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Choroid Plexus Papillomas of the Foramen of Luschka: MR Appearance

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Choroid plexus papillomas are rare intracranial neoplasms that are most commonly found in the trigone of the lateral ventricles or in the fourth ventricle. Rare cases of extraventricular choroid plexus papillomas have been reported within the cerebellopontine (CP) angle cistern [1–6]. Such cases have been described as either extending from the fourth ventricle via the foramen of Luschka or arising directly from the CP angle cistern.

We report MR findings in two unusual cases of choroid plexus papillomas that appeared to arise from within the foramen of Luschka without an associated intraventricular component. In the first case, the tumor presented as an expansile mass of the foramen of Luschka, whereas in the second case the tumor arose from the foramen of Luschka and extended to the CP angle cistern.

Case Reports

Case 1

A 52-year-old woman presented with a 2-month history of progressive headaches and ataxia. Physical examination was unremarkable. T1-weighted MR images revealed a mass of low signal intensity expanding the right foramen of Luschka (Fig. 1). The mass appeared as inhomogeneous high signal intensity on intermediate and T2-weighted images and enhanced homogeneously on T1-weighted images obtained after administration of gadopentatate dimeglumine. Moderate hydrocephalus was present without evidence of ventricular obstruction. At surgery, a choroid plexus papilloma was found expanding the foramen of Luschka and was completely resected. The patient's hydrocephalus resolved postoperatively, but recurred several months later, necessitating shunt placement.

Case 2

A 55-year-old woman presented with a 2-year history of progressive dizziness and clumsiness in writing with her right hand. Physical examination was normal except for right finger-to-nose difficulty. T1-weighted MR images showed a multilobulated mass of low signal intensity expanding the right foramen of Luschka and extending to the right CP angle cistern (Fig. 2). The mass appeared inhomogeneously bright on intermediate and T2-weighted images. On T1-weighted images obtained after administration of gadopentatate dimeglumine, the mass enhanced markedly except for persistent components of low signal intensity. Subsequent CT scans showed the mass to contain multiple foci of calcifications of variable size.

At surgery, a choroid plexus papilloma was found arising from within the foramen of Luschka. The tumor was very vascular.

Discussion

Choroid plexus papillomas are rare, histologically benign intracranial neoplasms that typically arise from an intraventricular location. They occur commonly in children and less frequently in adults. They represent 3–5% of intracranial tumors in children and 0.5% in adults [7–9].

These tumors occur most frequently in the lateral ventricles followed by the fourth and third ventricles in order of decreasing frequency [7–9]. In children they are found most often in the trigone of the lateral ventricles, whereas in adults they are more likely to be found in the fourth ventricle. Involvement of the CP angle cistern occurs in up to 9% of choroid plexus papillomas, either from direct extension of a fourth ventricular papilloma, from distant CSF seeding, or as a primary CP angle lesion possibly arising de novo from embryonic choroidal remnants [1–5, 10]. In a recent review of the literature, Martin et al. [1] found 20 reported cases of primary unconnected papillomas. Most of these were thought to have developed from the small choroidal tuft that normally protrudes from the foramen of Luschka. Both our cases appear to have arisen from this foraminal tuft. In the first case the tumor remained within the foramen as an expansile mass, whereas in the second case the tumor extended to the CP angle.

There have been two recent case reports of choroid plexus papillomas of the CP angle at MR imaging [1, 6]. In the first report, the tumor extended to the CP angle cistern from the fourth ventricle via the anterolateral recess and the foramen of Luschka [6]. This tumor was described as isointense with brain on T1-weighted MR images and as homogeneously enhancing on contrast-enhanced CT scans. T2-weighted im-

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In the second case report, the tumor arose de novo in the CP angle and extended into the adjacent cavernous sinus [1]. This latter tumor was described as isointense with high signal hemorrhagic foci on T1-weighted images and as heterogeneously hyperintense on T2-weighted images. On CT studies, the tumor appeared calcified and moderately dense on precontrast scans and enhanced markedly on postcontrast scans. Both our cases were hypointense relative to brain on T1-weighted images, heterogeneously hyperintense on T2-weighted images, and enhanced brightly on contrast-enhanced studies.

At CT, choroid plexus papillomas are usually homogeneous in appearance in children, but inhomogeneous in adults, often related to cystic degeneration or calcification [11]. Tumoral calcification has been reported in 4.1% [9] to 20% of intraventricular papillomas, but has been more frequent in the rarely reported CP angle papillomas [1, 12]. Multifocal calcifications were present in one of our two cases.

Clinically, patients with choroid plexus papilloma of the CP angle usually present with ataxia and palsy of the fifth, seventh, or eighth cranial nerves. Both our patients had forms of ataxia, but neither manifested a cranial nerve deficit. Hydrocephalus commonly occurs in patients with choroid plexus papilloma and results from overproduction of CSF [9, 13]. Additionally, hydrocephalus in these patients may be due to ventricular outlet obstruction resulting from the tumor mass itself or from prior hemorrhage and adhesions [3, 6, 9, 14]. Hydrocephalus that could not be explained by ventricular outlet obstruction was a clue to the correct diagnosis in the first case.

The differential diagnosis for similarly located lesions on MR images include hemangioblastoma, ependymoma, meningioma, neurinoma, giant aneurysm, and metastatic disease. Hemangioblastomas commonly demonstrate serpentine flow voids caused by prominent feeding vessels and often have a prominent cystic component [15]. Aneurysms similarly demonstrate either flow characteristics or evidence of thrombosis at MR [16, 17]. Ependymomas of the fourth ventricle more commonly arise during childhood and tend to have more irregular margins with surrounding edema [18]. Meningiomas of the CP angle generally have a broad dural attachment and are more commonly isointense with gray matter on T2-weighted images [19, 20]. Neurinomas of the eighth cranial nerve extend from the internal auditory canal whereas those of the fifth nerve can be seen to arise along its more rostral course [21]. Both neurinomas and meningiomas may demonstrate calcification and a cystic component.

Finally, interventricular extension has been stressed as a sign of choroid plexus papilloma [2, 22]. Extension through ventricular outlet foramen should not be confused with exten-
sion outside of the ventricle across its ependymal lining, a sign of choroid plexus carcinoma [23].

In summary, the diagnosis of choroid plexus papilloma should be considered in an adult with an expansile mass at the foramen of Luschka that expands the foramen or extends to the CP angle. The association of nonobstructive hydrocephalus is a helpful clue to the correct diagnosis.

REFERENCES

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