Mature Teratoma Arising From the Sella
—Case Report—

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Abstract

A 26-year-old, short statured, obese male presented with a mature teratoma located entirely within the dural confines of the sella manifesting as headaches and progressive loss of vision. He had pan-hypopituitarism. Magnetic resonance imaging showed a large sellar-suprasellar but entirely infradiaphragmatic tumor of varying consistency. The tumor was resected through a trans-sphenoidal route. The tumor had elevated the diaphragma sellae to a significant extent but did not pass through. Histological examination confirmed a mature teratoma.

Key words: teratoma, sella, suprasellar region, pituitary gland

Introduction

Teratomas are benign tumors derived from the pluripotent cells consisting of elements from all three layers, the ectoderm, mesoderm, and endoderm. Teratomas are commonly located in the gonads, anterior mediastinum, retroperitoneum, and the sacrococcygeal region, and occur more frequently in children and young adults than in the older population. Intracranial teratomas are rare and usually found in the pineal region followed by the suprasellar/hypothalamic region.14,17 Teratomas comprise about 0.5% of all intracranial tumors.17 Occasional cases of teratoma are reported in the basal ganglia, cavernous sinus, fourth ventricle, and the sylvian fissure.1,4,15,16,19 Teratomas arising within the sella turcica are uncommon.23 Intrasellar teratoma comprising of only mature elements is extremely rare. We report a case of mature teratoma located entirely in the sellar-suprasellar infradiaphragmatic region.

Case Report

A 26-year-old, short statured, obese male patient presented with complaints of generalized headaches for the past 6 years. He had suffered progressive worsening of vision in both eyes and excessive weight gain for 2 years. Neurological examination revealed severe bilateral, horizontal gaze-evoked nystagmus. Visual acuity was 6/60 in both eyes with bitemporal field deficit. Fundoscopy showed primary optic atrophy. Hormonal assay revealed panhypopituitarism: adrenocorticotropic hormone 11 pg/ml (normal range 10–50 pg/ml), cortisol 1.8 mg/dl (4–18 mg/dl), thyroid-stimulating hormone less than 0.2 μU/ml (0.2–5 μU/ml), free T3 0.2 ng/ml (2.47–4.34 pg/ml), free T4 less than 0.2 ng/ml (0.95–1.8 ng/ml), growth hormone 0.3 ng/ml (2–5 ng/ml), prolactin 7 ng/ml (1.5–9.7 ng/ml), luteinizing hormone less than 0.6 mIU/ml (2–5 mIU/ml), follicle-stimulating hormone less than 0.7 mIU/ml (3–8 mIU/ml), and testosterone 32.8 ng/dl (250–1000 ng/dl). The adenohypophysial hormones showed no response to triple stimulation test (0.5 mg of thyrotropin-releasing hormone, 0.1 mg of luteinizing hormone-releasing hormone, and 6 U of regular insulin). Computed tomography (CT) of the brain showed a hypodense tumor in the sella with a peripheral rim of calcification (Fig. 1A). The suprasellar extension is better visualized on the coronal images (Fig. 1B). Magnetic resonance imaging showed a large mixed intensity sellar-suprasellar tumor extending up to the floor of the third ventricle (Fig. 2). The pituitary gland was not seen distinctly from the mass.

Replacement therapy with hydrocortisone and thyroid hormone was initiated. The sublabial trans-sphenoidal approach exposed the tumor with soft and firm areas. The tumor contents were mainly fat,
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Fig. 1 A: Axial computed tomography (CT) scan of the brain showing a predominantly hypodense sellar mature teratoma with a peripheral rim of calcification. B: Coronal CT scan showing suprasellar extension of the intrasellar mature teratoma.

Fig. 2 A: Axial T₁-weighted magnetic resonance (MR) image of the brain showing a well-defined rounded hyperintense mature teratoma in the suprasellar cistern. B: Sagittal T₁-weighted MR image showing the sellar mature teratoma with a large suprasellar extension up to the floor of the third ventricle.

Fig. 3 Postoperative computed tomography scan showing complete excision of the mature sellar teratoma.

Fig. 4 A: Photomicrograph of the tumor specimen showing mature adipose tissue (arrow), glandular tissue (thick arrow), and well-developed nervous tissue and nerve fibers (arrowhead). HE stain, ×160. B: Photomicrograph of the tumor specimen showing the columnar cell lining. HE stain, ×400.

bony septation, and keratinaceous flakes resembling epidermoid. The entire tumor was resected. The bone within the tumor could be held, dissected from the adjoining structures, and removed in relatively large pieces. The tumor was located underneath the markedly elevated diaphragma sellae. The tumor was adherent to the diaphragma sellae and was torn during dissection, resulting in egress of a large amount of cerebrospinal fluid. The intracranial structures were seen vividly through the window in the diaphragma sellae. Normal pituitary gland could not be identified. Fat was harvested from the thigh and packed in the cavity.

The postoperative course was uneventful. The patient's vision and visual fields improved immediately after surgery. The visual fields returned to near normal and vision was 6/18 in both eyes at follow-up examination 6 months after surgery. He had no diabetes insipidus. Postoperative CT confirmed complete excision of the tumor (Fig. 3). Histological examination showed abundant adipose tissue with foci of calcification (Fig. 4A). A cyst lined by stratified squamous epithelium containing keratinaceous debris was seen. Few spaces lined by pseudostrati-
fied columnar cells resembled respiratory epithelium (Fig. 4B). Collagen tissue and a few nerve fibers were also seen. There was no evidence of any primitive tissue or malignant component. The histological features suggested mature teratoma. Hydrocortisone and thyroid hormone replacement therapy was con-


dition is commonly seen in patients with parasellar teratomas. Our patient presented with panhypopituitarism. Absence of diabetes insipidus after radical clearance of tumor from the sella is unusual. However, preservation of a part of the normal compressed pituitary gland within the sella, presence of pituitary rest cells in the supradiaphragmatic region, or some kind of a compensatory mechanism could be responsible. Sellar teratomas are infrequent and can mimic pituitary adenoma, craniopharyngioma, or a dermoid cyst. Neuroimaging demonstrates mature teratoma as a well-delineated mixed density mass with cysts and calcification. Immature form teratoma appears with smaller and less prominent cysts and calcification but may be associated with peritumoral edema.

The understanding of the natural history of intracranial teratomas is still incomplete, partly due to the low incidence. Intracranial mature teratoma has the potential to undergo ossification, teeth formation, melanotic progonoma, and malignant transformation. One case of a highly ossified endosuprasellar mature teratoma contained more than 150 teeth macroscopically identified during the operation.

Mature teratomas are benign and usually radioresistant. Radical excision is advocated despite the benign histological appearance since long-term outcome after such treatment is excellent. Tumor markers can be helpful in long-term follow up of patients with germ-cell tumors. Deoxynucleic acid flow cytometric studies and tumor markers have identified aneuploidy and malignant phenotype in residual mature teratomas which further emphasizes the importance of complete removal of mature teratomas. A study of 153 intracranial germ cell tumors at different sites showed that 10-year survival rate for mature and immature teratoma is 93% and 86%, respectively. Patients with a teratoma showing malignant transformation had a 3-year survival rate of 50%.

**References**

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