Pineocytoma Mimicking a Pineal Cyst on Imaging: True Diagnostic Dilemma or a Case of Incomplete Imaging?

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AJNR Am J Neuroradiol 2008, 29 (1) 159-163
doi: https://doi.org/10.3174/ajnr.A0750
http://www.ajnr.org/content/29/1/159
Pineocytomas are rare tumors that arise from pineal parenchymal cells. Imaging features are nonspecific; classically, they present as well-circumscribed homogeneously enhancing solid masses, centered on the pineal gland. However, there is considerable variability in this appearance, and certain pineocytomas may have a cystic or partially cystic appearance. In some cases, the appearance may be similar to a pineal cyst, and ocytomas may have a cystic or partially cystic appearance. In solid masses, centered on the pineal gland. However, there is considerable variability in this appearance, and certain pineocytomas may have a predominantly cystic appearance on imaging. This has led some to suggest that differentiation between pineocytomas and pineal cysts may not be possible. We have attempted to determine if cystic pineocytomas could be found in a series of these tumors evaluated by CT and MR imaging.

MATERIALS AND METHODS: We searched the radiology literature as well as the medical records from our own institution for pathologically proved pineocytomas with available preoperative imaging or imaging reports, with specific focus on whether postcontrast MR imaging was included. In cases in which images were available, they were evaluated by a Certificate of Added Qualification (CAQ)-certified neuroradiologist and a radiology resident, who attempted to determine if the pineocytomas had any MR imaging characteristics of typical pineal cysts. To be considered a typical pineal cyst, an area of signal-intensity abnormality must be centered on the pineal recess, demonstrating internal homogeneity on T2-weighted imaging, following CSF signal intensity on T1- and T2-weighted images, without any marginal lobularity or nodular contrast enhancement and a wall thickness of <2 mm. In cases in which imaging was not available, radiology reports and/or descriptions provided in the radiology literature were reviewed by a CAQ-certified neuroradiologist. For any lesion described as cystic, we again attempted to elucidate the exact extent of imaging that was performed, note specific lesion characteristics, and determine if the lesion met the criteria described previously. Finally, for tumors in which image size was provided, the mean value of maximal tumor dimension, SD, median, and range were calculated.

RESULTS: Forty-four pathologically proved cases of pineocytomas from the radiology literature, as well as 8 pathologically proved cases of pineocytomas from our institution with available imaging studies and/or reports, were reviewed. Of these, 23 were solid masses, and 7 were partially solid and cystic, whereas 14 tumors could not be completely characterized due to incomplete imaging evaluation. Eight were primarily cystic; however, none of these could be confidently characterized as meeting the criteria for a typical cyst.

CONCLUSION: In our analysis, no truly cystic pineocytomas were identified.

Materials and Methods

Literature Review

A PubMed search was performed on May 23–24, 2006, and subsequently repeated on April 24–25, 2007, to ensure that no cases were missed, by using the keywords “pineocytoma,” “pineal cyst,” “pineal tumor,” “pineal neoplasm,” “pineocytoma MR,” “pineocytoma imaging,” “pineal cyst imaging,” and “pineal MR.” The literature was
searched for case reports and case series from the past 20 years in English-language articles describing the appearance of pineocytomas on MR imaging.

**Internal Patients**

Our institution has in place an “honest broker” system to comply with the Health Insurance Portability and Accountability Act regulations, and this study was performed within the guidelines of that system. A search was performed of our composite electronic medical records, encompassing 20 academic and community hospitals affiliated with our institution in an effort to identify patients who had pathologically proved pineocytomas with available imaging studies and/or reports. Radiology reports from January 1, 1991, to October 4, 2006, were searched by using the keywords “pinealcytoma,” “pineocytoma,” “pinealblastoma,” “pinealoblastoma,” “pineal parenchymal,” and “pineal tumor.” Pathology reports from January 1, 1991, to October 4, 2006, were searched by using keywords “pineocytoma” and “pineal parenchymal tumor” to attempt to uncover any additional cases. A total of 322 cases were reviewed, excluding duplicated names between the 2 searches. Final cases were selected that met the following 3 criteria: A CT and/or MR imaging was performed before surgery, the images and/or radiology reports were available, and there was an available pathology report confirming the diagnosis of pineocytoma.

