เทพธารินทร์
ศูนย์เบาหวาน ไทรอยด์ และต่อมไร้ท่อ
Service Education Research

Interhospital Endocrine Conference

A 33-year-old Indonesian male presented with polyuria and polydipsia for 3 months

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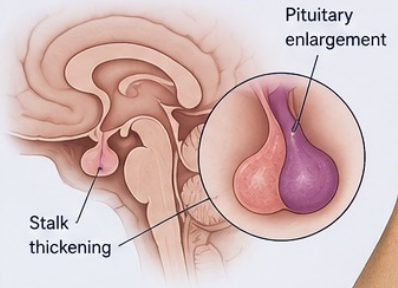
IgG4-related hypophysitis

IgG4-RELATED HYPOPHYSITIS

A rare form of autoimmune hypophysitis part of IgG4-related disease (IgG4-RD)

PATHOPHYSIOLOGY

Immune-mediated infiltration of IgG4-positive plasma cells and fibrosis in the pituitary gland, leading to mass effect and pituitary dysfunction.



CLINICAL FEATURES

- **Headache**
- **Visual disturbance**
- **Hypopituitarism** (e.g., fatigue, decreased libido, secondary adrenal insufficiency)
- **Diabetes insipidus** (less common)

COMMON ASSOCIATIONS WITH IgG4-RELATED DISEASE

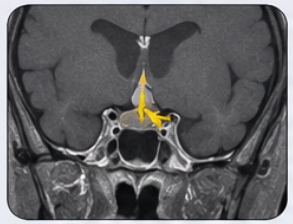
- Mikulicz disease (lacrimal & salivary gland enlargement)
- Autoimmune pancreatitis
- Retroperitoneal fibrosis
- Sclerosing cholangitis
- Renal involvement

DIAGNOSIS

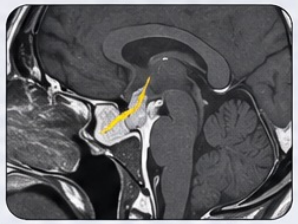
- **MRI:** Pituitary enlargement, stalk thickening, homogeneous enhancement
- **Serum IgG4 elevation** (may be normal in some cases)
- **Other organ involvement** supports the diagnosis
- **Definitive diagnosis:** Histopathology showing lymphoplasmacytic infiltration rich in IgG4-positive plasma cells with storiform fibrosis

MRI FINDINGS (EXAMPLE)

T1-weighted (contrast)
Coronal view



T1-weighted (contrast)
Sagittal view



Enlarged pituitary gland with homogeneous enhancement and thickened infundibulum

LABORATORY FINDINGS

- Elevated serum IgG4 level (>135 mg/dL is suggestive)
- Other pituitary hormone deficiencies as appropriate

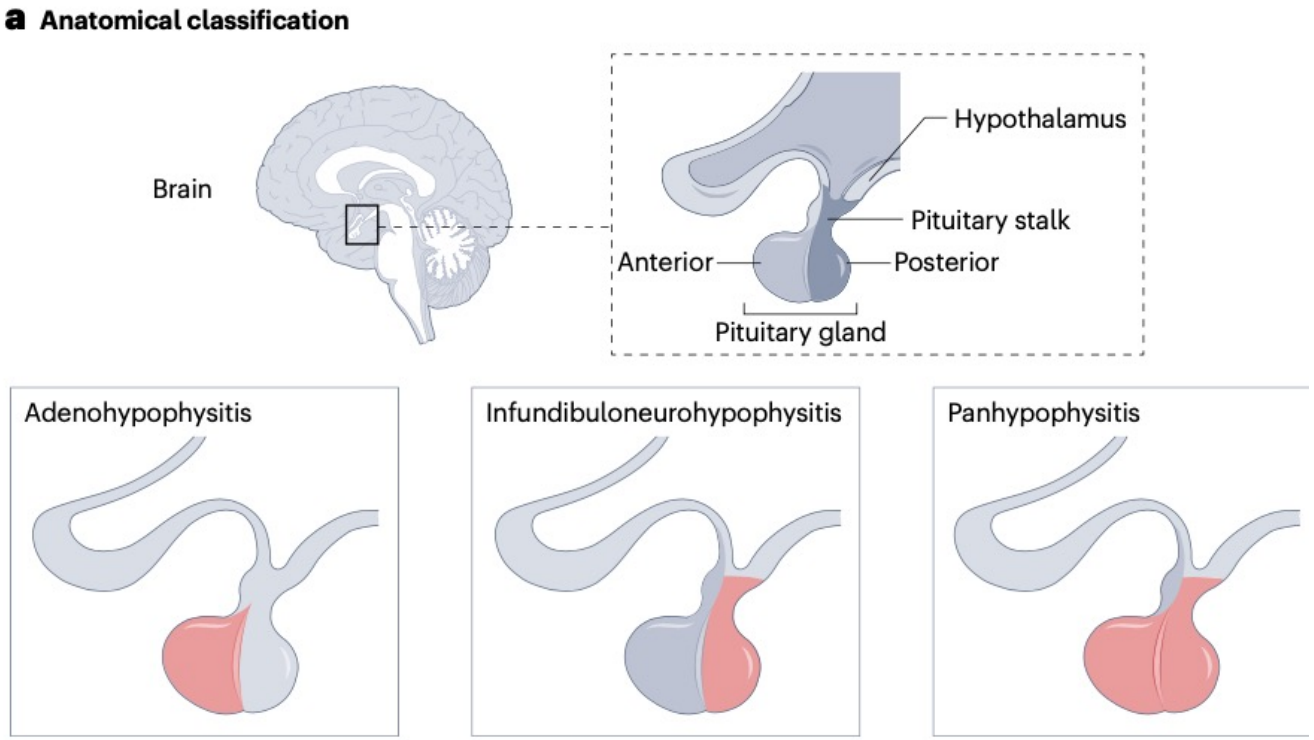
IgG4 level
256 mg/dL
(elevated)

