Transsphenoidal surgery of intrasellar germinomas

Report of two cases

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Two patients with diabetes insipidus, hypopituitarism, and an enlarged sella turcica underwent a transsphenoidal operation for the treatment of intrasellar germinomas. Successful transsphenoidal treatment of such neoplasms has not been reported previously. The cases indicate that the diagnostic possibility of intrasellar germinoma should be considered in young patients with combined diabetes insipidus and hypopituitarism, even when the sella is markedly expanded.

KEY WORDS diabetes insipidus • hypopituitarism • intrasellar germinoma • sellar enlargement • transsphenoidal microsurgery

Sellar enlargement that is not associated with increased intracranial pressure, craniopharyngioma, or pituitary adenoma is rare in patients in the first and second decades of life. We report two patients presenting with diabetes insipidus, hypopituitarism, and an enlarged sella who underwent successful transsphenoidal treatment of large intrasellar germinomas.

Case Reports

Case 1

This 20-year-old man was admitted to the Neurosurgical Service of the University of California, San Francisco (UCSF), on May 22, 1982, for the evaluation of progressive loss of vision. During February, 1982, he noted that he was easily fatigued and had global headaches, loss of appetite, polydipsia, and polyuria. Over the next 3 months, he lost 40 lb and noticed that his facial hair was becoming sparse. He experienced decreasing libido, difficulty obtaining and maintaining an erection, and a decrease in testicular size. Four weeks before his admission to UCSF, the headache became more severe, with the addition of a sharp pain behind the left eye. One week later, he noted decreased peripheral vision, which progressed steadily over the next 3 weeks to the point that he could no longer read or drive an automobile. The polyuria had become more severe. The patient sought medical attention at another institution, where an initial evaluation revealed an intrasellar mass with suprasellar extension. He was referred to our service for treatment.

Examination. Physical examination revealed decreased body hair, small testes, and a dense bitemporal hemianopsia. The visual loss in the left eye included the entire inferior nasal quadrant as well as the temporal field. Central vision was reduced in both eyes, and bilateral band atrophy was noted on funduscopic examination. Both optic discs were pale, the left more so than the right. Laboratory evaluation revealed hypopituitarism. The values obtained were: morning cortisol level < 1.0 μg/dl (normal 5 to 20 μg/dl); follicle-stimulating hormone (FSH) < 3 mIU/ml (normal 4 to 20 mIU/ml); luteinizing hormone (LH) 5 mIU/ml (normal 5 to 25 mIU/ml); growth hormone (GH) < 1.0 ng/ml (normal 0 to 9.9 ng/ml); testosterone < 0.1 ng/ml (normal 3 to 10 ng/ml); and prolactin (PRL) 25 ng/ml (normal 0 to 20 ng/ml). Thyroid function tests were normal. Polytomography demonstrated an enlarged sella with truncation of the dorsum sellae. Focal thinning of cortical bone was seen in the anterior and inferior aspects of the sella, slightly to the left of midline. A computerized tomography (CT) scan showed a large
Case 1

A mass arising from the sella with a significant suprasellar component extending into the region of the hypothalamus (Fig. 1).

Operation. On May 24, 1982, the patient underwent transsphenoidal exploration of the sella. The anterior wall and floor of the sella consisted of only thin bone scattered in isolated chips. As the incised dura was removed, tumor herniated through the dural aperture and into the sphenoid sinus. The tumor had two components. The intrasellar portion, which was present slightly to the left of midline, was firm and yellow. The suprasellar component was soft and gray with dense fine fibrous strands. The intrasellar component was totally removed, but the suprasellar portion was adherent and was removed incompletely. The sella was packed with fat and its anterior wall was reconstituted with nasal cartilage.

Histological analysis of the surgical specimen revealed a typical germinoma. A lumbar puncture was performed, and the cerebrospinal fluid contained no tumor cells.

Postoperative Course. The patient’s vision improved dramatically over the next 48 hours, and continued to improve until it was normal by the time he was discharged on the 7th postoperative day. His diabetes insipidus was unchanged at the time of discharge.

The patient received radiation therapy on an outpatient basis. This therapy consisted of a total of 5560 rads to the pituitary fossa given initially through parallel opposed ports with two subsequent boosts through progressively smaller ports centered on the pituitary fossa. He has completed this course of therapy, and at this time is in good health. His vision has remained normal, but his diabetes insipidus and hypopituitarism are unchanged.

Case 2

This 9-year-old girl was admitted to UCSF on February 2, 1976, for evaluation of an enlarged sella. She had begun to have headaches, personality changes, and occasional nausea and vomiting in 1973, at which time a diagnosis of pseudotumor cerebri was made. The patient was treated with adrenal corticosteroids, which resulted in resolution of her symptoms. Three years later, her parents noted that she had polydipsia and polyuria, which became progressively worse over the next 4 months. An endocrine evaluation revealed diabetes insipidus and panhypopituitarism, and the patient was started on desmopressin acetate (DDAVP) and hydrocortisone. X-ray films of the skull showed an enlarged sella with thinning of the dorsum sellae and posterior clinoid processes. A CT scan showed no evidence of suprasellar extension. A pneumoencephalogram showed slight suprasellar extension with no involvement of the hypothalamus (Fig. 2). The patient was referred to UCSF for further evaluation.

Examination. The general physical and neurological examinations showed no abnormalities, and the
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The hallmark of suprasellar germinomas, regardless of type, is the triad of visual disturbance, diabetes insipidus, and hypopituitarism.

Radiological evidence of sellar abnormalities has been noted in patients with suprasellar germinomas.2-5,9,11 The tip of the dorsum sellae can be eroded without expansion of the sella as a direct effect of the suprasellar mass, but in most cases the changes are those resulting from increased intracranial pressure rather than from an expanding intrasellar mass. In the reported cases in which sellar extension was visualized during a therapeutic surgical procedure, the radiological evaluation had often revealed a normal sella.5,7,15 Of the relatively few reports of germinomas in an intrasellar location, most reflect observations made at the time of craniotomy when the surgeons noted apparent intrasellar extension of the suprasellar tumor.2-7,13,15 Reports of postmortem documentation of intrasellar germinomas are rare.1,3,5,7,11

The sella was enlarged in both of our patients. In Case 1, the tumor appears to have extended into the sella either through a patulous sellar diaphragm or through invasion of the pituitary stalk. In Case 2, the suprasellar cistern was nearly normal radiologically. These characteristics indicate that the tumor had either a primary intrasellar origin, or an origin in the infundibular region with downward growth solely through the pituitary stalk into the sella.

To our knowledge, this is the first report of transsphenoidal surgical intervention for intrasellar germinomas. Although this neoplasm occurs rarely in the sella, the possibility should be considered when evaluating young patients with intrasellar pathology and diabetes insipidus in the absence of calcification within the mass. The early history of diabetes insipidus, present in both of our patients, strongly suggests an infiltrative process involving the infundibular and hypothalamic regions, even when the sella is obviously expanded and when radiographic studies do not confirm the presence of suprasellar or hypothalamic involvement.

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References


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