Quadrigeminal Plate Hamartoma

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A hamartoma is a benign tumorlike nodule composed of an overgrowth of mature cells and tissues that normally occur in the affected part; often one element predominates. In the case presented, the hamartoma is an overgrowth of neuronal tissue in the quadrigeminal plate, a hamartoma location that has only been reported once previously in the literature [1].

Case Report

At a routine eye examination of an 11-year-old girl, papilledema was found and diagnosed clinically as pseudotumor cerebri. Three years later, the patient was admitted to our institution because of recurring, mostly temporal headaches. There was no focal neurologic deficit or gaze abnormality (specifically Parinaud phenomenon).

The initial CT and the MR images are shown in Figures 1A, 1B, and 1C. Bilateral carotid and right vertebral angiograms disclosed no abnormality. A ventricular shunting procedure (Fig. 1C) and biopsy of the lesion (Fig. 1D) were performed. Since surgery, the patient's headaches have improved considerably.

Discussion

CNS neuroepithelial hamartomas are rare lesions that result from faulty embryologic development. They produce variable neurologic deficit depending on their location. Hamartomas, though rare in the CNS, are encountered relatively more frequently in the hypothalamus and in the temporal lobe [2, 3]. Most hypothalamic hamartomas cause isosexual precocious puberty of central origin. Seizures, intellectual impairment, and behavioral problems occur less frequently. The temporal lobe hamartomas frequently cause epilepsy. To our knowledge, a hamartoma of the quadrigeminal plate has been described only once before in the literature [1]. In that case, the patient was a 35-day-old boy with hydrocephalus diagnosed by CT. After death due to unrelated circumstances, an autopsy revealed a hamartoma of the quadrigeminal plate.

In our case, no distinctive CT and MR features indicated a histologic diagnosis of hamartoma. All we could suspect from CT and confidently conclude from MR was the presence of an intraaxial (parenchymal) lesion of the tectum of the quadrigeminal plate. MR excluded from consideration the entire genre of tumors of pineal gland origin. We had diagnosed the lesion as a type of glioma, either astrocytoma or ependymoma. The MR signal characteristics of the lesion were such that lipoma or a type of cystic lesion warranted no consideration. One clue to the exact histology of the lesion might have been the Chiari I malformation, another developmental anomaly. In Chiari II malformation, there is hyperplasia of many brain structures, including the massa intermedia, the Meynert commissure, and the mesencephalic tectum (mesencephalic beaking) [1, 4]. Mesencephalic beaking is encountered in 89% of patients with Chiari II malformations [4]. One possibility is that the quadrigeminal plate hamartoma seen concurrently with a Chiari I malformation, as in our case, might be a variant of mesencephalic beaking seen in Chiari II malformations.

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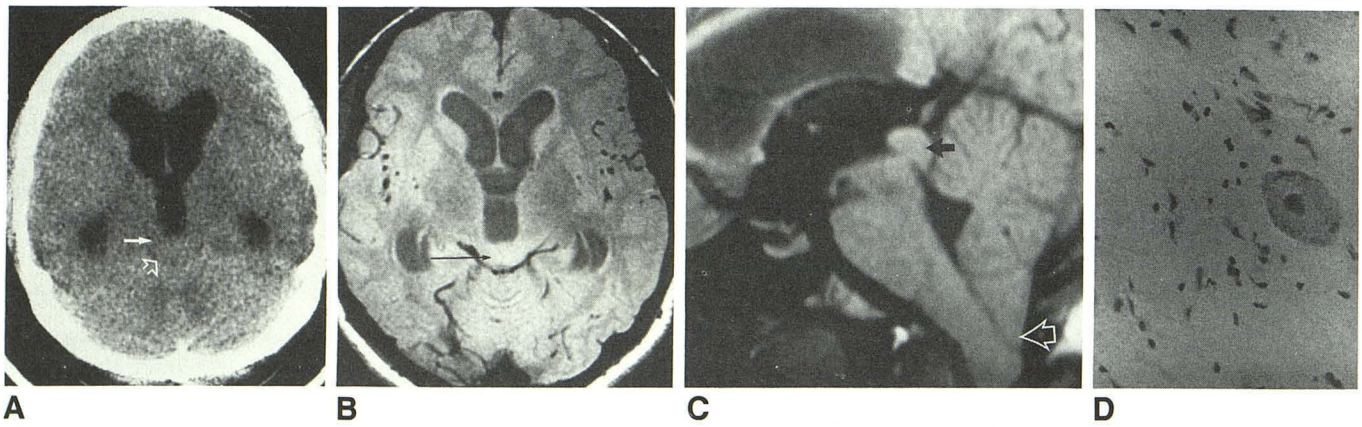

Fig. 1.—*A*, Plain CT scan performed before ventricular shunting shows lesion at quadrigeminal plate (*closed arrow*) compressing quadrigeminal plate cistern (*open arrow*) and causing hydrocephalus.

***B*, Axial MR (2000/20) performed before ventricular shunting shows quadrigeminal plate lesion (*arrow*) much better than does CT (*A*).**

***C*, MR sagittal view (600/25) shows quadrigeminal plate lesion clearly (*closed arrow*). Lesion is causing aquiductal obstruction, which caused hydrocephalus. Incidentally noted is a Chiari I malformation (*open arrow*).**

***D*, Histologic section of the specimen shows a large neuron with numerous astrocytes and reveals reactive astrocytosis with a few disorganized neurons and reactive microglial cells.**