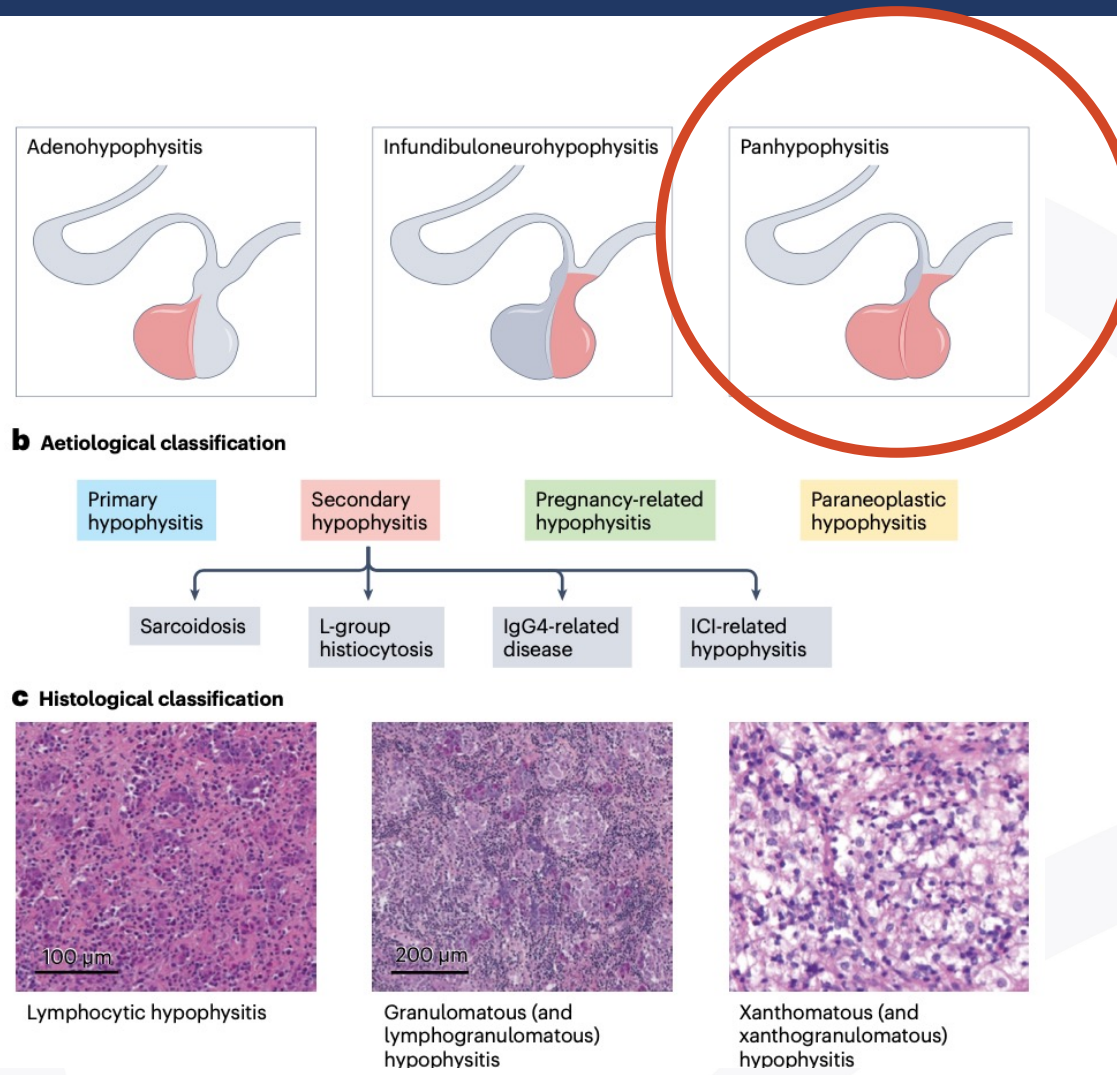
TREATMENT

- **First-line: Glucocorticoids** (e.g., Prednisone 0.6 mg/kg/day) with slow taper
- **Steroid-sparing agents** (e.g., Azathioprine, Mycophenolate mofetil) for relapsing or refractory cases
- Monitor clinical symptoms, pituitary function, serum IgG4, and MRI

KEY POINT IgG4-related hypophysitis should be suspected in patients with pituitary enlargement and hypopituitarism, especially when there is IgG4-RD in other organs. Early diagnosis and steroid treatment can improve outcomes.

