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Case Report

Pineal Cystic Germinoma with Syncytiotrophoblastic Giant Cells Mimicking MR Imaging Findings of a Pineal Cyst

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Summary: We report a case of precocious puberty in a 4-year-old boy. Contrast-enhanced T1-weighted MR imaging suggested a pineal cyst with enhancement of the slightly thickened wall and focal wall irregularity. Three-dimensional constructive interference in a steady-state imaging revealed a focal lobulation and a nodule-like area in the lesion. The lesion mimicking a pineal cyst proved to be a germinoma with syncytiotrophoblastic giant cells, on the basis of biopsy and tumor marker results.

Primary pineal tumors are rare in all primary intracranial neoplasms, and most include germ cell tumors, pineal parenchymal tumors, and gliomas (1). Germ cell tumors are classified into six histologic types: germinoma, teratoma (mature and immature teratomas and teratoma with malignant transformation), embryonal carcinoma, choriocarcinoma, yolk-sac tumor (endodermal sinus tumor), and mixed (2). They mainly affect children and adolescents. The germinoma may be divided into two categories: pure germinoma and germinoma with syncytiotrophoblastic giant cells (STGCs; 2, 3). Although clinical diagnostic criteria for germinoma with STGCs have not been firmly established, tumor markers are important for the diagnosis. When patients with germinomas show mild elevation of human chorionic gonadotropin (hCG) levels in serum or CSF, they are generally regarded as germinoma with STGCs (4, 5).

Pineal tumors have various neuroimaging findings by histologic heterogeneity in the pineal region (1, 6). Most pineal tumors grow as solid masses, and a cystic appearance is unusual (6–9). Although germinoma with cystic components has been reported (10–12), the cystic component is seldom confused with a pineal cyst. We herein report a case of a germinoma with STGC that was difficult to differentiate from a pineal cyst on the basis of routine MR imaging findings.

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Case Report

A 4-year-old boy presented with precocious puberty such as a rapid increase of body height (10 cm/y), enlargement of the penis, and growth of pubic hair. The boy also had a severe headache and nausea in the morning a few weeks before admission. The boy did not demonstrate neurologic signs or symptoms at the time of admission. The results of endocrine examination revealed a high level of serum testosterone (413 ng/dL), serum hCG (17.1 mIU/mL), serum hCG-β (9.8mlU/mL), and CSF hCG-β (20.7 mlU/mL). The serum alpha-fetoprotein (AFP) level was within the normal range.

CT showed a simple, well-defined cyst without calcification in the pineal region. There were no apparent masses in the testis and adrenal gland on the whole-body CT scans. T1- and T2-weighted MR images showed a $16 \times 13 \times 13$ mm cyst in the pineal region. The content of the pineal cyst was homogeneous and isointense relative to CSF. Contrast-enhanced T1-weighted imaging demonstrated a pineal cyst with enhancement of the slightly thickened wall and focal wall irregularity (Fig 1A). 3D constructive interference in a steady-state (CISS) imaging revealed a focal lobulation and a nodule-like area in the lesion (Fig 1B). Thickness of the wall was about 2 mm, and the size of the nodule-like area was $1.8 \times 1.8 \times 3.0$ mm. Although it was difficult to distinguish the lesion from a pineal cyst on the basis of MR imaging findings, the CISS images demonstrated detailed internal structures of the lesion.

The patient underwent a biopsy for diagnosis. Histologic examination of the biopsy specimen was consistent with a germinoma. It consisted of uniform cells with large, vesicular nuclei, prominent nucleoli, and clear, glycogen-rich cytoplasm. The large round cells had intracytoplasmic globules that stained positively with periodic acid Schiff stain. Immunohistochemical study was positive for placental alkaline phosphatase and negative for hCG and AFP in the limited biopsy samples. The Mib-1 proliferation index of the lesion was 10–15%. Although STGCs were not observed in the biopsy specimens, the lesion was conclusively diagnosed as a germinoma with STGCs on the basis of the results of the tumor markers.

