RadioGraphics

Best Cases from the AFIP

Fourth Ventricle Epidermoid Tumor: Radiologic, Intraoperative, and Pathologic Findings[1]

Editor's Note.—Everyone who has taken the course in radiologic pathology at the Armed Forces Institute of Pathology (AFIP) remembers bringing beautifully illustrated cases for accession to the Institute. In recent years, the staff of the Department of Radiologic Pathology has judged the "best cases" by organ system, and recognition is given to the winners on the last day of the class. With each issue of *RadioGraphics*, one or more of these cases are published, written by the winning resident. Radiologic-pathologic correlation is emphasized, and the causes of the imaging signs of various diseases are illustrated.

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History

A 28-year-old woman with no known medical history presented with a history of progressive gait imbalance and left upper limb incoordination over a period of 2 years to the University Health Network in Toronto. The remainder of the patient's history was noncontributory. At physical examination, gait ataxia, nystagmus, and left cerebellar signs including left arm dysmetria and dysdiadochokinesia were observed.

Imaging Findings

Magnetic resonance (MR) imaging of the brain was performed with a superconducting 1.5-T MR system (Signa Echospeed, version 8.2.3–11.2 software; GE Medical Systems, Milwaukee, Wis) with a standard head coil. The examination demonstrated a large, insinuating extra-axial mass centered within and expanding the fourth ventricle (Fig 1). The anteroposterior, transverse, and craniocaudal dimensions of the mass measured 3.9 × 4.5 × 4.5 cm and extended through the foramina of Luschka and Magendie with caudal extension as far as the foramen magnum. Although the midline cerebellar structures were compressed, no associated edema was seen. There was no obstructive hydrocephalus.

Abbreviations: CSF = cerebrospinal fluid, DW = diffusion weighted, FLAIR = fluid attenuated inversion recovery