For both the cases from the literature and the internal patients, when images were available, they were reviewed by a Certificate of Added Qualification–certified neuroradiologist and a third-year radiology resident, and the imaging features of these lesions were evaluated with respect to the presence of cysts, enhancement pattern, lesion size, and wall characteristics. Note was made of any limitations of the available images. When only reports were available, we attempted to obtain relevant information regarding these features. If available, lesion size was noted or measured, and average maximal tumor diameter with SD, median, and range was calculated. Where possible, we noted as many as possible of the following technical scanning criteria: section thickness, sequences used (for MR imaging), imaging plane, use of contrast, dose, matrix size, FOV, any delay between injection and imaging, scanner make and model, and field strength (for MR imaging).

Finally, we attempted to classify tumors as solid or cystic, keeping in mind our goal of attempting to determine whether a truly cystic pineocytoma existed and if it could possibly mimic a typical pineal cyst on imaging. For the purposes of this study, the criteria of a typical pineal cyst put forth by Barboriak et al were used.7 A typical pineal cyst was considered present if it met 4 MR imaging criteria: a round or ovoid area of signal-intensity abnormality centered on the pineal recess, the area of concern demonstrating hypointensity to white matter on T1-weighted images and isoointensity with CSF on T2-weighted images, the area of concern being internally homogenous on T2-weighted images, and last, no marginal lobularity or nodular contrast enhancement demonstrated. A rim of T2-hypointensity or contrast enhancement on T1-weighted images was permitted if the rim measured <2 mm thick.7

**Results**

**Literature Review**

Twelve case reports or case series that included MR imaging of pineocytomas, some of which also included CT imaging, were found and yielded 44 pathologically proved cases of pineocytomas. Various imaging techniques, including CT with and without contrast and T1-weighted, T2-weighted, and postcontrast T1-weighted MR imaging, were performed. Thirteen of 44 cases were imaged with non-contrast-enhanced CT. Sixteen of 44 cases were imaged with contrast-enhanced CT, and 37 of 44 were imaged with MR imaging. Of those, 34 of 44 were imaged with T2-weighted MR pulse sequences, 36 of 44 were imaged with T1-weighted MR pulse sequences, and 21 of 44 cases were imaged with T1-weighted MR pulse sequences following the administration of gadolinium contrast.

Unfortunately, 5 of the 12 reviewed case reports or case series provided no specific information regarding scanner type used or scanning parameters. The remaining 7 studies provided differing levels of detail regarding scanning parameters. MR imaging was performed on 0.5T, 1.5T, and 2T scanners. T2-weighted images were acquired with a conventional spin-echo technique with TRs ranging from 2000 to 4800 ms and TEs ranging from 80 to 108 ms. Section thickness (provided in only 2 studies) ranged from 3 to 7 mm. For T1-weighted spin-echo sequences, TRs ranged from 400 to 600 ms and TEs ranged from 11 to 30 ms. Section thickness ranged from 3 to 7 mm. Information was provided in only 1 study regarding FOV, 22 × 22 cm for both T1 and T2 sequences, and matrix size, 256 × 256 for T2 sequences and 256 × 296 for T1 sequences. No information was provided regarding the specific type of gadolinium-based contrast used, volume of contrast, or time elapsed between contrast administration and imaging.5,14,18–22

Only a single reviewed study provided information regarding CT scanning parameters, though even then specific information on scanner type was not provided. In that study, 10-mm sections were obtained in the supratentorial region, and 5-mm sections, in the infratentorial region, using 200–275 mAs and 100–120 KV(p), with a 512 × 512 matrix. Information was not provided in any study regarding type or volume of iodinated contrast used or time from contrast administration to imaging.18

Of the tumors evaluated, 16 were solid masses, whereas 6 were partially solid and partially cystic. These tumors were well-circumscribed isodense-attenuated-to-hypodense masses, centered on the pineal region or pericranial CT, with marked enhancement on postcontrast CT. On MR imaging, they were well-circumscribed masses, demonstrating low-to-isointense signal intensity on T1-weighted imaging, and heterogeneous, isointense-to-high signal intensity on T2-weighted imaging. Eleven of the aforementioned 22 solid or partially solid masses were imaged by using postcontrast T1-weighted imaging, all of which demonstrated enhancement of the solid portion of the mass. Unfortunately, 14 tumors could not be precisely classified because they were incompletely evaluated, lacking either T1-weighted, T2-weighted, or postcontrast T1-weighted MR imaging.

