

Unsuspected Recurrent Pituitary Adenoma Presenting as an Orbital Mass

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Purpose: Orbital invasion of pituitary tumors is rare and usually accompanied by optic nerve head pallor and visual loss. We describe a case of unilateral massive orbital invasion by a recurrent pituitary tumor with preserved visual acuity and normal optic nerve appearance.

Methods: Case report.

Results: Progressive proptosis developed 15 years after transphenoidal removal of a pituitary tumor. Based on the radiological appearance and the clinical history, the patient was suspected to have a sphenoid wing meningioma secondary to previous radiation treatment. A combined neurosurgical and orbital approach was used to remove the intraorbital mass, which extended from the cranial cavity through the superior orbital fissure and the optic canal. Histopathologic examination demonstrated a recurrent nonsecreting pituitary adenoma.

Conclusions: Orbital extension of a recurrent pituitary adenoma should be considered in the differential diagnosis of progressive proptosis even in the absence of significant optic neuropathy.

A 47-year-old woman sought treatment in January 1998 with a 2-year history of right-sided proptosis and diplopia; no pain or loss of vision was reported at that time. Her past medical history included hypertension, obesity (163 cm tall, 100 kg) and “emotional instability” that had interfered with her ability to maintain employment and interact socially with others. She also had a history of a nonsecreting corticotrophic pituitary adenoma, which occurred in 1983 with amenorrhea, hypothyroidism, and breast discharge. At that time she underwent a transfrontal craniotomy and surgical resection, but intraoperatively the tumor was found to be infiltrating the dura and not completely resectable. Postoperative irradiation was given. The patient was subsequently treated by an endocrinol-

ogist with daily prednisone and thyroxin replacement. The prednisone was discontinued in 1990 when her basal cortisol levels rose to 21.5 $\mu\text{g}/\text{dl}$ (normal). Endocrinologic evaluation in January 1998 revealed normal serum prolactin, thyroid, and cortisol measurements.

At the time she was seen by an outside ophthalmologist in January 1998, the patient reported a 6-month history of rapidly increasing proptosis of the right eye. Visual acuity was 20/25 OU. Hertel exophthalmometry was 28 mm OD and 15 mm OS. The right pupil was 2 mm compared with 4 mm on the left and very poorly reactive to light. No relative afferent pupillary defect was present. Examination of extraocular motility showed a right exotropia with limitation of elevation and adduction of the right eye. The patient cooperated poorly for automated visual field testing. She had multiple losses of fixation when testing the right eye and refused to submit to visual field testing of her left eye. Despite its unreliability, the visual field in the

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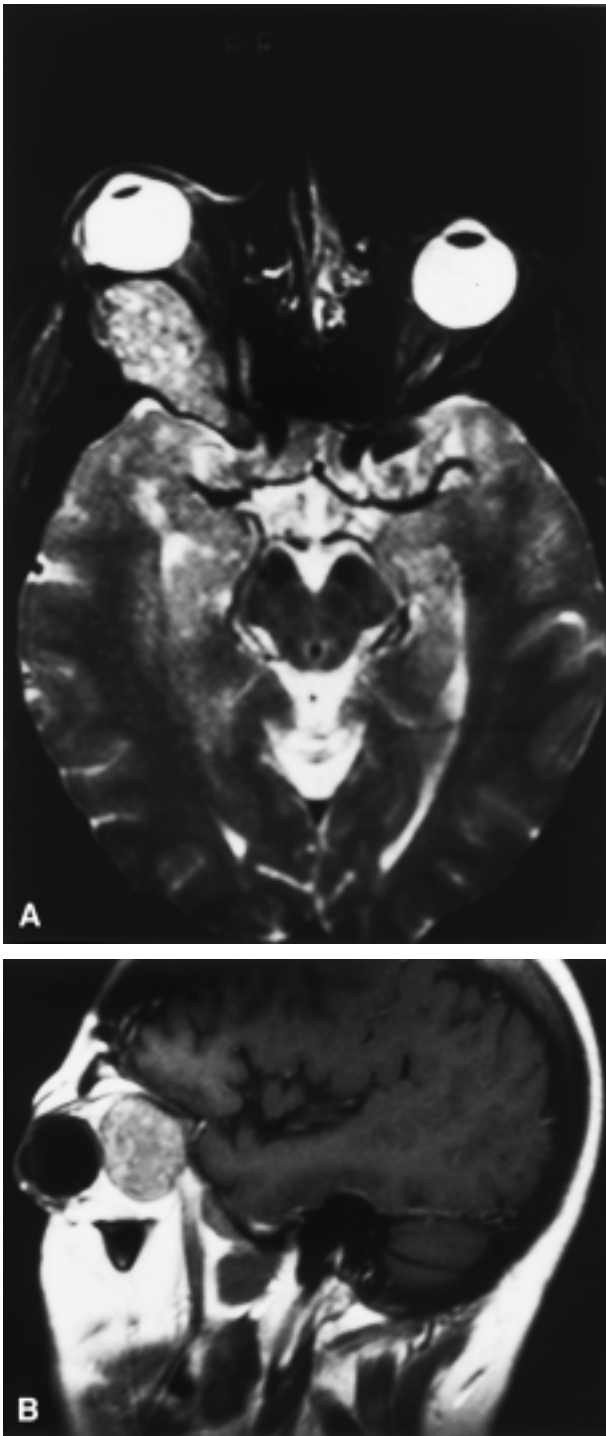


FIG. 1. **A.** Axial magnetic resonance imaging T2-weighted scan showing the intraconal mass extending into the apex of the orbit. **B.** Sagittal magnetic resonance imaging T1-weighted scan of the orbital mass.

right eye suggested a superotemporal scotoma. Color plates and fundus examination showed nothing abnormal.

