Intra- and Suprasellar Germinoma Mimicking a Pituitary Adenoma

—Case Report—

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Abstract

A 25-year-old female presented with amenorrhea and bitemporal hemianopsia without diabetes insipidus due to an intra- and suprasellar dumbbell tumor. The preoperative and operative diagnosis was pituitary adenoma, but the histological diagnosis was germinoma. Differential diagnosis between germinoma and pituitary adenoma based on computed tomograms and magnetic resonance (MR) imaging features and signal intensity characteristics was difficult in this patient. An enhanced mass posterior to the normal pituitary gland on the mid-sagittal MR image normally indicates an intrasellar germinoma rather than a pituitary adenoma, but this finding was equivocal in our patient.

Key words: diabetes insipidus, germinoma, intrasellar tumor, magnetic resonance imaging, pituitary adenoma

Introduction

Most intra- and suprasellar germinomas are correctly diagnosed based on the characteristic initial symptom of diabetes insipidus in teenagers. Early diagnosis of such germinomas is of the utmost importance as they are highly radiosensitive, but a correct diagnosis may occasionally be difficult to achieve in a patient with a pituitary region tumor without diabetes insipidus.

Intrasellar germinomas are classified into two types: tumors growing primarily in the sella turcica and tumors originating in the suprasellar region and extending into the sella turcica. A definite diagnosis of the former is difficult because almost all such tumors also have a significant amount of suprasellar component. Previously reported magnetic resonance (MR) imaging findings of intrasellar germinomas in five patients were not pathognomonic.^{1,2,4,8,9)}

We report a female patient who presented with amenorrhea and bitemporal hemianopsia but

without diabetes insipidus, due to a large intra- and suprasellar dumbbell-shaped germinoma, mimicking a pituitary adenoma on neuroimages.

Case Report

A 25-year-old female consulted an ophthalmologist for evaluation of failing vision. She was found to have bitemporal hemianopsia, and was referred to us for further examinations in October, 1991. She had become amenorrheic 5 years previously. At that time, her luteinizing hormone (LH) level was below 0.5 mIU/ml and follicle-stimulating hormone (FSH) level was low at 0.8 mIU/ml, but her prolactin (PRL) level was within normal limits, so no treatment was offered. She had remained amenorrheic since. She had no history of polyuria and polydipsia. Her family history was non-contributory.

Physical examination on admission found she was 149 cm in height and weighed 38 kg. No galactorrhea

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Fig. 1 Precontrast CT scans at two contiguous levels, showing a homogeneous, slightly high density mass in the intra- and suprasellar region.

was observed. Her pubic hair was scanty. Her visual acuity was correctable by lenses. Neurological examination found no abnormalities except for the visual field defects. Upward gaze was normal. Her daily urinary output ranged from 1300 to 1500 ml and the urine specific gravity was around 1.010 g/cm³. Endocrinological examinations showed low base values of LH (<0.5 mIU/ml), FSH (<0.5 mIU/ml), and thyroxine (0.94 ng/dl) but the PRL level was normal. Triple loading tests with thyrotropin-releasing hormone, LH-releasing hormone, and insulin showed poor responses of the adrenocorticotrophic hormone-cortisol system. Steroid administration elicited neither hypernatremia nor an increase in urinary output.

Plain skull x-ray films showed an apparently normal sella turcica and no abnormal calcifications. Computed tomography (CT) showed an intra- and suprasellar mass extending toward the foramen of Monro, which showed homogeneously slightly high density (Fig. 1). Postcontrast CT was not performed preoperatively, but the residual tumor was homogeneously enhanced postoperatively.

The mass was isointense to the gray matter on T₁- and T₂-weighted MR images, and markedly enhanced by gadolinium-diethylenetriaminepentaacetic acid (Gd-DTPA). Retrospectively, a high intensity slit-like structure could barely be seen anterior to the intrasellar mass on the mid-sagittal T₁-weighted MR image, which was enhanced by Gd-DTPA (Fig. 2A, B). There were two cleavage planes or notches, one corresponding to the anterior cerebral arteries and the other to the diaphragma sellae (Fig. 2C, D).

