Differential Diagnosis of Sellar Lesions

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Differential Diagnosis of Sellar Masses

1. Cystic Lesions
   Rathke’s cleft cyst, Craniopharyngioma, Arachnoid Cyst

2. Other Neoplasms
   Meningioma, Germ Cell Tumor, Chordoma, Granular Cell Tumor, Pituicytoma, Glioma, Metastases, Lymphoma

3. Inflammation/Infection
   Sarcoidosis, Tuberculosis, Pituitary Abscess, Langerhans’ Histiocytosis, Lymphocytic Hypophysitis

4. Empty sella syndrome

5. Pituitary Adenomas (91%)
Case

34 yo F presents with headache, that eventually led her primary care physician to get a brain MRI. Prolactin 55 ng/mL, other hormones within normal limits.

Rathke’s cleft cyst

Originate between anterior and posterior lobe from embryologic Rathke’s pouch. Anterior lobe forms from anterior wall of Rathke’s pouch and pars intermedia forms from posterior wall of Rathke’s pouch.

Rathke’s Cleft Cyst

Incidence – 13-22% of autopsies and “volunteer” MRIs
Symptoms – headache (60%) most common; 35-50% hormonal dysfunction; 20-50% visual disturbance

Rathke’s Cleft Cyst - MRI

Differential diagnosis is usually cystic adenoma vs Rathke’s cleft cyst, Rathke’s cleft cyst defined by:

1. Stalk typically midline
2. abnormality between anterior and posterior lobe

1. Two types:
   i. T1 dark, T2 bright (one third) – fluid like CSF
   ii. T1 bright, T2 variable (two third) – fluid is mucus, higher rate of presenting with h/a (100%) or gland dysfunction (40%); higher rate of postop diabetes insipidus than adenomas

Source: JNS 102: 189, 2005
**Rathke’s Cleft Cyst Treatment**

1. transsphenoidal surgery if larger than 1 cm or clearly symptomatic (microscope usually used in lieu of endoscope if cyst is intrasellar)
2. access cyst through vertical midline cut in gland or by elevating gland to get under it.
3. need to drain cyst and remove part of wall
4. Try to send portion of cyst contents to path to look for inflammation and squamous metaplasia
5. alcohol irrigation if no CSF encountered
6. Gelfoam in cyst cavity rather than fat graft
7. Consider immediate postop MRI given rate of recurrence

**Rathke’s Cleft Cyst**

Cyst wall typically has ciliated columnar or cuboidal epithelium with interspersed mucin-producing goblet cells, but squamous metaplasia can occur and increases risk of recurrence by as much as 3- to 4-fold

**Effect of surgery on Rathke’s symptoms**

1. rate of headache improvement 60-80%
2. rate of hormone dysfunction improvement 20%

Source: JCEM 84: 3972, 1999

**Rathke’s Cleft Cyst Recurrence**

20% recur within 5 years, most within first 2 years

Source: JCEM 84: 3972, 1999
Factors predicting recurrence

118 cases from USC retrospective review
Radical resection of whole wall with no impact on recurrence, but increases postop DI from 9% to 42%
Fat graft increases recurrence rate
Squamous metaplasia increases recurrence

Source: JNS 102: 189, 2005

Pituitary Arachnoid Cyst

Congenital or acquired; herniation of arachnoid membrane through incompetent diaphragm or arises from arachnoid rests below the diaphragm
Symptoms – 90% have headaches, 35% have visual field defect, 35% have hypopituitarism
MRI – similar to Rathke's cleft cyst containing watery contents rather than mucus, T2 bright T1 dark
Treatment – transsphenoidal drainage and wall biopsy, headache gets better but endocrine function improvement worse than with other cysts. 20% recurrence at 10 years

Craniopharyngioma

Age 5 to 10
Histologically benign looking tumor (developing from nests of epithelium derived from Rathke's pouch) with malignant behavior due to invasion of surrounding structures and recurrence after gross total resection.
Single large or multiple cysts filled with turbid, proteinaceous brown-yellow material that sparkles due to cholesterol crystals.
Usually start sellar, can have suprasellar extension with invasion into hypothalamus.

Craniopharyngioma

Radiation used for residual tumor after surgery, although hypopituitarism and other side effects in the pediatric population will ensue.
10-year recurrence rate – 83% for tumors larger than 5 cm, 20% for tumors smaller than 5 cm
Overview of cystic pituitary lesions

Psychiatric disturbances unique to craniopharyngioma, get better with surgery
Arachnoid cyst patients present older (fifth decade of life)
Endocrine dysfunction rare in arachnoid cyst, if present in arachnoid cyst won’t get better with surgery

Source: JCEM 84: 3972, 1999

Cystic Pituitary Lesions - Recurrence

Arachnoid cysts – 20% recur after 10 years; Rathke’s – 20% recur within first year or 2; Craniopharyngioma – most recur, spread over 10 years

Source: JCEM 84: 3972, 1999

Case

33 year old female with severe headaches, n/v, weight loss, hyperprolactinemia, hypoadrenalism, hypothyroidism.

