Pilocytic astrocytoma arising from the pineal gland

A 36-year-old man presented with a one-month history of diplopia of sudden onset, fluctuating on and off with spontaneous remission. Two weeks prior to hospital admission, he developed mild bitemporal headaches and dizziness. Neurological examination was normal. Magnetic resonance imaging (MRI) revealed a round, well-demarcated cystic lesion arising in the pineal gland, causing it to expand and, thus, compress the mesencephalon, posterior recess of the third ventricle, splenium of the corpus callosum, thalamus and cerebellar vermis, leading to compensated obstructive hydrocephalus. The lesion was hypointense compared with the gray matter on T1-weighted images (Fig. 1A), isointense compared with CSF (cerebrospinal fluid) on T2-weighted images and hyperintense compared with CSF on proton density-weighted images (Fig. 1B), and showed only focal peripheral enhancement (Fig. 1C) after administration of gadolinium diethylenetriaminepentaacetic acid.

The lesion measured \(2.5 \text{ cm} \times 3.2 \text{ cm} \times 2.9 \text{ cm}\). Serum levels of alpha-fetoprotein (AFP), beta-human chorionic gonadotropin (β-hCG) and carcinoembryonic antigen (CEA) were normal. A suboccipital craniotomy was performed and CSF was collected directly from the cistern of the foramen magnum. The tumor was approached via a supracerebellar infratentorial route and microsurgical gross total resection was achieved by the senior author (A.R.V.) after evacuation of the citrus-yellow cystic content.

Histological examination showed a cystic mass with a wall that was thin in parts, and thick and more cellular in the rest. The cellular area was composed of bipolar astrocytes in a dense fibrillary matrix with scattered granular eosinophilic bodies and Rosenthal fibers, all features of a pilocytic astrocytoma (Fig. 2). There was evidence of previous hemorrhage into the cyst. Residual pineal parenchyma was seen at the periphery of the tumor with occasional pineocytes interspersed among the tumor cells. CSF examination was negative for neoplastic cells. Eight years after surgery without adjuvant therapy, the patient was well, with normal visual acuity and no radiological evidence of recurrence of the tumor (Fig. 3).

Pineal lesions include benign or malignant tumors [5] and non-neoplastic cysts. Tumors at this location comprise 0.4—1.0% of intracranial tumors, and can be classified into two major types: germ-cell tumors and neoplasms derived from pineal parenchymal cells[3]. Although the pineal gland contains a significant number of astrocytes, the occurrence of a primary low-grade astrocytoma of the pineal gland is rare. To our knowledge, there are only 10 cases of well-documented low-grade glial tumors definitively arising from the pineal gland [1—3]. Difficulty in the diagnosis most frequently arises in distinguishing low-grade astrocytomas in the pineal region from the more common, non-neoplastic pineal cysts. Imaging studies are useful, but cannot reliably make the diagnosis. Both lesions typically appear cystic,
although low-grade astrocytomas may have higher signal intensity on MRI or be identified by the presence of a mass lesion. In some cases, as with our patient, the definitive diagnosis is only possible by pathological examination.

Although diffusion-weighted imaging was not performed, we believe it would be useful to make quantitative diffusion-weighted imaging studies with ADC measurements in such cases in future, as this can help to differentiate pilocytic astrocytomas from non-neoplastic cysts and high-grade gliomas [4] on the basis of tumor cellularity.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

References


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Figure 3  Postoperative MRI shows no residual lesion.