Both computed tomographic and magnetic resonance image scans showed a homogeneous, slightly enhancing mass filling the right orbit almost entirely and markedly displacing the eye forward (Fig. 1A,B). The mass was noted to elevate the optic nerve superomedially and to extend posteriorly into the apex of the orbit and through the optic canal. No bony erosion or evidence of tumor extension into the suprasellar region was noted (Fig. 2). Neurosurgical consultation evoked the suspicion of a middle cranial fossa meningioma extending into the orbit thought to have occurred secondary to a previous radiation therapy. Despite her previous history, a recurrence of a pituitary adenoma was thought to be unlikely because of the recent onset of proptosis after a long asymptomatic interval, absence of systemic systems, relatively normal visual function, and massive extension unilaterally into the orbit with the normal radiological appearance of the sella. The patient refused proposed surgical resection of the tumor.

Three months later, the patient returned to her ophthalmologist reporting a painful, red right eye. She had increased proptosis with lagophthalmos and exposure keratopathy with an inferior epithelial erosion (Fig. 3). The patient refused testing of visual acuity, pupils, and extraocular motility. She was urgently referred to us because of the keratopathy. The patient was unable to tolerate an attempted tarsorrhaphy under local anesthesia, so a 90% permanent tarsorrhaphy was placed over the

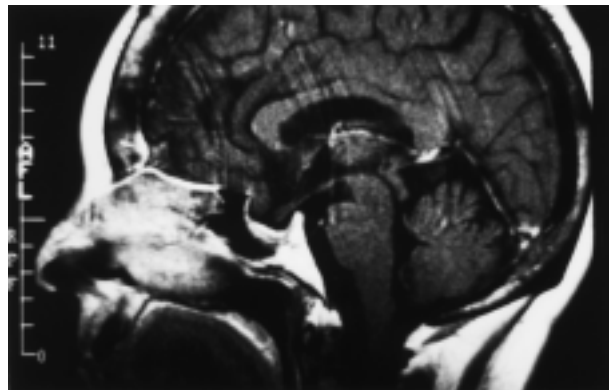


FIG. 2. Sagittal magnetic resonance imaging demonstrating enlargement of the sella turcica and displacement of the optic chiasm into the sella, but no evidence of residual pituitary neoplasm.

right eye using general anesthesia. The corneal defect healed. The patient again refused proposed surgical intervention aimed at alleviating the progressive proptosis, but after additional counseling she consented to combined orbital and neurosurgical removal of the intracranial and orbital mass through a right frontotemporal craniotomy with orbitozygomatic avulsion on July 31, 1998.

A nonencapsulated mass invading the right cavernous sinus, the orbit, and the frontal lobe dura was subtotally resected. Histopathologic examination demonstrated a recurrent nonsecreting pituitary adenoma.

Postoperatively the patient maintained 20/25 visual acuity OU and had no relative afferent pupillary defect. Color vision remained normal in each eye, but the patient refused to undergo automated perimetry. The optic nerve head appearance remained normal. The patient had a large alternating exotropia (Fig. 4) but experienced complete relief of proptosis (Hertel: 16 mm OD, 15 mm OS). When the patient was last examined in July 1999, there was no clinical or radiographic evidence of residual or recurrent tumor.

DISCUSSION

Orbital invasion of pituitary tumors is rare; to our knowledge, only seven cases of orbital extension of a pituitary tumor have been reported since Jackson¹ reported two cases in 1960. These nine previous cases included three prolactinomas,^{2,3} one thyrotropin-secreting hormone-secreting adenoma,⁴ and five reports that did not specify whether an endocrinologic disorder was caused by the tumor.^{1,5-7,10} Ap-



FIG. 3. Patient at the time of referral with marked proptosis, inferior scleral show, conjunctival injection, and exposure keratopathy.



FIG. 4. Appearance of patient 1 year postoperatively, with relief of proptosis but alternating exotropia.

proximately 75% of pituitary adenomas are endocrinologically active. Prolactin-secreting tumors are the most common, followed by adenomas secreting growth hormone, adrenocorticotrophic hormone and, less commonly, thyroid-stimulating hormone.⁸ Nonsecreting adenomas tend to be larger and have a higher recurrence rate (20% between 6 months and 6 years).⁹ In all previous reports of pituitary tumors invading the orbit, visual loss and optic nerve pallor were the common findings in association with unilateral proptosis, extraocular muscle limitation, and systemic symptoms related to the endocrinologic dysfunction. The present case is notable because of the relatively preserved visual function in the affected eye and the normal appearance of the optic nerve head despite massive orbital invasion. Furthermore, the orbital extension dramatically overshadowed the intracranial tumor remnant to such an extent that the radiological and clinical suspicion anticipated a radiation-induced meningioma. The likelihood of recurrent pituitary adenoma was further reduced by the normal endocrinologic state maintained during the 15 years after her initial craniotomy and resection and the long period of stability before the recent onset of rapidly developing proptosis.

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