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A Choroid Plexus Papilloma Arising from an Incidental Pineal Cyst

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Summary: We report a case of an incidental pineal cyst discovered on MR imaging that progressed to a choroid plexus papilloma. The MR features of pineal cysts, the difficulty of distinguishing benign cysts from cystic neoplasms, and the implications for surgical management are discussed.

Index terms: Choroid plexus, neoplasms; Pineal gland, cysts

Benign pineal cysts are common findings in as many as 40% of autopsy specimens (1, 2). Although these cysts are reported as incidental findings in 1.4% to 4.3% of magnetic resonance (MR) examinations, they are almost always asymptomatic (3, 4). Rare symptomatic cysts are usually caused by mass effect or hemorrhage (5). The fact that MR imaging can distinguish benign pineal cysts from pineal neoplasms is of obvious clinical importance. We present a case report of an apparent pineal cyst that progressed to a choroid plexus papilloma. Our case demonstrates the difficulty in characterizing pineal mass lesions on the basis of MR imaging and, in particular, the difficulty in distinguishing pineal cysts from pineal neoplasms.

Case Report

In 1991, a 24-year-old woman presented with a history of headache, bilateral central scotomas, photophobia, and visual aura. A clinical diagnosis of migraine was made. Computed tomography (CT) performed at that time showed a well-circumscribed, 1-cm-diameter, nonenhancing cyst in the location of the pineal gland. At MR imaging, performed at 1.5 T, the signal intensity of the cyst appeared to be slightly hyperintense relative to cerebrospinal fluid (CSF) on T1-weighted images (Fig 1A), proton density–weighted images (Fig 1B), and T2-weighted images. A T1-weighted image obtained after intravenous administration of gadopentetate dimeglumine revealed a thin, smooth rim of enhancement of the cyst wall (Fig 1C). The MR findings were otherwise normal.

The patient remained asymptomatic until 1994, when she presented with headache, nausea, vomiting, and diplopia. Physical examination revealed Parinaud syndrome, bilateral papilledema, ataxia, and hyperreflexia in the right extremities. A CT scan followed by MR examinations showed a 2.0 × 2.5-cm, well-circumscribed, nonhomogeneously enhancing, mixed cystic and solid pineal mass with secondary obstructive hydrocephalus (Fig 2A–D). Serum α-fetoprotein and human chorionic gonadotropin levels were normal. An open biopsy was performed, and histopathologic examination revealed a choroid plexus papilloma (Fig 2E).

Discussion

Benign pineal cysts may be seen in as many as 40% of autopsy specimens (1, 2). This high rate of occurrence is due to the inclusion of both microscopic and macroscopic cysts in the study series (2). MR studies have established a rate as high as 4.3%, with the majority of these cysts measuring less than 1 cm in diameter (4). Pineal cysts are almost invariably asymptomatic and are usually discovered incidentally during the investigation of unrelated neurologic disorders. Rarely, pineal cysts produce symptoms, and these usually result from mass effect or hemorrhage. Compression of the midbrain tectum may produce Parinaud syndrome. Aqueductal compression may produce hydrocephalus with signs and symptoms of raised intracranial pressure.

The MR features of pineal cysts have been described. Benign, asymptomatic pineal cysts are usually well-circumscribed, homogeneous, round lesions that are only slightly hyperintense relative to CSF on T1-weighted images. On T2-weighted and proton density–weighted images they are hyperintense relative to CSF (4, 6). Kjos et al (7) have attributed these features to

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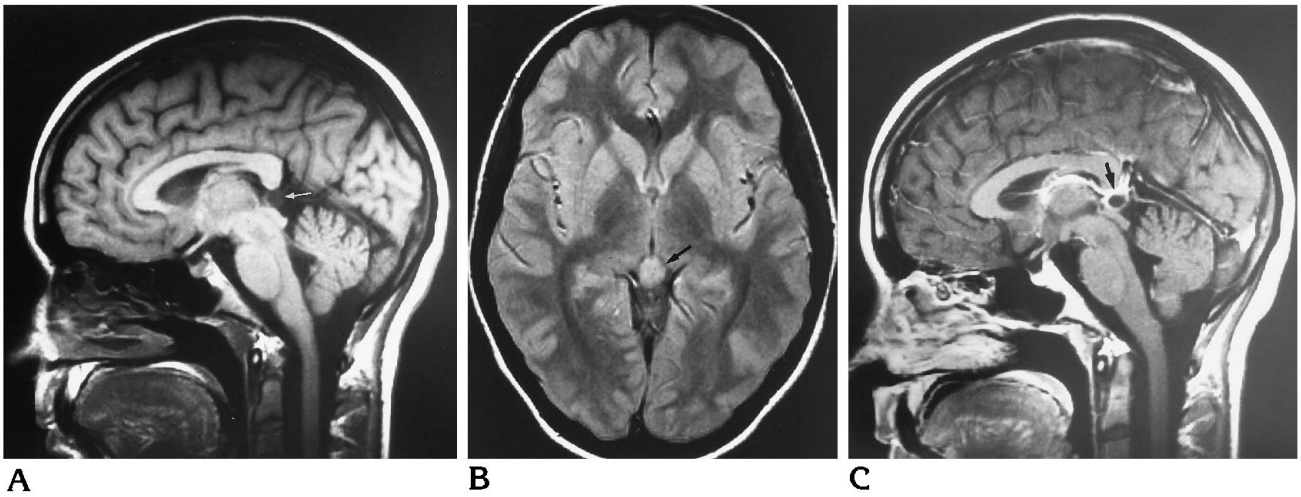

