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MR of Fourth-Ventricular Epidermoid Tumors

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Epidermoid tumors arising from the fourth ventricle are uncommon. Although they may grow to an enormous size in a very critical location, the majority of patients do not have the explosive symptoms and neurologic findings common to most fourth-ventricular tumors. We present one confirmed case and one highly suggestive case of epidermoid tumor of the fourth ventricle. Both were evaluated by MR imaging and demonstrate the typical features of these tumors.

Case Reports

Case 1

A 28-year-old woman presented to our institution with progressive ataxia and a right seventh nerve palsy. A CT scan revealed a cystic hypodense lesion in the posterior fossa that did not enhance with the administration of contrast material (Fig. 1A). A diagnosis of cysticercosis was initially suggested. The nature of the lesion was unclear, however, and an MR scan was obtained at our medical center.

The MR study revealed a fourth-ventricular lesion with inhomogeneous low signal intensity on the T1-weighted images (Figs. 1B and 1C) and extremely bright signal intensity on the T2-weighted images (Fig. 1D). The fourth ventricle was stretched and completely filled by this large lesion, which was somewhat molded to the shape of the ventricle. The superior margin of the lesion reached and expanded the posterior portion of the aqueduct. The brainstem, including the pons, medulla, and upper cervical spinal cord, were pushed anteriorly and demonstrated an irregular posterior border. Despite the large size of the lesion, no hydrocephalus or transependymal edema was noted. Since there was no thin-walled cystic structure identified and the signal intensity was not similar to CSF, a diagnosis of cysticercosis was not considered. Instead, a cholesterol-containing tumor in the fourth ventricle such as an epidermoid tumor was suspected preoperatively, especially with the flowing pattern of growth and the lack of hydrocephalus.

At surgery, a fourth-ventricular pearly tumor was readily apparent. The tumor mass was soft and peeled off adjacent neural structures without difficulty. Histopathology demonstrated a typical epidermoid tumor. Postoperatively, the patient's symptoms resolved.

Case 2

A 35-year-old woman presented with a history of headaches and blurred vision of approximately 12 years duration. Nine years previously a head CT scan at another hospital demonstrated hydrocephalus and a possible posterior fossa low-density tumor. At that hospital, no enhancement was seen with contrast material, and a shunt catheter placed into the lateral ventricles for decompression completely resolved the patient's symptoms. The patient refused to have surgery to remove the "tumor." She has been doing well and is followed by annual CT scans.

At the most recent evaluation, a follow-up CT scan with and without contrast enhancement demonstrated essentially no change in the appearance of the low-density posterior fossa tumor (not shown). A higher section showed a low-attenuating mass in the posterior third ventricle and aqueduct (Fig. 2A). The first MR scan obtained 3 years ago from our medical center demonstrated an inhomogeneous mass occupying the upper part of the fourth ventricle, the aqueduct, and the posterior part of the third ventricle. This lesion showed low signal intensity on the T1-weighted image (Fig. 2B) and a very bright signal intensity on the T2-weighted image (Fig. 2C).

Follow-up MR scans over the past 3 years have demonstrated no change in the pattern of tumor involvement. A presumptive diagnosis of a fourth-ventricular epidermoid tumor was made on the basis of the MR findings. Because of the patient's symptoms and the lack of change in the tumor mass over the past years, surgical intervention is being withheld for the present time.

Discussion

Intracranial epidermoid tumors (cholesteatomas) constitute 0.3–1.8% of brain tumors [1]. The great majority of epidermoid tumors arise from the skull base, particularly the cerebellopontine angle and parapituitary regions in the midline [2]. Fourth-ventricular epidermoid tumors are unusual and have only been described in a few reports [3–6].

Although epidermoid tumors are considered to be congenital, they are most frequently diagnosed in patients 25–40 years old (mean, 36 years) [7]. Embryologically, these tumors probably result from incomplete cleavage of neural ectoderm from cutaneous ectoderm with inclusion epiblasts in the neural groove at the time of closure (3–5 weeks gestation) [8].