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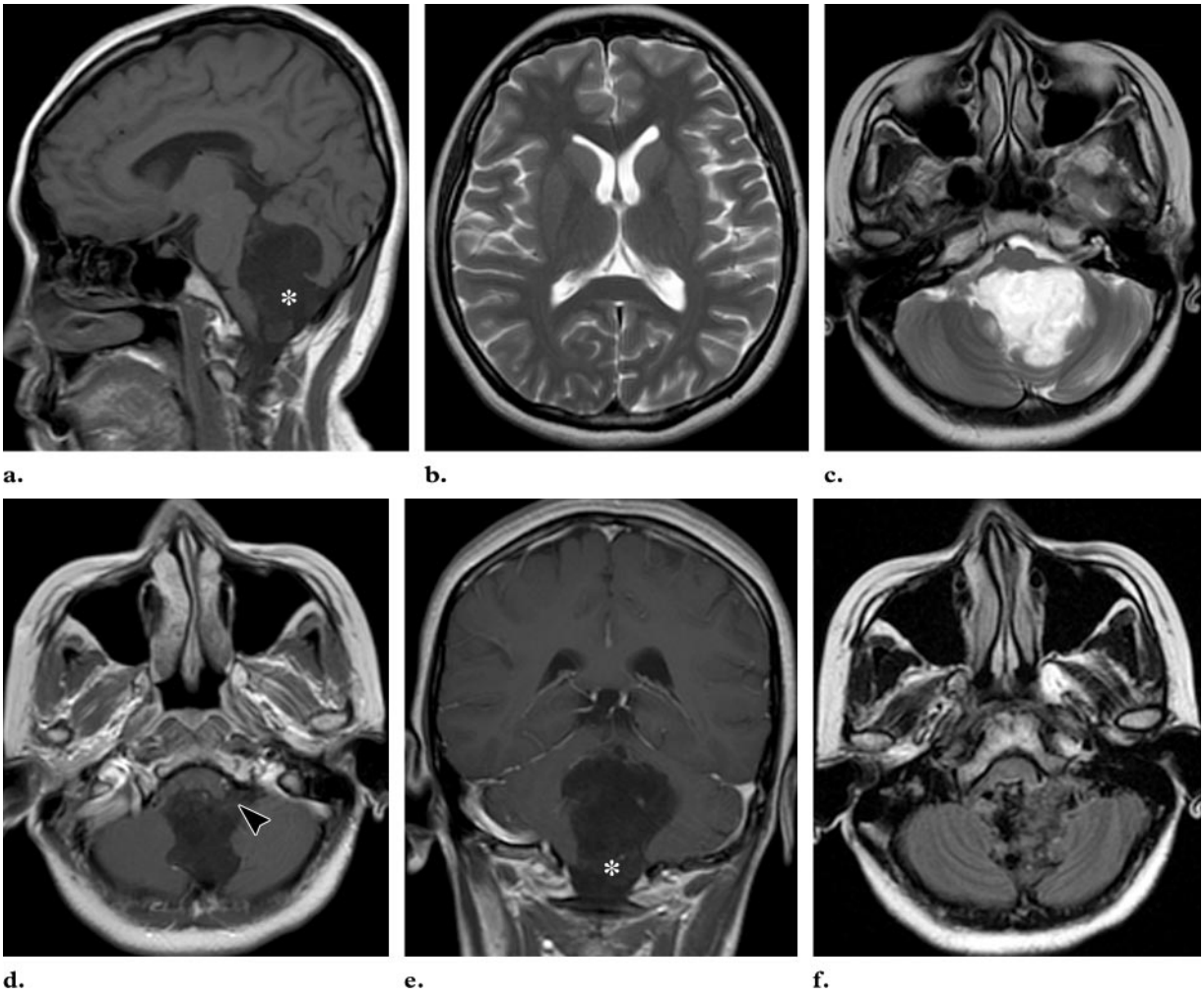
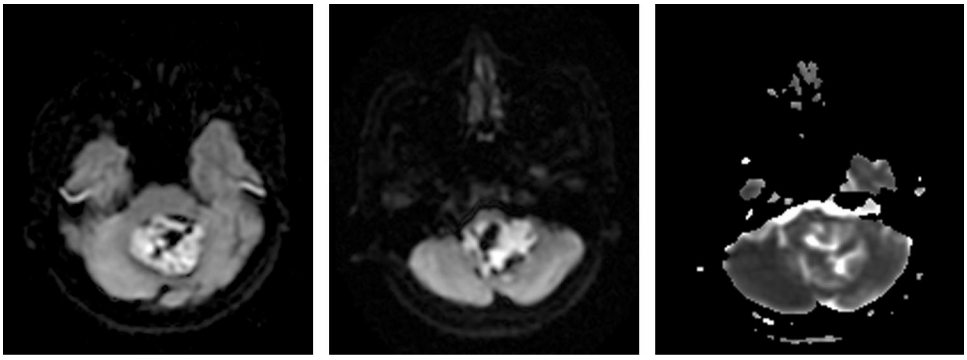


Figure 1. Epidermoid tumor depicted on sagittal T1-weighted **(a)**, axial T2-weighted **(b, c)**, axial **(d)** and coronal **(e)** gadolinium-enhanced T1-weighted, and axial fluid-attenuated inversion recovery (FLAIR) **(f)** images. A fourth ventricle mass is seen expanding, distorting, and compressing the brainstem without any evidence of obstructive hydrocephalus, as demonstrated by the normal size of the lateral ventricles superiorly **(b)**. The mass insinuates through the foramen of Magendie (∗ in **a** and **e**) and the foramina of Luschka (best seen on the left in **c**, **d**, **f**; arrowhead in **d**) and extends into the cisterna magna and caudally to the level of the foramen magnum **(a)**. On T1-weighted images **(a, d, e)**, its signal is only slightly hyperintense relative to cerebrospinal fluid (CSF). On T2-weighted images **(b, c)**, the mass has a hyperintense but heterogeneous signal, unlike the homogeneously high signal intensity of CSF. No significant enhancement is seen, and there is clear failure of suppression of signal on the FLAIR image. This appearance is characteristic of an epidermoid tumor.

The signal intensity characteristics of the mass were slightly hyperintense relative to CSF on T1-weighted images (Fig 1a) and were similarly hyperintense overall relative to CSF on T2-weighted images (Fig 1c). However, unlike the homogeneously hyperintense signal of CSF, the mass had a heterogeneous appearance on T2-weighted images. The heterogeneous signal intensity of the mass and its distinction from CSF were best appreciated on FLAIR images (Fig 1f). With application of the FLAIR pulse sequence, the mass demonstrated significant areas of hyperintensity that failed to suppress, unlike CSF. These



a. b. c.