Fig 1. Twenty-four-year-old woman at initial presentation.

A and *B*, Sagittal spin-echo T1-weighted (600/15/2 [repetition time/echo time/excitations]) *(A)* and axial proton density–weighted (2500/22/1) *(B)* MR images show a 1-cm-diameter pineal cyst *(arrow)*, which is slightly hyperintense relative to CSF.

C, Sagittal postcontrast T1-weighted MR image (600/15/2) shows a thin, smooth rim of enhancement of the cyst wall (*arrow*).

proteinaceous fluid content. Pineal cysts normally display a fine rim of enhancement after injection of contrast material. This has been attributed to the lack of a blood-brain barrier in the surrounding pineal tissue (8). Diffusion of contrast material into the cyst may cause more generalized enhancement if imaging is delayed (8). Fleege et al (9) reported that symptomatic pineal cysts may have an atypical MR appearance, including large size (mean, 1.6 cm), hemorrhage (18%), and nodular, irregular enhancement (58%). The MR appearance of symptomatic pineal cysts, particularly the nodular, irregular enhancement, overlaps with the appearance of pineal neoplasms. In the study by Fleege et al (9), an incorrect preoperative diagnosis of pineal neoplasm was made in 14 of 19 symptomatic pineal cysts on the basis of imaging findings and clinical criteria.

Our case illustrates the difficulty in differentiating pineal cysts from pineal region neoplasms on the basis of MR imaging. In 1991, our patient presented with clinical symptoms that were attributable to migraine. An MR examination revealed a 1-cm-diameter pineal lesion (Fig 1–C), characteristic of an incidental benign pineal cyst. Although we have been unable to find previous reports of a purely cystic choroid plexus papilloma, Nakagawa et al (10) reported a case of a purely cystic pineocytoma. Therefore, since a histologic specimen was not obtained at the time, the possibility that the pineal cyst identified actually represented an early purely cystic pineal neoplasm cannot be excluded. In 1994, our patient presented with clinical symptoms, including Parinaud syndrome, that were clearly related to the presence of a pineal mass lesion. An MR examination at that time showed a 2.0 × 2.5-cm mixed cystic and solid mass arising from the region of the pineal gland. On the basis of the clinical and MR findings, a pineocytoma was considered the most likely diagnosis. After a biopsy was performed, a diagnosis of choroid plexus papilloma was established on the basis of light and electron microscopic and immunohistochemical studies.

Choroid plexus papillomas are rare, accounting for only 0.4% of adult intracranial neoplasms (11). In adults, these tumors arise from the fourth ventricle in 70% of cases, the lateral ventricles in 24%, and, uncommonly, from the third ventricle or cerebellopontine angle cistern (11). The tumors of the third ventricle tend to arise posterosuperiorly (11). Animal studies have established an intimate relationship between the choroid plexus and the pineal gland (12). The development of a choroid plexus papilloma within the pineal gland is obviously rare but not totally unexpected given their intimate anatomic relationship. On MR images, choroid plexus papillomas typically appear as solid or, as in our case, mixed cystic and solid tumors.