Epidermoid tumors grow by the desquamation of epithelial cells, which break down into keratin and cholesterol within the tumor capsule. The soft, pliable substance produced by this process and the slow accumulation of the tumor mass allow it to extend into and conform to the shape of the available ventricular and subarachnoid space. Deformity and mass effect on the neural structures thus occur as a late

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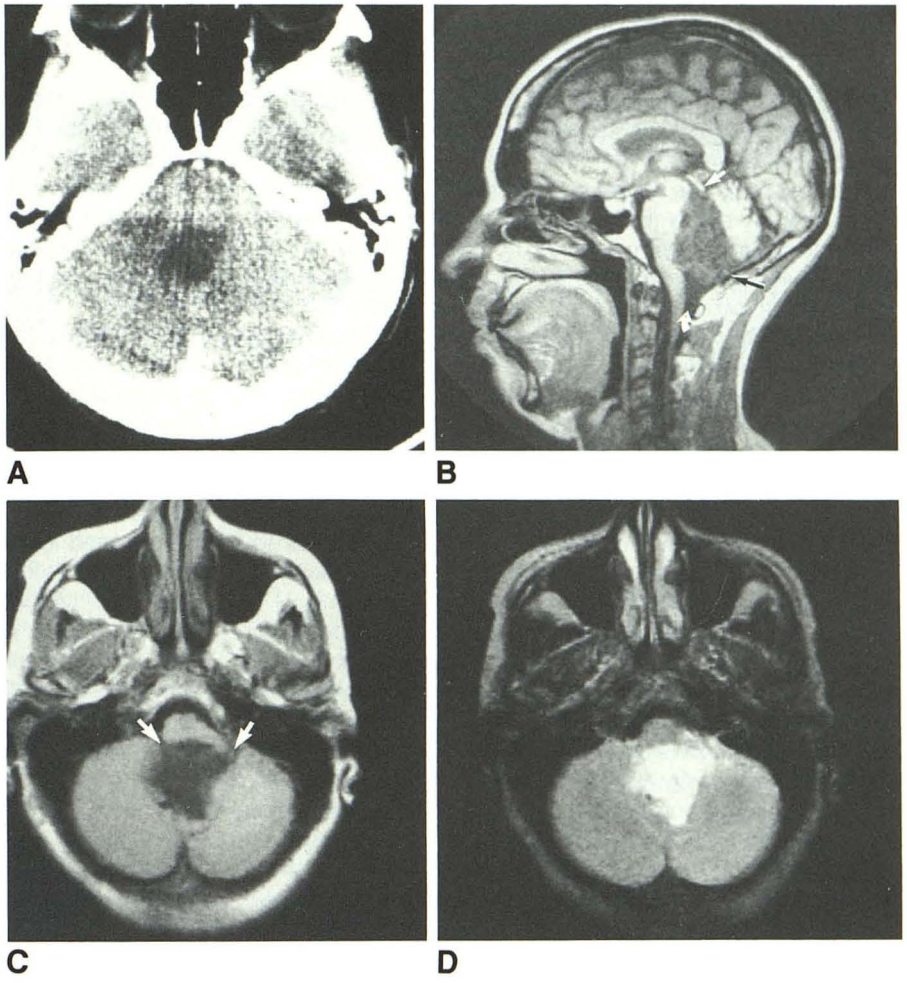

Fig. 1.—Case 1.

A, Axial CT scan. The fourth ventricle is completely filled by a large mass of low attenuation (values not available). Axial CT sections at a higher level demonstrate normal-sized lateral ventricles (not shown).

B, Sagittal T1-weighted MR image (SE 500/30). The fourth ventricle is stretched and completely filled by a large, inhomogeneous low-signal-intensity tumor. Superior margin of lesion reaches and expands posterior portion of aqueduct. The tectum is lifted superiorly and posteriorly (*straight white arrow*). Inferior portion of tumor widely stretches the foramen of Magendie (*black and white arrow*) with posterior and superior displacement of inferior vermis. Part of lesion extends into cisternal space through foramen of Magendie and inferiorly through foramen magnum (*curved arrow*).

