Intraventricular ganglioglioma

see also Third ventricle ganglioglioma.

Involvement of the ventricular system is rare.

Clinical features

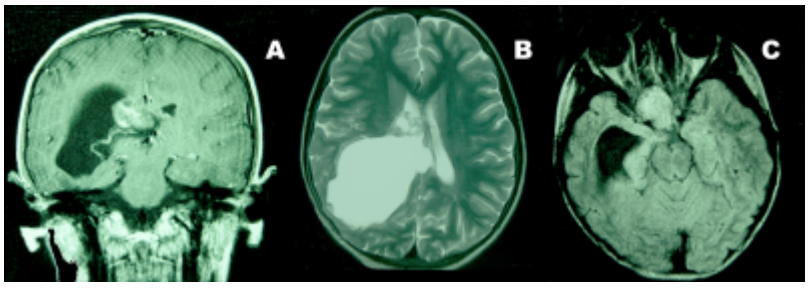
The symptoms are intracranial hypertension or seizure. The degree of hydrocephalus is closely related to the site of tumor's basement [1].

Outcome

The prognosis is good after total resection. The patients with GTR should be followed-up [2].

Diagnosis

The review of 25 previously reported intraventricular gangliogliomas found that their pre-surgical diagnoses were often incorrect, reflecting the difficulty of making the diagnosis with signs, symptoms, and imaging alone [3].



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(A) Coronal T1-weighted image and (B) Axial T2-weighted image showing a solid-cystic intraventricular lesion in the trigone of the right lateral ventricle. The solid component was isointense, and heterogeneous contrast enhancement was observed. Additionally, (C) Axial T1-weighted image showing lobulated isointense lesion in the optic chiasm.

Case series

A total of 7 cases with intraventricular ganglioglioma diagnosed by the surgical pathology examination from June 2004 to April 2011 in our center were retrospectively analyzed. The clinical data were collected from the clinical medical records, and the tumor site, size and basement of tumor were analyzed. Follow up was performed to obtain the clinical outcomes.

The 7 cases included 5 males and 2 females, with disease onset at 23.6 ± 14.9 years old. Epilepsy as the initial symptom was observed in 1 case. Reduced hearing, dizziness and weakness of both lower limbs were found in 1 case. Intracranial hypertension were detected in 5 cases, including 1 case complicating by decreased visual acuity. Tumors were located in the lateral ventricle in 5 cases, while 2 cases in the third ventricle. Hydrocephalus was observed in 5 cases, including 2 cases with severe hydrocephalus, and both underwent ventriculoperitoneal shunting. Total resection of tumors was performed in 5 cases, and 2 cases underwent gross total resection. The mean duration of follow-up was 28.7 months (8-90 months). Intracranial hypertension in all cases disappeared. Even radiotherapy post-surgery, one case with GTR relapsed 1 year later. However, the other 6 cases didn't relapse.

Ganglioglioma in ventricular system is extremely rare, mainly with the symptoms of intracranial hypertension or seizure. The degree of hydrocephalus is closely related to the site of tumor's basement. The prognosis is good after total resection. The patients with GTR should be followed-up [4].

Case reports

Chatrath et al., from the University of Virginia School of Medicine, Bethesda, MD Anderson Cancer Center Houston, report a case of an intraventricular ganglioglioma involving the septum pellucidum in a pediatric patient with history of optic glioma. Only one other pediatric intraventricular ganglioglioma arising from the septum pellucidum has been reported previously.

The patient initially presented at 9 months of age with a pilocytic astrocytoma centered on the optic chiasm, treated with chemotherapy and radiation at 3 years of age. Routine follow-up imaging at 13 years of age revealed the development of a mass in the septum pellucidum, which was subtotally resected endoscopically because of its proximity to the fornices. Pathology confirmed a ganglioglioma positive for the BRAF V600E mutation. The tumor residual progressed and was treated with stereotactic radiosurgery. The patient was asymptomatic at her 6-month follow-up visit and the size of the nodule remained stable.

The review of the 25 previously reported intraventricular gangliogliomas found that their pre-surgical diagnoses were often incorrect, reflecting the difficulty of making the diagnosis with signs, symptoms, and imaging alone. Patients can be reassured that the prognosis is generally favorable following uncomplicated neurosurgical resection [5].

Samdani et al., from the Shriners Hospital for Children, described an illustrative case of an intraventricular ganglioglioma with a prominent cystic component and enhancing mural nodule, which represents the classic radiographic appearance of gangliogliomas described in other locations. A superior parietal lobule approach offered excellent surgical access for tumor removal and the patient has remained free of neurological deficits following surgery. Regardless of location within the central nervous system, ganglioglioma should be on the differential diagnosis for any cystic mass with a mural nodule, particularly in the setting of epilepsy [6].

A patient with a ganglioglioma is presented in the previously unreported location of the anterior third ventricle at the foramen of Monro, mimicking a colloid cyst. We review all other reported cases of intraventricular ganglioglioma (n=6) to characterize this entity. Intraventricular gangliogliomas

typically affect younger patients with female predominance (male:female, 2:5; median age 25 years). Symptoms occur secondary to obstruction of physiological cerebrospinal fluid circulation. Complete surgical resection with re-establishment of cerebrospinal fluid drainage is the goal of treatment [7].

References

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