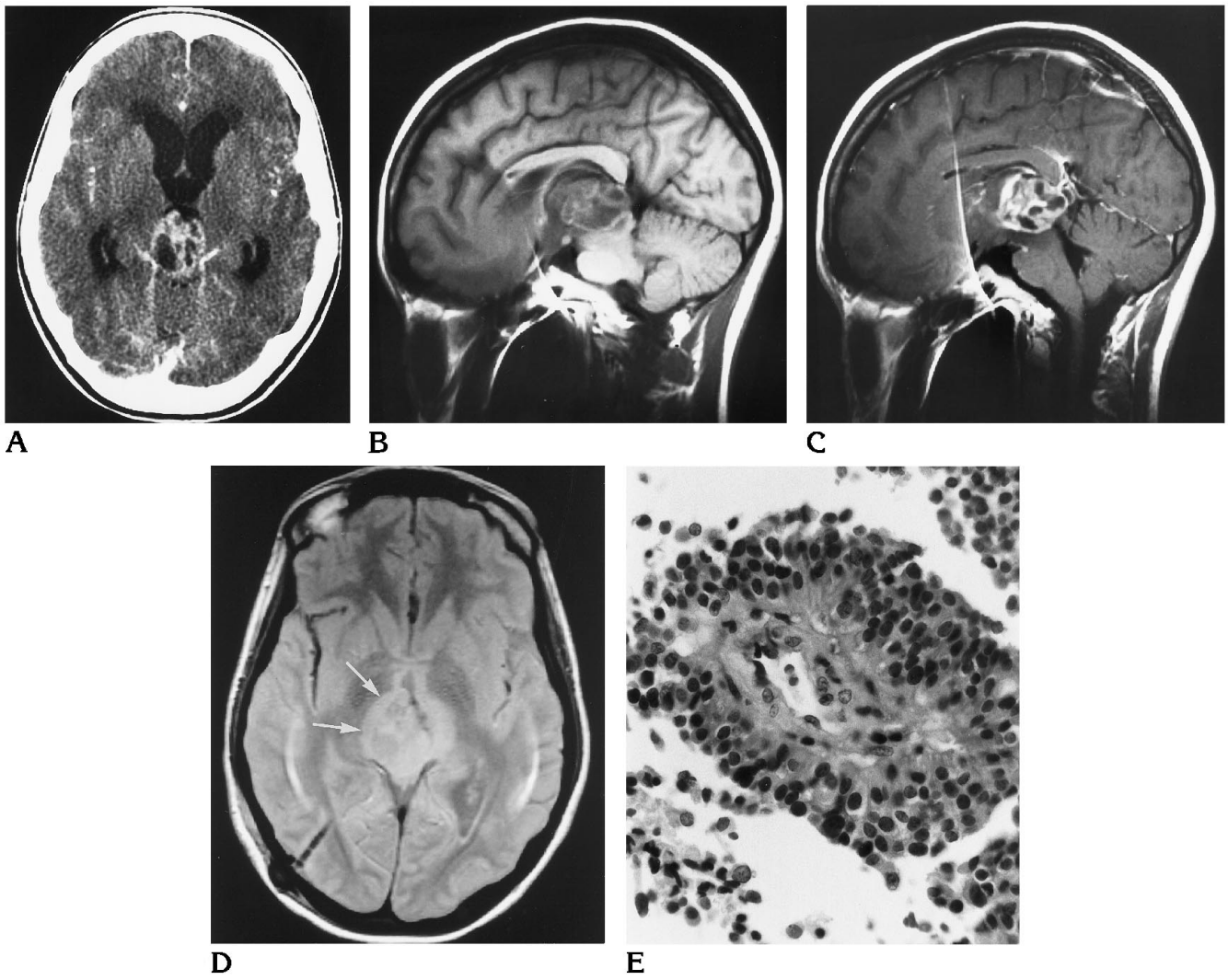

Fig 2. Same patient, 3 years later.

A, Contrast-enhanced axial CT scan shows a 2.5-cm-diameter mixed cystic and solid mass in the pineal region with obstructive hydrocephalus.

B, Noncontrast sagittal T1-weighted MR image (480/14/3) shows a 2.5-cm-diameter cystic and solid mass compressing the midbrain tectum. Note the metal artifact arising from the patient's dental amalgam.

C, Sagittal postcontrast T1-weighted MR image (480/14/3) shows a nonhomogeneously enhancing tumor.

D, Axial proton density–weighted MR image (2500/22/1) shows a large mass in the pineal region (*arrows*).

E, Photomicrograph of the tumor shows a single papilla with a fibrovascular core distinctly separated from the epithelial cells, which were immunohistochemically shown to contain cytokeratins and to react for S-100 protein (hematoxylin-eosin stain, magnification ×640). Results of glial fibrillary acidic protein tests were negative. Electron microscopy of the same specimen revealed surface microvilli and apical tight junctions with no evidence of mucin. These findings were consistent with choroid plexus papilloma.

The solid components of the tumor usually enhance intensely.

Metastatic involvement of the pineal gland has been well described. Pathologic studies in our patient, however, were most consistent with a primary choroid plexus tumor. The patient is currently responding well to radiation therapy, and there is no clinical evidence of a neoplasm elsewhere in her body.

In summary, we have presented an unusual case of a choroid plexus papilloma arising from an incidental pineal cyst. Our case illustrates the difficulty in distinguishing pineal cysts from pineal region neoplasms on the basis of MR imaging. Although it would be impractical to perform follow-up MR examinations on all incidentally discovered asymptomatic pineal cysts, we believe that cysts with atypical imaging fea-

tures should be followed up closely. Any symptomatic pineal lesion, whether cystic or solid, requires definitive diagnosis and treatment. Stereotaxic management of symptomatic benign pineal region cysts has been reported (13). If stereotaxic aspiration is performed, then a purely cystic neoplasm may be overlooked, since a histologic specimen is usually not obtained and the diagnosis depends on cytologic examination of cystic fluid. Therefore, if stereotaxic aspiration is performed, close follow up with MR imaging is recommended.

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