Rare Pediatric Central Neurocytomas

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KEYWORDS

• Central neurocytoma • Children • Treatment

KEY POINTS

- Central neurocytoma is extremely rare in children.
- Surgical resection is the mainstay of treatment.
- The indication for adjuvant radiation therapy/radiosurgery should be studied further.

Neurocytoma is an uncommon brain tumor that occurs anywhere in the central nervous system. Classically, neurocytomas develop in the lateral ventricles (central neurocytoma), but neurocytoma located outside the lateral ventricles are being increasingly reported (extraventricular neurocytoma [EVN]).[1] Neurocytoma is considered a disease of young adults, because about two thirds of patients are between the ages of 20 and 40 years.[2] It is extremely rare in the pediatric age group. There is no exact data on the incidence of neurocytoma in children. Rades and colleagues[3] reported a clinical series of neurocytoma of children in which data from 59 patients were collected from the literature. This article remains as the largest case series for this rare neoplasm in children under 18 years. According to the study, the median age of affected children was 16 years (range, 1–18).[3] Only 13 patients (22%) were less than 10 years at diagnosis. In the same study, more than one half of the patients had a tumor located mainly in the lateral ventricles and/or third ventricle. Parenchymal (extraventricular) tumor locations in brain or spinal cord were diagnosed in 18 patients (31%).

In 2007, the 4th edition of the World Health Organization classification of tumors of the central nervous system officially adopted EVN as a new entity.[4] EVNs have more diverse histologic features than central neurocytomas. Nishio and colleagues[5] detailed the pathologic features of 'nerve cell tumors' of the cerebrum. Nerve cell tumors are predominantly composed of ganglion cells (gangliocytoma), small neurocytes (neurocytoma), or multiple nerve cell lines (ganglioneuroblastoma, ganglioneurocytoma). In this scheme, cortical tumors consisting of small neurocytes could be called EVNs.[5] However, the diagnostic criteria of EVN need to be further refined, especially among children, because many kinds of dysplastic lesions develop in this age group.[6]

In the authors' institute (a tertiary referral children's hospital), only 2 patients were diagnosed with central neurocytoma from 2001 to 2012. During the same period, 871 patients were diagnosed with brain tumors after operative intervention. Therefore, central neurocytoma represents about 0.2% of pediatric brain tumors. The ages of patients with central neurocytoma were 10 years and 14 years at diagnosis. The 10-year-old boy presented with a generalized tonic clonic seizure. MRI shows a gadolinium-enhancing mass in the left trigone. The mass was well-demarcated and there was no accompanying hydrocephalus (**Fig. 1**). Initially, the tumor was partially removed via a transcortical approach at other hospital.

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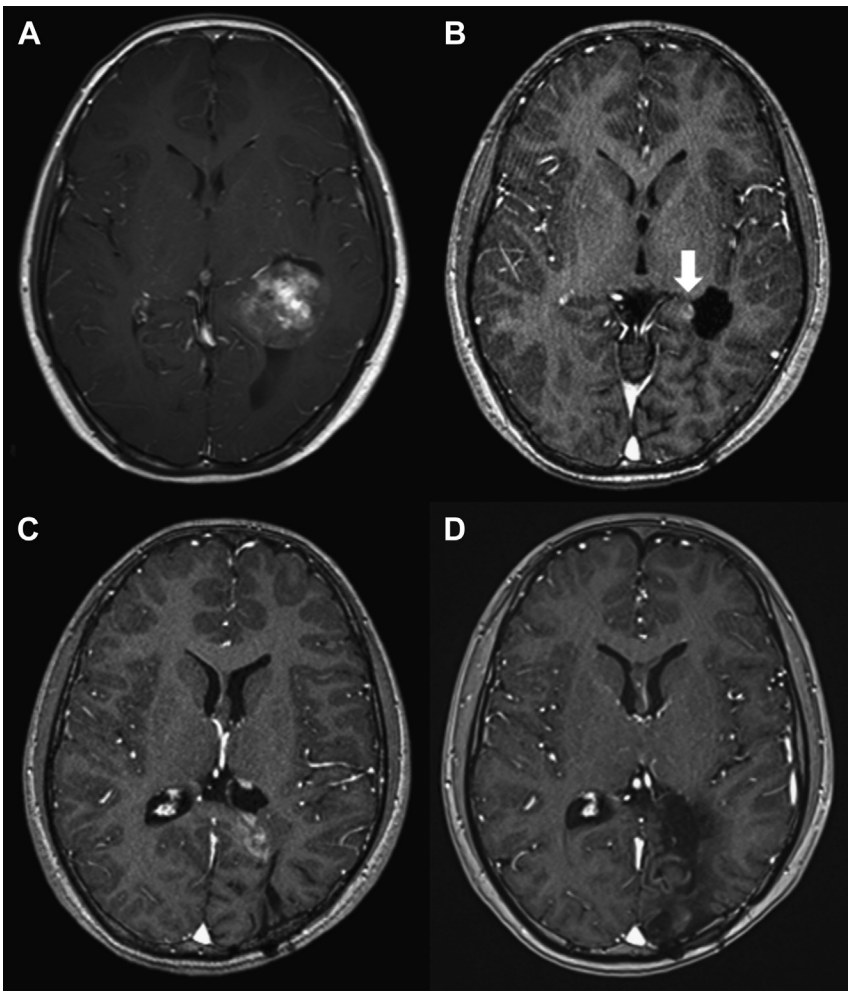

Fig. 1. MR images from a 10-year-old boy. (*A*) Initial preoperative MRI shows a well-demarcated mass in the left trigone. (*B*) The mass was completed removed in the second operation. A small enhancing nodule (*arrow*) developed after 17 months and the patient underwent radiosurgery for the lesion. (*C*) The lesion increased in size and extent 1 year after the radiosurgery. He received a third operation and proton beam therapy. (*D*) Two years after a third operation, there is no recurrence of tumor.

The size of the residual tumor increased for 2 months postoperatively. A second operation was performed at our institute and the residual tumor was completely removed. However, a small enhancing nodule emerged in the tumor bed 17 months later. Although gamma knife radiosurgery was performed for the recurred tumor, the lesion continued to increase in size. A third operation was performed at 37 months after the second operation. After total resection of the tumor, proton beam therapy was given to the tumor bed. No further recurrence was observed for 2 years after the last operation.

The second patient, a 14-year-old girl, developed headache and vomiting; MRI showed a giant mass in the right trigone extending into the parietal lobe. The mass was mixed solid and cystic, and gadolinium enhancement was observed in the solid portion and cyst walls (**Fig. 2**). The tumor was completely removed via a transcortical route. The patient was followed for 9 years and there was no tumor recurrence.

Surgery is the mainstay of treatment for central neurocytoma. After complete tumor resection, the patient can be followed without adjuvant radiation therapy or radiosurgery. In the review by Rades and colleagues,[3] there was no difference in local control and survival between a complete resection group and a complete resection plus radiation therapy group. Incomplete tumor resection frequently leads to tumor recurrence and radiation therapy or radiosurgery may enhance local