C, Axial relatively T1-weighted MR image (SE 550/26). This inhomogeneous low-signal-intensity lesion stretches the fourth ventricle and foramen of Luschka (*arrows*).

D, Axial T2-weighted MR image (SE 2000/80) at same level as *C*. At this level the lesion shows extremely high signal intensity as compared with CSF without any transependymal edema.

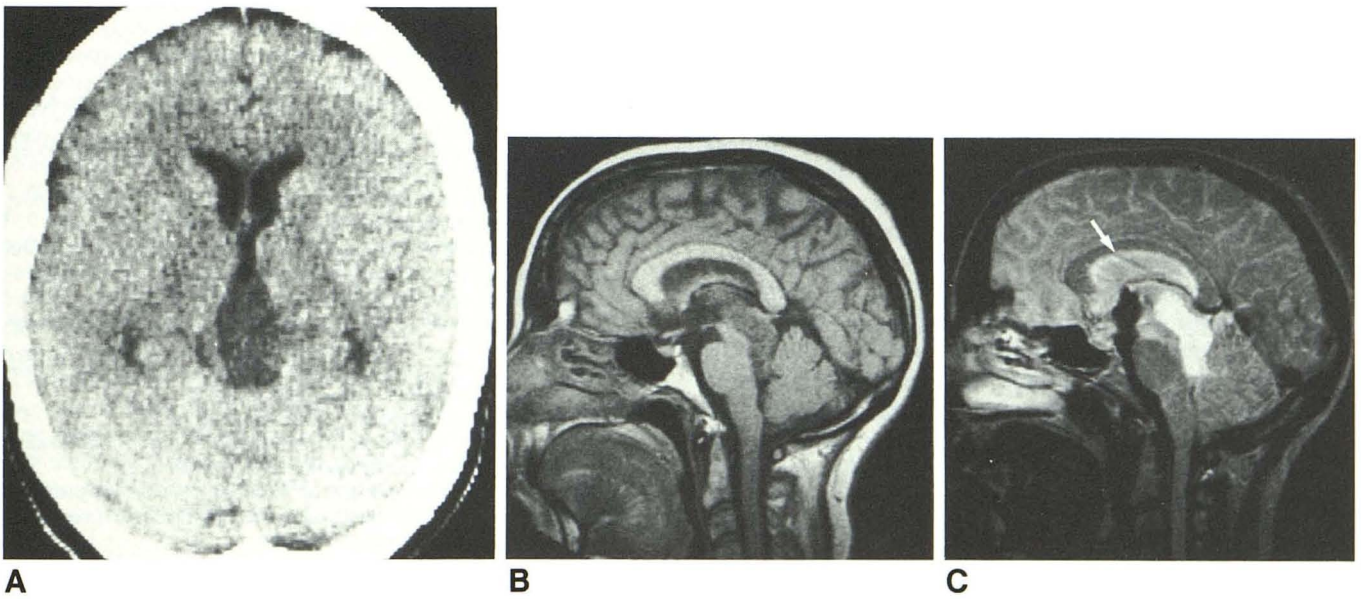

Fig. 2.—Case 2.

A, Axial CT scan. A low-attenuating mass is seen extending into posterior third ventricle.

B, Sagittal T1-weighted MR image (SE 550/26). An inhomogeneous low-signal-intensity lesion is occupying upper part of fourth ventricle, aqueduct, and posterior part of third ventricle. The lesion is stretching upper part of fourth ventricle and aqueduct, which is totally packed and distended. Superior vermis is displaced posteriorly and inferiorly, and the tectum is lifted superiorly. The lesion appears to arise from upper part of fourth ventricle and grows into the aqueduct as well as extends into posterior part of third ventricle.

