



# IgG4-Related Hypophysitis Masquerading as a Pituitary Macroadenoma: A Case Report



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## Abstract

IgG4-related hypophysitis is a rare inflammatory condition that can closely mimic pituitary macroadenoma on imaging. A 44-year-old male presented with progressive visual field loss and was found to have a 14 × 15 × 20 mm enhancing sellar/suprasellar mass compressing the optic chiasm. He underwent endoscopic endonasal resection with intraoperative navigation. Pathology revealed dense lymphoplasmacytic infiltrate, storiform fibrosis, and >40% IgG4-positive plasma cells, consistent with IgG4-related hypophysitis, along with a coexisting Rathke's cleft cyst. The patient recovered well and was referred for systemic evaluation. This case underscores the importance of considering IgG4-related disease in the differential for sellar lesions and highlights the role of histopathology in guiding diagnosis and management.

## Introduction

- IgG4-related disease (IgG4-RD) is a systemic fibroinflammatory condition that can affect nearly any organ, but pituitary involvement is rare.
- IgG4-related hypophysitis (IgG4-RH) is an uncommon cause of sellar masses, often mimicking pituitary macroadenomas on imaging.
- Patients may present with visual field deficits, hypopituitarism, or nonspecific symptoms such as fatigue.
- Radiologic features are often indistinguishable from neoplastic lesions, leading to frequent preoperative misdiagnosis.
- Histopathologic confirmation is essential, revealing a dense IgG4-positive plasma cell infiltrate with fibrosis.
- Early identification is critical to enable systemic evaluation and guide non-surgical treatment options such as corticosteroids.

We present a unique case of IgG4-RH coexisting with a Rathke's cleft cyst in a 44-year-old male with progressive visual decline.

## Methods and Materials

**Patient:** 44-year-old male with progressive peripheral visual field loss (left > right) and fatigue.

**Imaging:** MRI revealed a 14 × 15 × 20 mm avidly enhancing sellar/suprasellar mass compressing the optic chiasm, initially presumed to be a non-functioning pituitary macroadenoma.

**Surgical Approach:**  
Endoscopic endonasal transsphenoidal resection  
Intraoperative stereotactic neuronavigation  
Skull base reconstruction using mucoperiosteal graft  
No lumbar drain or fat graft was required

**Histopathology:**  
Dense lymphoplasmacytic infiltrate  
Storiform fibrosis  
40% IgG4-positive plasma cells per high-power field  
Coexisting Rathke's cleft cyst  
No evidence of adenoma or malignancy

**Postoperative Course:** Uneventful; patient discharged on postoperative day two with stable neurologic status and improved visual fields.

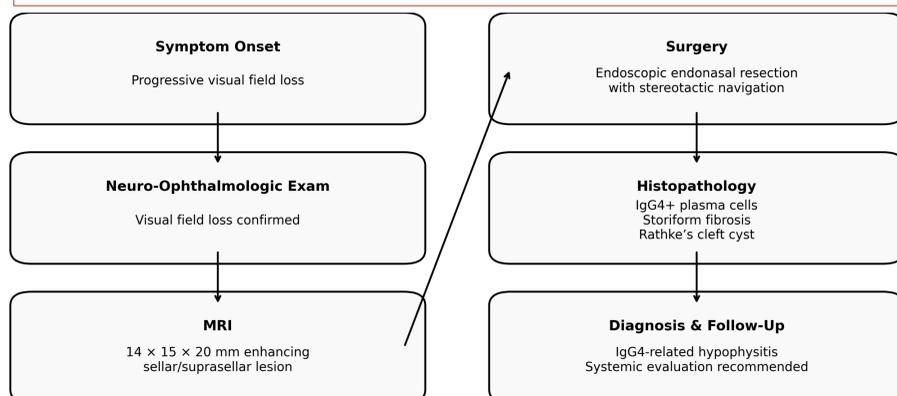


Table 1. Clinical Course Overview.

## Results

The patient underwent successful **endoscopic endonasal transsphenoidal resection** of the sellar/suprasellar mass.