Two cases series described primarily cystic pineocytomas. The first described 6 pineocytomas that were primarily cystic.5 One was septate and eccentric with a thick wall, 1 had a dorsal nodule, 3 had walls that were thicker than 2 mm (more precise measurements were not provided), whereas 1 may have been a simple cyst with a thin wall (it is difficult to determine this from the published data). Unfortunately, specific information regarding wall thickness, specifically whether the enhancing
wall was greater or less than 2 mm, was not provided, nor were images provided for independent review. The second described 2 cystic pineocytomas, both of which were symptomatic—headaches and tics in 1 case and headache, vertigo, Parinaud syndrome, and balance disturbances in the second—and both of which demonstrated enhancement of the cyst wall. Unfortunately, again further information regarding wall nodularity or wall thickness, specifically whether the enhancing wall was greater than or less than 2 mm, was not provided; images were not provided for independent review, nor were scan parameters available.

Very few reviewed cases provided detailed measurements of tumor size. Only 8/44 cases provided tumor measurements in 2 or 3 dimensions, whereas 2/44 cases provided only the maximal tumor dimension.\textsuperscript{6,19,22} The remaining cases either classified the tumors as "small," "medium," "large," or ">20 mm," or did not comment on tumor size.\textsuperscript{8,19-21,23-27} Given these limitations, we calculated the average maximal dimension of the 10 tumors with provided measurements to be 22.1 mm with an SD of 13.2 mm. Values ranged from 9 to 50 mm, with a median value of 22.5 mm.

**Internal Patients**

The records of our institution yielded 8 pathologically proved cases of pineocytomas with available MR imaging studies and/or reports. Various imaging techniques, including CT without contrast and T1-weighted, T2-weighted, and post-contrast T1-weighted MR imaging, were performed. Three of 8 cases were imaged with non-contrast-enhanced CT, 4/8 were imaged with T2-weighted MR pulse sequences, 4/8 were imaged with T1-weighted MR pulse sequences, and 8/8 cases were imaged with T1-weighted MR pulse sequences following the administration of gadolinium (though in 2 cases tumor enhancement was noted with T1-weighted MR imaging, and 8/8 cases were imaged with T2-weighted MR imaging, all tumors demonstrated enhancement of the cyst wall).\textsuperscript{7,16,17} Clearly any patient presenting with symptoms referable to a pineal cyst (or other pineal mass) deserves imaging—most of these tumors bear little resemblance to typical pineal cysts.

A typical pineal cyst, as defined by Barboriga et al.,\textsuperscript{7} can have a thin <2-mm rim of enhancement. It is the lack of a blood-brain barrier surrounding the pineal gland that allows the walls of these benign cysts to enhance because the wall is composed of pineal tissue.\textsuperscript{17} It has been theorized that the delayed internal enhancement of pineal cysts is most likely due to passive diffusion of contrast material from surrounding pineal tissue with a possible component of active secretion, as well; however, the exact mechanism has yet to be fully described.\textsuperscript{17} This same mechanism is responsible for the increasingly homogeneous pattern of enhancement of pineal cysts, possibly resembling a solid mass, on significantly (60–90 minutes) delayed postcontrast imaging.\textsuperscript{17} This is a potential pitfall and may result in confusion when interpreting delayed images (which are not routinely or intentionally obtained at most imaging centers). Therefore, it is important to take into account the time elapsed after contrast administration in such unusual cases.

The imaging features of atypical pineal cysts, including irregular nodular enhancement and hemorrhage into a typical pineal cyst, have been previously described by multiple authors.\textsuperscript{7,9,14,15,17} Large pineal cysts may cause mass effect and compression on the quadrigeminal plate or vein of Galen, leading to lethal increases in intracranial pressure and neurologic devastation.\textsuperscript{17} Some authors advocate imaging follow-up for pineal cysts >14 mm for this reason, whereas other authors report that pineal cysts are unlikely to change in size with time and may even involute, and they advocate follow-up on a clinical basis alone, with future imaging based on patient symptoms.\textsuperscript{2,16,17} Clearly any patient presenting with symptoms referable to a pineal cyst (or other pineal mass) deserves imaging follow-up. To our knowledge, however, no study has specifically correlated cyst size with malignant potential.