C, Sagittal T2-weighted MR image (SE 2000/80) at same level as *A*. The lesion shows inhomogeneous, extremely high signal intensity as compared with CSF. Note shunting tube in lateral ventricle (*arrow*).

event and symptoms become apparent only when the tumor has enlarged sufficiently to cause hydrocephalus or local neural compression.

The diagnosis of fourth-ventricular epidermoid tumors based only on clinical findings is difficult because the symptoms and signs are intermittent and nonspecific. Patients may present with symptoms of either classic midline posterior fossa tumors; cerebellopontine angle tumors associated with lateral extension; or progressive dementia, ataxia, and urinary incontinence resulting from communicating hydrocephalus. Other symptoms and signs may include headache with or without vomiting, mental status changes, psychiatric problems, cranial nerve palsy, and hemiparalysis [2–5]. The intermittent nature of the neurologic symptoms produced by epidermoid tumors has even led to reports of long-term spontaneous remission. The remitting symptoms may also lead to the erroneous diagnosis of demyelinating disease, such as multiple sclerosis [5, 9].

Epidermoid tumors usually contain various amounts of cholesterol. CT scans frequently have negative attenuation numbers and are not enhanced with contrast. Epidermoid tumors typically have prolonged T1 and T2 relaxation times. Although these tumors invariably contain cholesterol, they do not produce bright signals on T1-weighted images in contrast to some craniopharyngiomas. Intensity (or T1 relaxation time) differences are thought to relate to differences in the physical state of the cholesterol [10]. Epidermoids contain solid cholesterol, whereas craniopharyngiomas contain liquid cholesterol. Absence of hydrolyzed cholesterol in epidermoids may contribute to a prolonged T1 [10]. The differential diagnosis of low-density, unenhanced fourth-ventricular tumors on CT includes epidermoid cyst, dermoid cyst, arachnoid cyst, cysticercosis cyst, and rarely cystic ependymoma, cystic astrocytoma, and cystic hemangioblastoma.

Dermoid cysts are usually calcified and are frequently associated with a dermal sinus or other anomalies of the vertebral axis. Arachnoid cysts and cysticercosis cysts usually have well-defined, rounded thin margins and are more frequently associated with hydrocephalus than are epidermoid tumors. Both cysts usually have a homogeneous signal intensity similar to CSF on T1- and T2-weighted images as opposed to the inhomogeneous low signal intensity on the T1-weighted image and extremely high signal intensity on the T2-weighted image found in the epidermoid cyst. Most posterior fossa arachnoid cysts occur in the midline between the two cerebellar hemispheres or at the cisterna magna, and frequently cause compression of the fourth ventricle with symptoms early in childhood [11]. Arachnoid cysts of the fourth ventricle are rare but have been reported by Di Rocco et al. [12], who demonstrated compression and narrowing of the fourth ventricle by the arachnoid cyst in one of their patients. Cystic astrocytoma, cystic ependymoma, and cystic hemangioblastoma should demonstrate a greater degree of obstruction and elicit more profound edema and mass effect than seen with epidermoid tumors. The clinical history would be progressive with rapid neurologic impairment.

At surgery, total excision of the epidermoid tumor is frequently unsuccessful because of the close adherence of the tumor to adjacent structures and the widespread nature of the tumor. Residual epithelium may lead to recurrence of tumor, with the slow growth rate similar to normal epidermis [8]. It is essential to make an early diagnosis of this tumor in order to achieve complete resection and reduce irreversible damage to the CNS.

In conclusion, the characteristic MR findings of these tumors include: (1) inhomogeneous low signal intensity on T1-weighted images and marked increase in signal intensity on T2-weighted images, (2) minimal mass effect, (3) a flowing pattern of growth, and (4) frequently absent or minimal hydrocephalus. Although none of the above findings is pathognomonic for epidermoid tumor, they can facilitate the early diagnosis, determine the extent of involvement, and lead to the institution of prompt resection or shunting to revert CNS damage and decrease surgical morbidity.

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