Figure 2. DW images **(a, b)** and the corresponding apparent diffusion coefficient map **(c)** demonstrate significant areas of restricted diffusion within the fourth ventricular mass (high signal intensity on DW images, low signal intensity on apparent diffusion coefficient map). Pockets of free diffusion (relatively low signal intensity on DW images) are seen within the mass that likely represent CSF channels trapped within the tumor and probably reflect the basis for the absence of obstructive hydrocephalus in patients with these tumors.

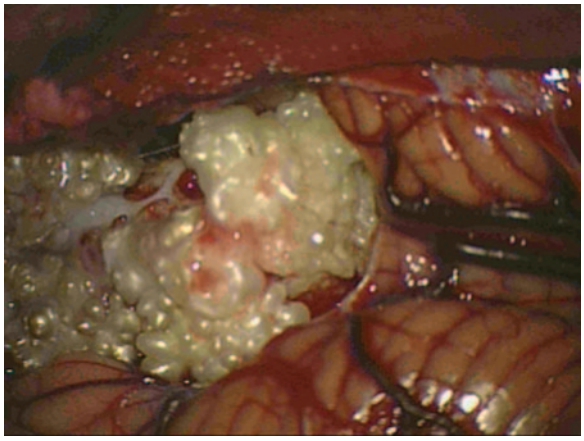


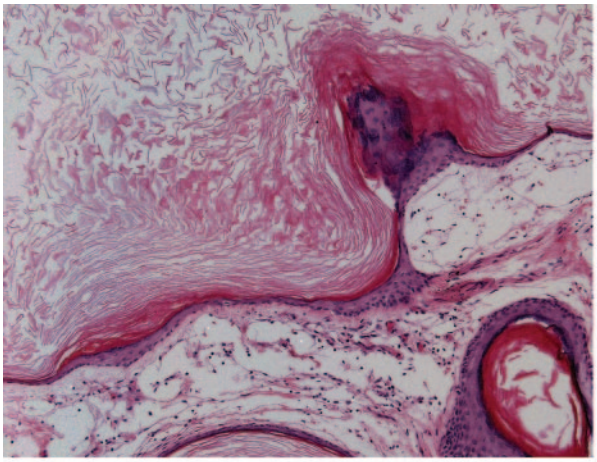
Figure 3. Intraoperative photograph shows an irregularly lobulated, cauliflower-like pearly tumor, an appearance characteristic of an epidermoid.

hyperintense areas were mixed with areas of low signal intensity. On diffusion-weighted (DW) images, overall restricted diffusion with areas of free diffusion that likely represent CSF channels trapped by the tumor (Fig 2) was seen. No significant enhancement of the tumor was seen in images obtained after intravenous administration of gadolinium contrast material (Fig 1d, 1e).

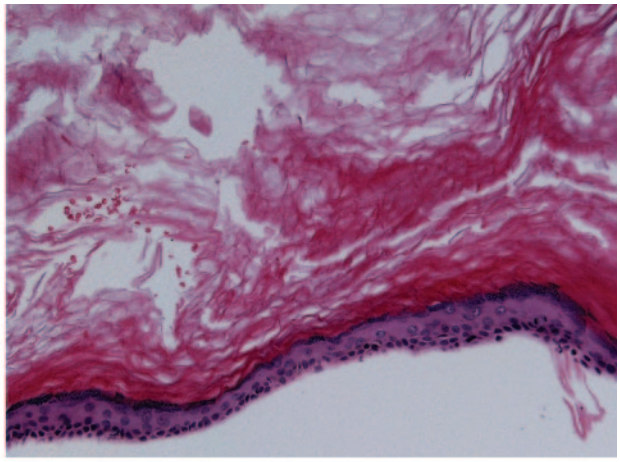
Pathologic Evaluation

A posterior fossa craniectomy was performed to resect the tumor, which was compressing the brainstem. Following the suboccipital craniectomy and upon opening the dura mater, a pearly structure with an appearance characteristic of an epidermoid tumor was seen extending almost to the occipitocervical junction (Fig 3). A subtotal resection (the lining was left intact) of the entirely solid tumor was performed. Good decompression of the brainstem was achieved without jeopardizing vital brainstem structures. The patient had an unremarkable postoperative course and was discharged from the hospital on the 3rd postoperative day.