**Gross total resection** was achieved without intraoperative complications. Estimated blood loss was 50 mL.

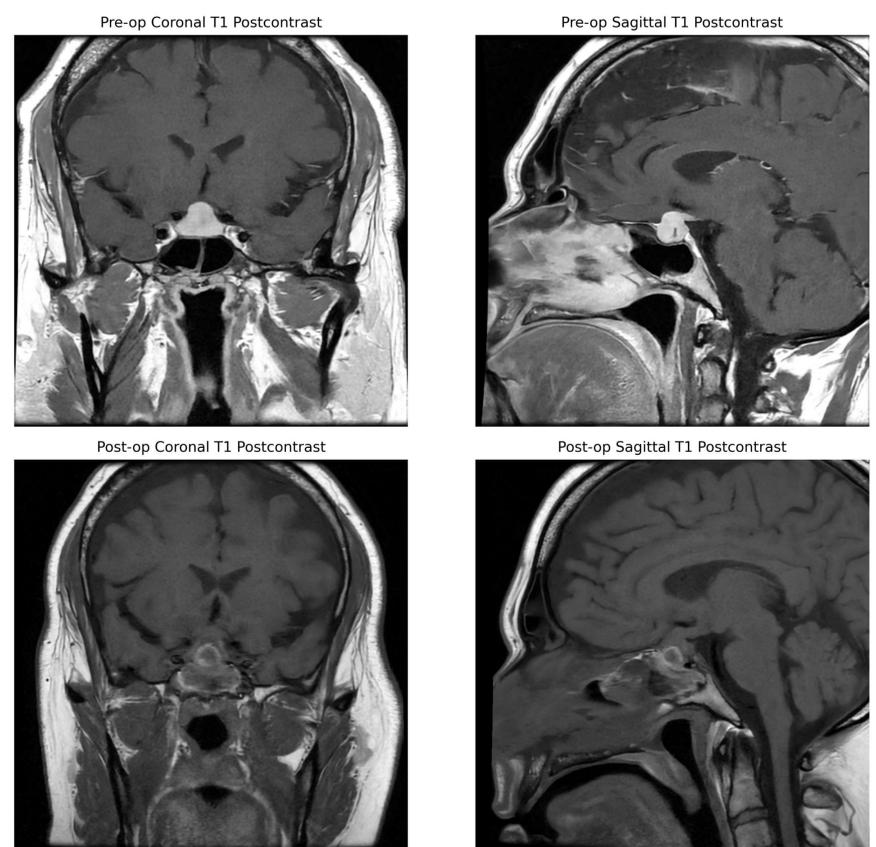
**Postoperative MRI** showed no definite residual enhancing tissue, with changes consistent with resection and reconstruction.

**Histopathology** revealed:

- Dense lymphoplasmacytic infiltrates
- Storiform fibrosis
- 40% IgG4-positive plasma cells per high-power field
- Focal cystic changes consistent with a Rathke's cleft cyst
- No evidence of pituitary adenoma or malignancy

The patient had an **uneventful recovery**, was discharged home on postoperative day two, and reported **improved visual symptoms** on follow-up. He was referred for **rheumatologic evaluation** to assess for systemic IgG4-related disease.

## Pre- and Postoperative MRI Images



## Discussion

IgG4-related hypophysitis is a rare inflammatory condition that closely mimics pituitary macroadenoma in clinical and radiographic presentation, often leading to misdiagnosis. In this case, surgical resection was performed for decompression and diagnosis, revealing classic histopathologic features of IgG4-RD alongside a coexisting Rathke's cleft cyst. While surgery was appropriate, recognition of IgG4-RH is important, as it may respond to corticosteroid therapy and alter long-term management. Postoperative systemic evaluation is essential given the potential for multi-organ involvement. Multidisciplinary follow-up ensures appropriate monitoring and coordination of care.

## Conclusions

This case highlights IgG4-related hypophysitis as a rare but important differential diagnosis for sellar and suprasellar masses. Despite radiographic similarity to pituitary macroadenoma, accurate diagnosis relies on histopathology. Early identification can guide systemic evaluation and potential immunosuppressive therapy. Skull base surgeons should remain aware of this entity, especially in cases with atypical features or unexpected intraoperative findings.

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