vessels.

CT. On CT, choroid plexus papillomas may appear as well-defined, homogeneous, usually hyperdense intraventricular masses. Tumor margins can be smooth (29%), lobulated (19%), or irregular (52%). Irregular margins are found in 71% of malignant tumors and 43% of benign lesions. Papillomas are isodense or hyperdense to parenchyma in 75% of cases, and 25% are hypodense or heterogeneous (17). Tumor calcification on CT has been reported in 24% of choroid plexus tumors (2). Hemorrhage within the tumor may also be seen. A mottled appearance seen in the majority of choroid plexus papillomas is caused by CSF trapped between the papillae (8) (Figs 4A and B). Choroid plexus papillomas may show limited parenchymal invasion (16), and it may be difficult to distinguish benign from malignant choroid tumors on imaging studies.

The differential diagnosis of third-ventricular tumors in children includes choroid plexus papilloma, glioma, craniopharyngioma, colloid cyst, ependymoma, teratoma, dermoid, epidermoid, metastasis, and tumors deposited by CSF seeding. Metastases are unlikely in a young patient. Colloid cyst, meningioma, epidermoid, dermoid, teratoma, and most craniopharyngiomas can be excluded by their CT and magnetic resonance features (2, 9, 18, 19). Supratentorial ependymomas usually have an intraventricular portion smaller than the component of the tumor mass invading the parenchyma. Choroid plexus papillomas and gliomas posses similar features and may be confused with each other radiographically (2).

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