Discussion

Although cystic components have been thought to be rare in germ cell tumors other than teratomas (6–9), recent studies have reported that germ cell tumors frequently have cystic components (10–12). Engel et al (9) reported the findings of MR imaging and pathologic examination in 13 cystic pineal lesions. The histologic findings revealed six pineocystomas, four glial cysts, an arachinoid cyst, a low-grade astrocytoma, and a teratoma. Five of six pineocytomas were well-defined, cystic lesions of 8–20 mm in diameter, which were indistinguishable from glial cysts at MR imaging. Liang et al (10) described that cystic

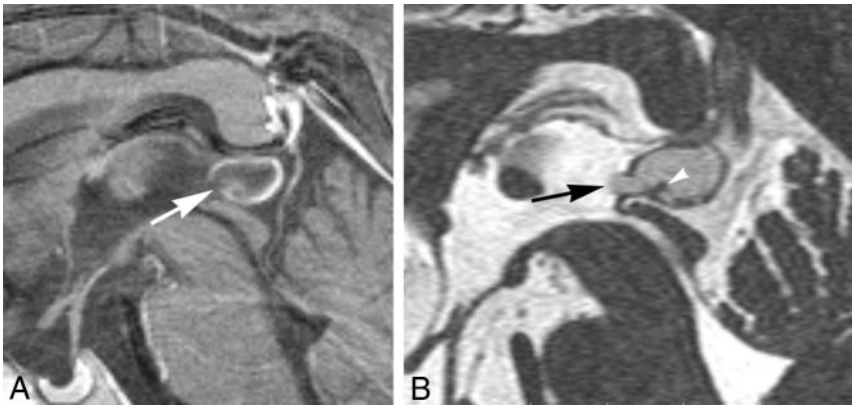

FIG 1. Sagittal MR images obtained in a 4-year-old boy with precocious puberty.

A, Sagittal contrast-enhanced T1-weighted MR image shows a 16 × 13 × 13 mm pineal cyst with enhancement of slightly thickened wall and focal wall irregularity (*arrow*).

B, 3D CISS imaging reveals a focal lobulation (*arrow*) and a nodule-like area (*arrowhead*) of the lesion. Although it is difficult to distinguish the lesion from a pineal cyst even on the CISS images, the morphological description of the lesion is atypical for a pineal cyst.

components were found in 17 of 27 patients with germ cell tumors. The incidence of having cystic components was 44% in germinomas and 100% in nongerminomatous germ cell tumors. The size and number of the cysts were variable. Liang et al concluded that the high incidence is probably due to the improved spatial resolution of MR imaging; however, a germinoma mimicking a pineal cyst has not been described in their studies.

Unlike pineal tumors, pineal cysts are benign, usually asymptomatic, and found incidentally at MR imaging. The prevalence found by using MR imaging in healthy subjects is 1.4–4.3% (13). The pineal cysts are lined by collagenous fibers, glial cells, and normal pineal parenchymal cells (13). Although the size of the cyst may change at follow-up, they are not associated with specific clinical findings (14). The pineal cysts are round and smoothly marginated at MR imaging and have thin smooth walls no more than 2 mm thick (15, 16). The contents of the pineal cyst are usually homogenous and are either isointense relative to CSF or diffusely hyperintense (1, 14). Nodular, rim, or irregular enhancement of the cyst wall may be seen (16, 17). The enhancement pattern may be due to fragmentation of the pineal parenchyma as the cyst enlarges (16). The variable MR imaging appearance of benign pineal cysts may make them indistinguishable from other pineal-region tumors (16). Although it has been recommended that particularly large pineal cysts (>1.4 cm) undergo follow-up imaging (18), no evidence has been presented to suggest that large cysts are more likely to grow or become symptomatic.

In our case, two things were unusual as a pineal cyst. The first was that the patient had specific symptoms that could be referred to a suprasellar or pineal tumor. Second, MR imaging findings were not typical, but were rather atypical, for a pineal cyst. The overall size of the cyst was also atypical for a pineal cyst. On the basis of these unusual conditions for a pineal cyst, surgical biopsy was performed.

CISS images provided additional anatomic details of the cystic lesion compared with conventional MR images in this case. CISS images have been applied for the assessment of small structures adjacent to or in the CSF (19). Because the pineal gland is a small structure and surrounded by the CSF, high-spatial-resolution images like those of CISS might be more useful for identifying internal structures of the pineal lesions. Although CISS usually shows the pineal cyst as smooth and thin with a hypointense wall containing homogeneous hyperintense components similar to those of CSF, further studies regarding the CISS appearance of benign pineal cysts are needed.

Conclusion

We reported a germinoma with STGC that was not completely distinguishable from a pineal cyst on the basis of routine MR imaging findings. CISS imaging was useful for identifying detailed internal structures of the lesion. Tumor markers and biopsy were essential for accurate diagnosis.

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