Macroscopic inspection of the resected specimen revealed a mass measuring 3.8 × 3.5 × 1.4 cm that had a smooth, delicate, translucent capsule and that contained white flaky material. Histopathologic evaluation showed typical features that were consistent with members of the family of pearly tumors (epidermoid tumors), such as attenuated squamous epithelium and abundant anucleate squamae that constituted the bulk of the mass (Fig 4a). The epithelial layer showed regular maturation with no evidence of atypia (Fig 4b). In many regions, cell ghosts (or "shadow cells") resembling vegetable matter were noted (Fig 4c). No sweat glands or sebaceous glands (findings suggestive of a dermoid) were seen.



a.



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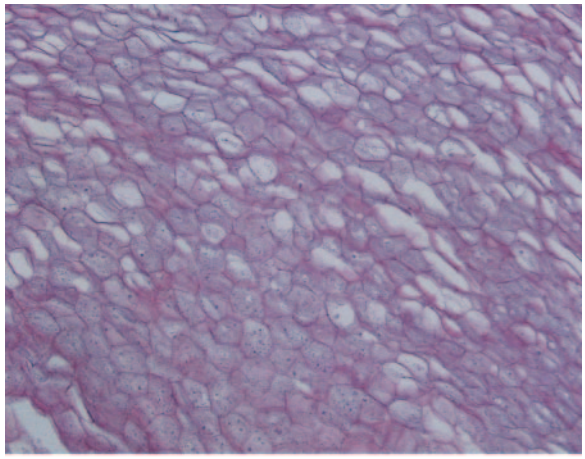

c.

Figure 4. Photomicrographs (hematoxylin-eosin stain) demonstrate an attenuated squamous epithelium with abundant anucleate squamae (original magnification, ×10; **a**), regular maturation of the epithelial layer (original magnification, ×20; **b**), and many regions containing cell ghosts or "shadow cells" (original magnification, ×20; **c**). Note the absence of dermal appendages. These findings are typical features for a member of the family of pearly tumors (epidermoid tumor).

Discussion

Intracranial epidermoid tumors are congenital inclusion cysts that constitute 0.2%–1.8% of primary intracranial neoplasms (1,2). They were first described in 1807 by an artist in a French medical school (3), and the first full description was given by the French pathologist Cruveilhier in 1829 (4). They are believed to form between the 3rd and 5th week of embryonic development as a result of displaced epithelial remnants that remain after the neural tube closes (1,3,5). Although acquired epidermoid tumors may develop as a result of trauma, this is uncommon in the brain. Epidermoids are composed of an outer capsule of connective tissue that surrounds a layer of keratinized stratified squamous epithelium and inner cystic fluid that usually includes debris, keratin, water, and cholesterol. As the epithelial layer desquamates, the cells accumulate and form a cholesterol-rich layer that gives the tumor its characteristic pearly white appearance. These tumors do not contain dermal appendages. They grow—or "insinuate"—within CSF spaces, surrounding and encasing adjacent vessels and nerves. To the surgeon or pathologist, the tumor has an irregular and lobulated cauliflower-like outer surface that glistens like pearls.

Most intracranial epidermoid tumors are intradural tumors. The most common intracranial location for epidermoid tumors is the cerebellopontine angle cistern, which accounts for approximately 40%–50% of cases (1). Epidermoid tumor is the third most common cerebellopontine angle internal auditory canal mass after vestibular schwannoma and meningioma. Although there are only about 100 reported cases of epidermoid tumor in the fourth ventricle, this location is the second most common for an epidermoid tumor in the posterior fossa (6). Another common location is the sellar and parasellar region. Less common locations include intraparenchymal locations (epidermoid tumors have been reported in all lobes of the cerebral hemispheres), the pineal gland, the thalamus, and the septum pellucidum (3). These tumors may also be seen intrinsically within the brainstem (5). In rare cases, they have been reported in other locations such as the lateral ventricles (7). Ten percent of epidermoid tumors are extradural, located in the skull or spine (1).