Hypophysitis Differential diagnosis





Miquel L, Testud B, Albarel F, Appay R, Graillon T, Cuny T, Dufour H, Ebbo M, Brue T, Jarrot PA, Schleinitz N, Castinetti F. An update on hypophysitis. Nat Rev Endocrinol. 2026 May;22(5):287-301. doi: 10.1038/s41574-025-01200-1. Epub 2025 Nov 27.

IgG-4 related hypophysitis

- IgG4-related disease (IgG4-RD)
  incidence 0.28/100,000 patients
- IgG4 hypophysitis 1.5% of IgG-4 RD
- Diagnosis criteria as in table
  Definite diagnosis: Histopathology
  Probable: MRI + biopsy
  Possible: MRI + IgG4 level + response to treatment
- IgG-4 level correlates with number of organs involved
- Management:
  start with Prednisone 30–40 mg/day or 0.6mg/kg/day

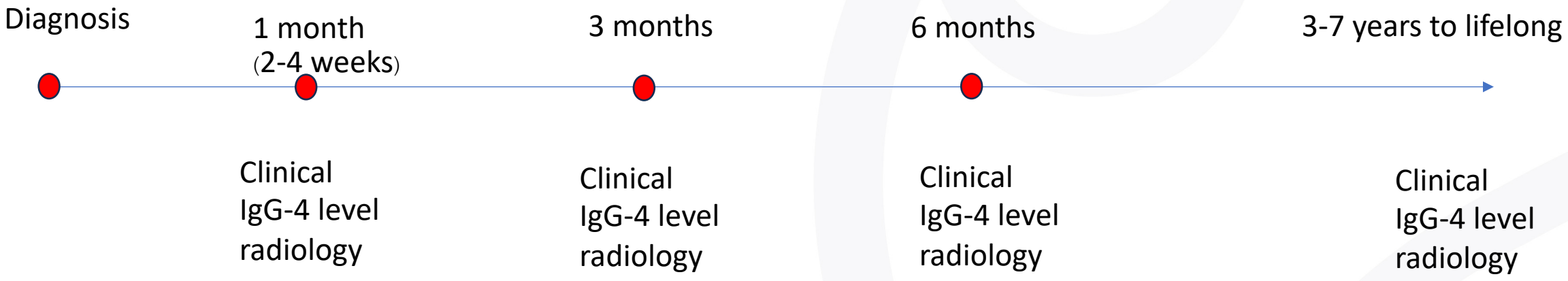
Criteria	
Histopathology	mononuclear infiltration of the pituitary gland, rich in lymphocytes and plasma cells, with >10 IgG4-positive cells/high-power field
MRI	sella mass or thickened pituitary stalk
Biopsy	involvement of other organs
Serology	IgG4 > 140 mg/dL
Treatment	clinical and radiological improvement with corticosteroids
Established diagnosis	1 or 2+3 or 2+4+5

Angelousi, A., et al. (2020). Hypophysitis (Including IgG4 and Immunotherapy). Neuroendocrinology, 110(9-10), 822–835.

Khosroshahi, A., et al (2015). International Consensus Guidance Statement on the Management and Treatment of IgG4-Related Disease. Arthritis & rheumatology (Hoboken, N.J.), 67(7), 1688–1699.

IgG4-related hypophysitis

- Clinical course
  > 50% relapsed-remitting,only 30% able to discontinue glucocorticoid
  Treatment
  First-line: prednisolone 30-40 mg/day then taper or to maintenance
  Steroid-sparing agent: add Rituximab or conventional immunosuppresive if failure to taper off prednisolone



Taper dose 10 mg/day in 2-4 weeks or longer then decreasing 5 mg/day q 2-4 weeks

Khosroshahi, A., et al (2015). International Consensus Guidance Statement on the Management and Treatment of IgG4-Related Disease. Arthritis & rheumatology (Hoboken, N.J.), 67(7), 1688–1699.

Take home messages

1. **IgG4-related hypophysitis**
   - MRI **smooth and Homogenous enhancement lesion**
   - MRI **decreasing in pituitary stalk thickness after treatment**
   - **Clinical, imaging, IgG4 level** responsive to corticosteroid treatment
2. **Tissue biopsy of Pituitary stalk lesion – challenging**
   - Multidisciplinary approach
   - High risk procedure (Mayo series 24%, other series 40-60% in PSL)

   >without any preoperative hypopituitarism, 30% new pituitary hormone abnormality
   >at least 1 pituitary hormone deficiency at baseline, 55% new horomone deficiency
   >without preexisting AVP-D, 46% develop AVP-D
   >other complication: CSF leak, infection
3. **Pituitary Stalk Lesion biopsy Indication**
   1. Progressive stalk lesion
   2. Suspected Metastases, Malignancy tumors
   3. Unresponsive to treatment