Currently, some authors advocate imaging follow-up for typical pineal cysts to document stability with time, particu-
larly in cysts larger than 10–14 mm, under the assumption that they may further enlarge or become symptomatic, whereas others advocate only clinical follow-up if there are no atypical features or symptoms related to the pineal cyst.\textsuperscript{7,9,15,16} Some radiologists also recommend follow-up under the assumption that if the cyst changes in size or develops atypical features with time, then it could represent a neoplasm such as a pineocytoma, though this is a specific recommendation made in only 1 of the series we reviewed, which recommended follow-up “for many years” for all pineal cysts.\textsuperscript{23}

The importance of postcontrast T1-weighted MR imaging in evaluating these tumors cannot be overstated because pineocytomas may have an imaging appearance identical to that of pineal cysts on noncontrast CT, T1-weighted precontrast MR imaging;\textsuperscript{5,23} all 3 of the tumors in question were noted to demonstrate cyst wall enhancement on postgadolinium imaging; however, specific characteristics of the cyst wall, namely if it was greater or less than 2 mm in diameter, were not mentioned. Unfortunately, the actual images were not provided for independent review in any of these cases. This finding appears to represent an unusual manifestation of a rare tumor because no additional such examples could be found in the literature or in our institutional records. It is unfortunate that these noteworthy cases are not more thoroughly presented because they are the only 3 cases in the literature describing an appearance of a pineocytoma identical to that of a simple cyst, so verifying that they truly met the criteria for simple cysts would be critical in the determination of whether pineocytomas can actually ever have this appearance. In 2 of the 3 cases, the pineocytoma caused clinical symptoms, which resulted in the patient’s seeking medical attention.

On the basis of our findings, we advocate that if a lesion meets the aforementioned criteria for a typical pineal cyst as set forth by Barboriak et al\textsuperscript{7} (<2 mm thick, no nodularity, etc), then no imaging follow-up is necessary to exclude a pineocytoma. There have been reports of pineal cysts enlarging with time, although this is rare and cysts may even involute; if a patient with a known pineal cyst develops new symptoms, then further imaging would be warranted. For a lesion that does not meet the criteria for a typical pineal cyst on MR imaging or causes clinical symptoms, further imaging and possibly even tissue sampling may be warranted. Although no specific recommendations regarding follow-up of cysts discovered incidentally on CT scans was garnered from our review of the literature, we advocate that if a lesion is 1 cm in minimal diameter or greater, then it is worthwhile confirming that the lesion has features of a simple cyst on MR imaging. Again, the importance of postcontrast thin-section MR imaging in this situation cannot be overstated because some pineocytomas may be indistinguishable from pineal cysts on CT and precontrast T1- and T2-weighted MR imaging; however, in the overwhelming majority of cases, they will show nodular wall or internal enhancement on postcontrast imaging. Although there is no clear lower limit in size for pineocytomas, most appear to be 1 cm or larger, and it would be impractical to perform MR imaging on every incidentally discovered pineal cyst. Therefore, 1 cm seems like a reasonable threshold.

The principal limitation of our study was the number of pineocytomas sampled. This is a reflection of the rarity of this disease and the infrequent occurrence of these tumors, even in large tertiary care centers. Further research in this area with a multicenter trial, pooling the resources of several large institutions in an attempt to obtain a more homogeneous group of pathologically proved pineocytomas with available CT and MR imaging to review, would be worthwhile. An associated
secondary limitation is the relatively poor description of the imaging appearance of pineocytomas and pineal cysts available in the literature reviewed, along with a lack of provided images in most cases (including potentially purely cystic ones), which makes subtle differentiation between these entities challenging. Also, the overall heterogeneity and lack of details of imaging techniques used add additional confounding factors, such as accurately determining wall thickness or the presence of subtle wall nodularity.

The data from our own institution are heterogeneous because many of our own patients with pineocytomas have received various types of imaging using different techniques; some patients had images available, while others did not. To some extent, this can be attributed to a transition within our institution from printed film to PACS, a change in scanners and protocols, the fact that some patients had full diagnostic imaging at outside institutions and were imaged at our institution only for stereotactic localization, and the fact that some of the older images were destroyed and not archived. Nevertheless, a more uniform imaging approach would certainly have been helpful because these factors limited our ability to fully evaluate the lesions, particularly for precise measurements of a cyst wall, though this was not an issue in these patients because none of these lesions were entirely cystic with a thin wall on postcontrast imaging and, therefore, did not require measurements for differentiation from a simple cyst.

Conclusion
In summary, although 2 case series described a single asymptomatic pineocytoma and 2 clinically symptomatic pineocytomas that were reportedly indistinguishable from typical pineal cysts on imaging, these cases were incompletely documented, and this seems to be an unusual occurrence of a rare tumor, on the basis of our experience and the remainder of reports in the literature. Most of these tumors will be at least partially enhancing masses that may contain areas of cystic change or at least nodularity of their walls, which should not be confused with typical pineal cysts. Furthermore, typical pineal cysts should not require imaging follow-up, unless the patient develops new symptoms.

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