At imaging, the classic appearance of an epidermoid tumor is that of a CSF-like mass insinuating into the cisterns and encasing adjacent vessels and neural structures (1). On computed tomographic (CT) scans, the typical appearance is that of a nonenhancing and hypoattenuating extra-axial mass. Calcifications may be seen within these tumors in 10%–25% of cases. MR imaging is the best modality for evaluating these tumors. Characteristically, they are either isointense or slightly hyperintense relative to CSF on T1- and T2-weighted images. In many cases, especially for tumors in the cerebellopontine angle cistern or the sellar-parasellar region, the main differential diagnostic consideration is an arachnoid cyst. The distinction is usually made with FLAIR and DW imaging. Arachnoid cysts follow the signal intensity patterns of CSF with all MR pulse sequences, whereas epidermoids are not hypointense on FLAIR images and display areas of hyperintense signal relative to CSF. On DW images, epidermoids typically show restricted diffusion, unlike arachnoid cysts. Finally, although most epidermoids do not enhance, up to 25% may show minimal rim enhancement (1).

Although most epidermoids have a fairly typical appearance and can be diagnosed with the above-mentioned criteria, their signal intensity characteristics may vary depending on the amounts of cholesterol and keratin within the tumor (4). These tumors may rarely appear hyperattenuated on CT scans due to high protein content (these are also referred to as white epidermoids) and show signal reversal with MR imaging pulse sequences, with high signal intensity on T1-weighted images and low signal intensity on T2-weighted images (1). In addition, epidermoids in uncommon locations (eg, intraparenchymal tumors) can pose a significant diagnostic challenge and could be misdiagnosed as astrocytomas preoperatively (3).

The clinical manifestations of epidermoid tumors usually relate to the mass effect of the tumor on adjacent structures. Gait disturbance is a common presenting symptom for patients with posterior fossa tumors (8); other common presenting symptoms include cranial neuropathies and, sometimes, seizures. Hydrocephalus is not commonly seen with intracranial epidermoid tumors, possibly due to fissuring of the cyst wall and the ability of CSF to decompress into the surrounding periventricular structures and outlet foramina (8). There may also be symptoms secondary to chemical meningitis caused by leakage of tumor contents into the subarachnoid space. For technical reasons and to avoid damaging vital structures, a subtotal resection is usually performed to alleviate compression of adjacent structures, with most cases having an overall good prognosis (6). The reported rate of recurrence of epidermoid tumors in literature is highly variable, and once the tumor is removed the brain tissue may re-expand slowly or not at all. Because of this, distinguishing a recurrence from a postoperative liquoral cyst has traditionally been cumbersome, and the decision for repeat resection is based mainly on the patient's symptoms (6). However, the diagnosis of tumor recurrence should now be easier with the application of DW imaging.

Malignant degeneration of an intracranial epidermoid tumor is an extremely rare event and should be considered when progressive neurologic symptoms with tumor enlargement are present and there is (new) contrast enhancement on serial images (9,10). Very rarely, there may even be leptomeningeal tumor dissemination (11). In one reported case of malignant degeneration of a cerebellopontine angle epidermoid tumor into a squamous cell carcinoma, retrospective evaluation of the initial MR images revealed a tiny area of enhancement (10). In their review of the literature, Hamlat et al (9) reported that out of 17 cases of malignant degeneration from the remnant of a benign lesion, six showed enhancement at the time of the initial CT study. Therefore, special note should always be made when any enhancement is seen on images of these tumors. Although benign cystic tumors may show some degree of associated enhancement, it has been suggested that surgery should focus on resection and histologic analysis of the enhancing area when encountered (9).

In summary, epidermoid tumors are benign slow-growing tumors that usually manifest as insinuating masses that can be accurately diagnosed with MR imaging. Their MR signal intensity characteristics are usually similar or slightly hyperintense relative to CSF on T1- and T2-weighted images, but these tumors are not hypointense on FLAIR images and show restricted diffusion on DW images, features that allow them to be distinguished from arachnoid cysts. Although most of these tumors do not enhance, minimal rim enhancement may be seen in up to 25% of cases. When atypical features such as associated enhancement are seen, special note

should be made to direct the surgeon's attention to the atypical area: Even though malignant degeneration is very rare, among such cases a significant proportion of tumors were initially associated with some degree of enhancement. When enhancement is seen, the surgeon may decide to focus on resection and histologic analysis of the enhancing area.

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