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Visual Diagnosis Pearl

Giant Prolactinoma Complicated by Cerebrospinal Fluid Leak

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CLINICAL PRESENTATION

A 43-year-old man with no significant past medical history presented to an otolaryngology clinic with four months of persistent clear nasal discharge and a salty taste in the posterior oropharynx. He denied fever, nasal congestion, sinus pressure, or allergic symptoms. Initial evaluation raised concern for chronic sinonasal pathology.

A non-contrast computed tomography (CT) scan of the sinuses unexpectedly revealed a large sellar and suprasellar mass exerting mass effect on the midbrain, pons, and left cerebellar hemisphere, with dehiscence of the sellar floor and communication with the paranasal sinuses. Subsequent magnetic resonance imaging (MRI) of the pituitary demonstrated a giant $8.1 \times 4.5 \times 5.1$ cm pituitary macroadenoma occupying the central skull base, sella, and suprasellar cistern, with extension into the posterior fossa and compression of the brainstem and left cerebellum (Figure 1).

The patient was referred to neurosurgery and endocrinology for further evaluation. On review of systems, he endorsed intermittent generalized headaches and decreased libido over several months but denied visual changes, diplopia, galactorrhea, dizziness, personality

changes, or altered level of consciousness. Formal visual field testing was unremarkable.

Laboratory evaluation revealed a markedly elevated serum prolactin level exceeding 9,400 ng/mL (reference range: 3.8–18.9 ng/mL), consistent with a giant prolactinoma. Total testosterone was decreased at 252.1 ng/dL (reference range: 300.0–890.0 ng/dL), with normal luteinizing hormone and follicle-stimulating hormone levels, consistent with secondary hypogonadism. Additional pituitary testing demonstrated low free thyroxine with an inappropriately normal thyroid-stimulating hormone level, while morning cortisol, ACTH, IGF-1, and growth hormone levels were within normal limits.

Given evidence of an active cerebrospinal fluid (CSF) leak due to erosion of the sellar floor, the patient underwent transsphenoidal resection of the pituitary mass with skull base reconstruction. Histopathology demonstrated a lactotroph adenoma, with immunohistochemical staining positive for prolactin and negative for ACTH, GH, FSH, LH, and TSH.

Postoperatively, the patient was initiated on cabergoline, titrated to 1.5 mg twice weekly, and levothyroxine 100 mcg daily. At three months, prolactin levels decreased to 146 ng/mL and normalized to 6.7 ng/mL by six months.

