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Congenital Choroid Plexus Papilloma of the Third Ventricle: Diagnosis with Real-Time Sonography and MR Imaging

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Congenital neoplasms of the choroid plexus are uncommon. A case of a surgically resected congenital third-ventricular choroid plexus papilloma is presented to demonstrate the appearance of this tumor using real-time sonography and MR imaging. The sonographic features of this tumor are reviewed and compared with previous reports. The MR characteristics are correlated with sonographic findings, with gross pathologic features, and with histology. A review of choroid plexus papilloma presenting within the neonatal period is also provided.

Case Report

A 3920-g male infant was born at 41.5 weeks gestation by spontaneous vaginal delivery. Physical examination was unremarkable except for moderate enlargement of the head circumference. Neurologic evaluation was normal. Real-time sonography revealed marked enlargement of the lateral ventricles, with a homogeneous, echogenic mass occupying the enlarged third ventricle and obstructing the cerebral aqueduct (Figs. 1A and 1B). An unenhanced CT scan revealed a homogeneously dense mass within the third ventricle (images were degraded by motion). The diagnosis of choroid plexus papilloma was suggested from the combined sonographic and CT findings. MR, performed with the baby under general anesthesia, sharply defined the third-ventricular neoplasm and allowed a confident preoperative diagnosis of choroid plexus papilloma (Figs. 1C and 1D).

A right frontal craniotomy was performed. Right frontal frontotomotomy and lateral ventricular approach, using 6× loupes, revealed a pink, soft, friable, papillary neoplasm filling the enlarged third ventricle and bulging into the foramina of Monro. The tumor was easily removed from the third ventricle and appeared to be totally removed from its choroidal origin at the venous angle. Histologic evaluation revealed a typical choroid plexus papilloma (Fig. 1E).

Postoperatively, the patient has been neurologically normal; however, a left frontal ventriculoperitoneal shunt was required 5 weeks after initial surgery because of communicating hydrocephalus, demonstrated by sonography. Follow-up MR at 9 weeks, performed with intramuscular Nembutal sedation, demonstrated complete absence of residual tumor and some ventricular decompression (Fig. 1F).

Discussion

Choroid plexus papilloma is an uncommon epithelial neoplasm of the CNS, accounting for 3–5% of brain tumors in children [1–9]. Twenty percent of all choroid plexus papillomas occur in patients under the age of 1 year. In a large series, these tumors represented 12.5% of verified neoplasms of the CNS [2].

Previous reports of sonographic findings of choroid plexus papilloma describe highly echogenic intraventricular mass lesions with irregular borders and associated hydrocephalus [10, 11]. Doppler findings have been described and may contribute significantly to the knowledge of tumor vascularity [10]. Although Doppler was not performed in our case, pulsatile vascular channels were observed within the mass during real-time sonography.

At the time this paper was submitted for publication, there were no published reports of the MR appearance of choroid plexus papilloma in the neonate. The MR signal intensities in this case are entirely nonspecific, showing intermediate T1 and long T2 signal characteristics. The signal intensity from the neoplasm is quite uniform (Figs. 1C and 1D). Prominent vessels are seen within the tumor mass, proved at surgery to be venous channels draining posteriorly into the internal cerebral veins.

The presence of marked third-ventricular expansion around the intraventricular neoplasm provides a major clue toward the diagnosis of choroid plexus papilloma (Figs. 1B–1D).

The differential diagnosis of intraventricular masses in the neonate must include hematomas and other less common lesions, such as astrocytoma, ependymoma, colloid cyst, and meningioma [2, 4]. MR will substantially enhance our ability to differentiate these specific lesions through a combination of morphology and signal intensity. Certainly, hematomas and colloid cysts will have strikingly different signal characteristics and appearances compared with other solid lesions. Cerebral arteriography may provide important diagnostic clues in difficult cases [4].

Postoperative follow-up sonography is a valuable examination that can be performed without sedation, with a minimum of patient discomfort and expense. In light of cost factors and risks to the patient, sonography is clearly the examination of choice for initial screening of the neonate and for routine follow-up. MR may be used whenever a mass is identified, in cases of occult hydrocephalus, or whenever more detailed
information than sonography can provide is required. MR can be performed successfully with oral, rectal, or intramuscular sedation in most circumstances.

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REFERENCES