The tumor, exposed *via* a right pterional approach, was grayish red and composed of a soft,

Fig. 2 A: T₁-weighted mid-sagittal MR image (repetition time 600 msec/echo time 300 msec, slice thickness 3.0 mm), showing an intrasellar mass with large suprasellar extension and a high intensity slit-like structure anterior to the mass (arrows). B: T₁-weighted mid-sagittal MR image with Gd-DTPA, showing marked enhancement. A slit-like structure anterior to the mass (arrows) is barely visible. C: T₁-weighted parasagittal MR image (7 mm right of the midline) with Gd-DTPA, showing two cleavage planes at the levels of the anterior cerebral arteries (upper arrow) and the diaphragma sellae (lower arrow). D: T₁-weighted coronal MR image with Gd-DTPA.

suckable portion and a fibrous, non-suckable portion. The intra- and suprasellar tumor was subtotally removed and the markedly elevated optic chiasm was decompressed. The histological diagnosis was germinoma with the typical "two-cell pattern" (Fig. 3).

Her postoperative course was uneventful except for a transient episode of polyuria. Bitemporal hemianopsia persisted.

Fig. 3 Photomicrograph of the tumor specimen, showing a typical "two-cell pattern" appearance. Hematoxylin and eosin stain, $\times 200$.

Human chorionic gonadotropin (HCG), alpha-fetoprotein, carcinoembryonic antigen, and placental alkaline phosphatase levels in blood and cerebrospinal fluid (CSF) were determined. All were normal except for a transient elevation of HCG (8.9 mIU/ml). Cytological examination of the CSF disclosed no abnormal findings either.

Postoperative irradiation (23.4 Gy) resulted in shrinkage of the residual tumor on the repeat postcontrast CT scan. After 52 Gy irradiation the tumor was scarcely visible by postcontrast CT. She has been well without hormonal replacement. Follow-up MR images and evaluations of tumor markers in her serum and CSF have remained normal for 31 months following radiotherapy. Bitemporal hemianopsia persisted. The effect of surgical manipulation on the chiasm cannot entirely be excluded.

Discussion

Reported intrasellar germinomas include detailed clinical information in 41 of 46 cases.¹⁻¹⁰ Initial symptoms appeared at ages ranging from 8 to 32 years (mean 15 yrs). There is a female predominance (25 females vs. 16 males).

Intrasellar germinomas have been classified into two types. Intra- and suprasellar germinomas generally tend to arise from the hypothalamus and extend to the posterior pituitary lobe through the pituitary stalk. Therefore, the earliest manifestation of tumors of primarily suprasellar origin (suprasellar type) is most often diabetes insipidus. This is usually also true for tumors of primarily intrasellar origin (intrasellar type), but diabetes insipidus is sometimes not apparent¹⁰ and visual disturbance

occurs in the late stage of the disease.⁸ Radiographically, an enlargement of the sella turcica is found frequently in the intrasellar type tumor, although a normal sella turcica has been reported in several such cases.^{2,4,9}

In our patient, the initial symptom was amenorrhea without diabetes insipidus. Diabetes insipidus is induced when the neurohypophysis and hypothalamus are affected but the normal functions of adenohypophysis remain. If both the neurohypophysis and adenohypophysis are disturbed, diabetes insipidus is masked.³ In our patient both the neuro- and adenohypophysis may have been disturbed at the beginning of the disease. Visual disturbance appeared 5 years later. These findings suggest that the tumor arose primarily in the sella turcica and extended suprasellarly. Nonetheless, the size and shape of the sella were radiographically normal.

MR imaging may provide some clues to the diagnosis of intrasellar germinoma, but only five previous papers have described MR imaging features.^{1,2,4,8,9} A small intrasellar germinoma was well visualized in the posterior pituitary on MR images with Gd-DTPA.⁹ An intrasellar germinoma was enhanced posterior to the normal pituitary gland on mid-sagittal MR images and had a cleavage plane at the level of the diaphragma sellae.² In retrospect, MR imaging of our patient also showed a slit-like structure anterior to the intrasellar mass, and two cleavage planes at the levels of the anterior cerebral arteries and the diaphragma sellae. A cleavage plane at the level of the diaphragma sellae is not specific to germinomas. The slit-like structure may have represented the displaced anterior pituitary, but the significance of the finding was not preoperatively recognized. The location of the tumor posterior to the normal pituitary gland and extending along the pituitary stalk could be important in establishing a diagnosis of intrasellar germinoma. However, a very large tumor extending suprasellarly in a dumb-bell shape as in our case would make determination of the site of tumor origin impossible on the basis of neuroimaging findings. Furthermore, differential diagnosis between large intra- and suprasellar germinoma and pituitary adenoma would also be quite difficult, as the CT densities and MR imaging intensities as well as enhancement patterns are similar for both germinomas and pituitary adenomas.

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