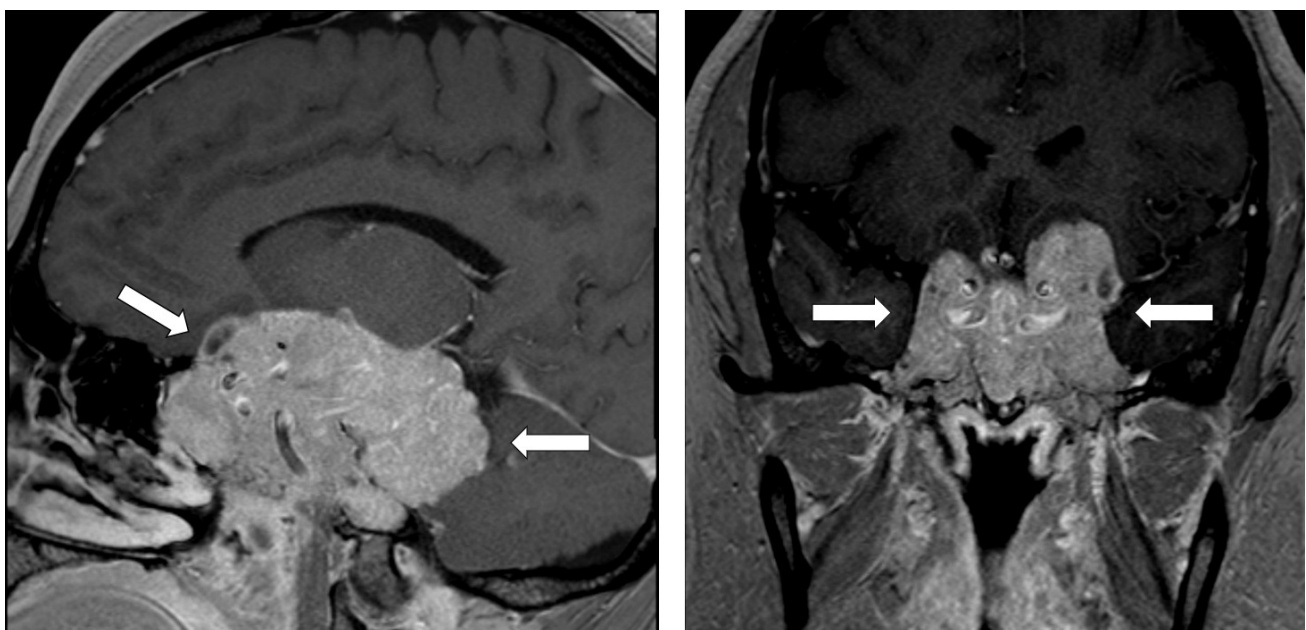


Figure 1. MRI of the pituitary gland (T1-weighted sagittal and coronal views) demonstrating a giant prolactinoma with extensive sellar and suprasellar involvement and posterior fossa extension (see arrows).

Persistent hypogonadism (testosterone 101.3 ng/dL at six months) prompted initiation of testosterone replacement therapy. Follow-up MRI obtained six months postoperatively demonstrated significant tumor debulking, with residual disease confined to the sella and cavernous sinuses and marked improvement in mass effect.

DISCUSSION

Prolactinomas are the most common functioning pituitary adenomas and are classified by size as microprolactinomas (<1 cm) and macroprolactinomas (>1 cm). Giant prolactinomas, defined as lesions exceeding 4 cm in diameter with markedly elevated prolactin levels and extensive extrasellar invasion, are rare and demonstrate a strong male predominance. These tumors tend to be more invasive and biologically aggressive compared with smaller adenomas.^{1,2}

Clinical manifestations of giant prolactinomas are often driven by mass effect rather than symptoms of hyperprolactinemia alone, including headaches, visual field defects, cranial nerve dysfunction, and pituitary hormone deficiencies.³ In this case, the patient's presentation with persistent rhinorrhea and a salty taste was attributable to a CSF leak caused by erosion of the sellar floor—an uncommon but clinically significant manifestation of invasive pituitary disease.

Dopamine agonists remain first-line therapy for giant prolactinomas, with cabergoline favored due to its superior efficacy and tolerability.² Long-term studies demonstrate that medical therapy frequently results in substantial tumor shrinkage and biochemical control, though residual disease is common and necessitates ongoing radiographic and biochemical surveillance.³

Surgical intervention is generally reserved for select indications, including pituitary apoplexy, progressive neurologic compromise, intolerance to dopamine agonists, or complications related to skull base invasion.² Notably, the presence of an active CSF leak represents a clear indication for early surgical intervention due to the risk of meningitis and the inability of medical therapy alone to restore skull base integrity.⁴ In such cases, surgery is typically aimed at tumor debulking and reconstruction, while adjuvant dopamine agonist therapy plays a critical role in achieving long-term disease control.

This case highlights an atypical presentation of a giant prolactinoma and underscores the importance of considering skull base pathology in patients presenting with persistent clear rhinorrhea. Early recognition and multidisciplinary management are essential to prevent complications and optimize clinical outcomes.

TEACHING POINTS

- Persistent clear rhinorrhea accompanied by a salty taste should prompt evaluation for a cerebrospinal fluid leak, even in the absence of trauma or sinonasal disease.
- Giant prolactinomas may present primarily with complications related to mass effect and skull base invasion despite minimal symptoms of hyperprolactinemia.
- An active cerebrospinal fluid leak is a key indication for early surgical intervention, even in tumors that are otherwise responsive to dopamine agonist therapy.



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