Third ventricular hemangioblastoma: MR appearance.

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Third Ventricular Hemangioblastoma: MR Appearance

Supratentorial hemangioblastomas of the third ventricle are particularly rare lesions. Of the three previously reported cases [1–3], one was associated with von Hippel-Lindau syndrome [1]. We present a fourth case and include the first published description of the MR appearance of third ventricular hemangioblastoma.

Case Report

A 15-year-old boy had an acute, severe change in personality and deterioration in mental status over a period of 2 weeks. Peculiar mannerisms and incoherent speech developed, and he had episodes of prolonged staring and seizures. He became withdrawn and solitary, and subsequently developed urinary and fecal incontinence. Routine laboratory studies were unremarkable. A CT scan showed a large lobulated contrast-enhancing lesion in the midportion of the third ventricle. The lesion was 4–5 cm in diameter and enhanced homogeneously. No evidence was seen of calcification or of a cystic component. MR imaging with IV gadopentetate dimeglumine showed a mass in the posterosuperior aspect of the third ventricle. The mass was inhomogeneous in appearance and approximately isointense with gray matter on noncontrast T1-weighted, proton-density, and T2-weighted images (Fig. 1). Bilateral periventricular edema was noted adjacent to the mass on T2-weighted images. After administration of gadopentetate dimeglumine, the mass enhanced intensely and homogeneously [2].

The tumor was resected via a transcallosal approach. The histologic specimen was characterized by numerous small blood vessels, with sparse intervening polygonal stromal cells. Lacy reticulin fibers were present, and the cytoplasm was vacuolated, consistent with a high lipid content. These findings were typical for hemangioblastoma. Angioblastic meningioma sometimes mimics hemangioblastoma histologically, but the absence of meningocytic whorls or psammoma bodies suggested that the tumor was not a subtype of meningioma.

Discussion

Hemangioblastoma is typically a tumor that becomes symptomatic in the fourth decade of life, and it is associated with von Hippel-Lindau disease in 15–20% of cases. Cases of this tumor account for less than 2% of intracranial neoplasms [4]. Multiple hemangioblastomas occur in 20% of patients who have von Hippel-Lindau disease and in 5% of otherwise normal patients. The prevalence of spinal hemangioblastoma in patients who have von Hippel-Lindau disease ranges from 8% to 35% [5].

Hemangioblastomas usually are located in the cerebellum. They are typically well circumscribed but lack a true capsule. About 60–70% of the tumors consist of a small hypervascular mural nodule with a cystic component. Solid tumors are more common in the supratentorial compartment [6].

Supratentorial hemangioblastomas are extremely rare and usually occur along the convexities, although they have been reported in numerous additional sites. The majority of these cases have been associated with von Hippel-Lindau disease [6]. Only three third ventricular hemangioblastomas have been reported previously, and of these, only one was associated with von Hippel-Lindau disease [1–3]. None of the tumors had a cystic component.

Previous reports indicated that the nodule always enhances either with iodinated contrast material on CT or with gadopentetate dimeglumine on MR. Serpentine vessels were well seen as flow voids against high-signal cyst or tumor on T2-weighted MR images [5]. In our case, the MR findings were nonspecific, and we did not identify serpentine vessels on T2-weighted images. No images were available from one [3] of the three previously reported cases, but the CT scan from the second case [2] showed a contrast-enhancing lesion in the anterior third ventricle with moderate hydrocephalus. Angiography revealed some vascular blush. In the third case [1], CT showed a large, isodense mass with a smooth border in the third ventricle, which enhanced homogeneously after administration of contrast material. Angiograms showed a homogeneous tumor stain. In our patient, the CT showed a solid, noncalcified mass 4–5 cm in diameter that enhanced homogeneously. As noted earlier, this is the first published description of the MR appearance of hemangioblastoma of the third ventricle. However, the MR features were nonspecific.

The differential diagnosis of an enhancing third ventricular mass is extensive, including ependymoma, choroid plexus papilloma, craniopharyngioma, germ cell tumor, meningioma, glioma, neurocytoma, oligodendroglioma, and metastasis [7]. We think it may be helpful to add hemangioblastoma to this list.

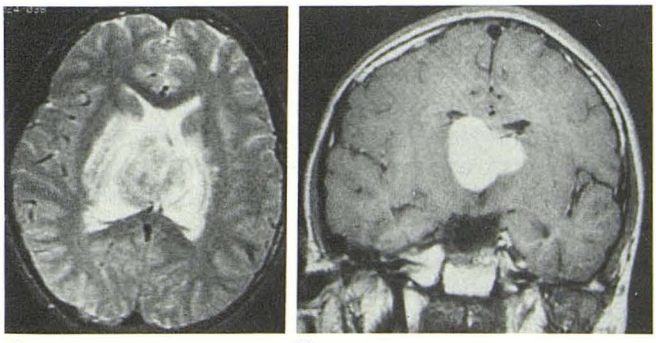

Fig. 1.—Third ventricular hemangioblastoma.
***A*, Axial T2-weighted MR image, SE 3000/80, shows a large heterogeneous iso- and hyperintense mass that displaces lateral ventricles laterally. Note surrounding periventricular edema.**
***B*, Coronal contrast-enhanced T1-weighted MR image, SE 800/20, shows a large homogeneously enhancing lobulated mass occupying third ventricle.**

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