MRI with intrinsically T1 bright, T2 intermediate signal 3-4 mm lesion in left inferoposterior sella

Pituicytoma

New diagnosis created by WHO in 2007 – WHO grade I tumor of the pituicytes, which are stromal (supporting) cells of the posterior lobe or stalk.
Present with visual field defect, or asymptomatic incidental finding on MRI
More vascular than pituitary adenoma and tough to access (posterior lobe, sometimes stalk) so surgery risky.
**Pituicytoma**

- Five cases at UCSF 2007-2009
- Mean age 53 (range 33-71)
- 3 men, 2 women
- 1 incidental finding; 2 panhypopituitarism, 1 visual symptoms; 2 with over 20 lb weight loss

**GFAP+**

- MIB-1 0.5-2.0%
- No hormones, no neurosecretory granules
- Well circumscribed
- Low recurrence rate but readily symptomatic due to location

**Metastasis to the Pituitary Gland**

1% of sellar lesions are mets - Breast, Lung most common

Posterior Lobe more common than anterior lobe (direct blood supply), also can grow along stalk

Patients 5-10 years younger than average brain met patient

Posterior lobe mets often present with diabetes insipidus and are often treated with radiosurgery

**Case**

33 year old male presents with right eye sealed shut for 10 days and several months of abdominal pain. Exam – R 3rd nerve palsy, but acuity intact. Hormone function reveals low testosterone, low thyroid hormone.
**Pituitary Lymphoma**

MRI – isointense to brain on T1 and T2, enhance homogeneously after gado (pituitary adenoma is hypointense on T1, hyperintense on T2, and have delayed gado enhancement compared to normal gland)

Treatment – transsphenoidal biopsy then chemotherapy

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**Pituitary Germinoma**

Most common causes of central DI in children/adolescents are Langerhans’ histiocytosis and germinoma.

MRI – thick homogeneously enhancing stalk; sometimes associated with pineal lesion.

Treatment – Transsphenoidal biopsy, then radiation to 40 Gy, 90% long-term survival

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**Case**

24 yo F with intermittent h/a for 6 months, 1 month of amenorrhea, 2 weeks of polyuria/polydipsia. No other sx. Sp grav 1.005, daily urine output 7 L, decreased estradiol, low T3/T4, prolactin 25 ng/mL, low GH, ESR 14, WBC 6.2

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**Pituitary Abscess**

Can be primary (in normal gland) or secondary (in pre-existing pathology like adenoma or Rathke’s cleft cyst).

Presenting symptom – hypopituitarism, headache, visual field cut. Fever and meningismus are rare.

Source of infection unclear – hematogenous versus local spread from sphenoid sinus both speculated
**Pituitary Abscess**

MRI - cystic lesion with heterogeneous signal and peripheral enhancement causing hypopituitarism. Restricted diffusion highly predictive when present.

Only 50% grow out organisms, when they do gram + and gram – equally common, fungi less common.

**Culture Negative May Still Benefit from Antibiotic Treatment**

Culture negative infected Rathke’s cysts recur more frequently than culture positive cysts treated with antibiotics; antibiotic treatment restores the recurrence rate to that of noncultured/noninfected cysts.

**Empty sella syndrome**

Sella filled with CSF causing pituitary gland to be flattened against floor of sella. In these patients, the diaphragm sellae is limited to a thin rim of tissue around a huge infundibular foramen.

Much more common in females than males. Seen in 6% of autopsies. 30% of empty sella patients are hypertensive.

Can be primary or secondary to burned out pituitary tumors after apoplexy, post-radiation.

Headache most common symptom. 30% GH deficient.

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Same case continued

One year later all hormone function except GH has resolved and MRI shows:
Case

28 yo F with severe h/a 4 days after normal vaginal delivery. Labs reveal low prolactin 0.5 ng/mL (normal 0-20) and low ACTH

Another case

39 year old male with 4 weeks of h/a, n/v, and left eye visual loss

To OR at another institution where left carotid injury occurred

Another case (continued)

Transferred to our institution for carotid injury/pseudoaneurysm management (stenting of left supraclinoid ICA)

On coumadin post-stent and on replacement steroid therapy.

3 month follow-up imaging

Lymphocytic Hypophysitis

Lymphocytic pituitary infiltration during or after pregnancy

Symptoms – present with ACTH deficiency (whereas nonfunctional adenoma presents with GH or FSH/LH deficiency), 1/3 get hyperprolactinemia (stalk effect) 2/3 get hypoprolactinemia (gland compression)

MRI – anterior gland enhancement that may extend to stalk

Diagnosed by transsphenoidal endonasal pituitary biopsy

Treated with steroids

3 weeks steroids
**Langerhans’ Histiocytosis**

In pediatric/adolescent patients, most common causes of DI are Langerhans’ histiocytosis and germinoma. MRI – thick stalk, posterior lobe loses its T1 brightness but now enhances. Need biopsy to differentiate from germinoma. Treat with alkylating agent + steroid. Prognosis good.

**Pituitary Sarcoidosis**

5% of sarcoid cases have CNS involvement, but hypothalamus, pituitary stalk, and pituitary are most common sites of CNS involvement. DI is most common presenting symptom, followed by anterior pituitary hormone deficiency. MRI with enhancing fullness of posterior gland. Treatment – transsphenoidal biopsy, treat with 40 mg prednisone daily, lesion will likely resolve, but patient will likely continue to need ddAVP if presented with DI.

**Case**

35 yo F with galactorrhea and amenorrhea. Labs reveal prolactin 43 ng/mL (normal 0-20), TSH 283 µU/mL (normal 0-3).

**Pituitary Hyperplasia**

Pituitary Adenoma - Nonsecretory

Radiographic features: abnormality in anterior lobe of gland usually anteroinferior aspect of gland; tend to be on one side and push stalk to opposite side; most are somewhat T2 bright indicating softness, fibrous adenomas can be T2 dark; characteristic T1 enhancement pattern causing adenoma to appear darker than adjacent gland.

Pituitary Adenoma - Nonsecretory

Stalk effect – increased prolactin due to tumor size, increased adenoma size up to 3 cm makes a higher prolactin attributable to stalk effect.

Pituitary Adenoma - Nonsecretory

Pathologist should stain tumors for hormones, especially ACTH, because “silent corticotrophic” adenomas may have higher risk of recurrence and risk of delayed Cushing’s disease.