Clinical Correlate: Pituitary Pathology

2018-2019 Mechanisms of Human Disease Course
Loyola University Chicago Stritch School of Medicine

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Mid-sagittal Plane

Traditional Open Approaches (Craniotomy)
Minimally Invasive Options

- Eyebrow Craniotomy
- Endonasal Approaches
  - Microscope
  - Endoscope
Minimally Invasive Options

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Mid-sagittal Plane
Endonasal Transphenoidal Approach

Endonasal Skull Base Bony Landmarks

Endonasal Transphenoidal Approach
Endonasal Transphenoidal Approach

Pituitary Gland

- Anterior Lobe (adenohypophysis)
  - True gland (oral mucosa – Rathke’s pouch, ectoderm)
  - Hormones: ACTH, TSH, PRL, GH, FSH, LH
- Posterior Lobe (neurohypophysis)
  - Axons of hypothalamic neurons
  - Hormones: Oxytocin, Vasopressin


Sphenoid Sinus:
The Sun in Our Solar System

Optic Pathways

- ORGANISATION OF OPTIC CHIASMA:
- In general nasal fibres from optic nerve cross in chiasma and temporal fibres remains uncrossed.
- Fibres from ext muscular / inf nasal retina cross and loop anteriorly to C/L optic nerve before they head in to optic tract forming *VON WILLEBRAND'S KNEE*.

INDIVIDUAL CASE PRESENTATIONS

1. Pituitary Macroadenoma
Pituitary Adenoma

- 10-25% of all intracranial tumors
- Prevalence: 17%
- Roughly 1 in 1000 adenomas require surgical treatment
- Three main categories:
  - Benign adenoma
  - Invasive adenoma
  - Pituitary carcinoma (0.1%)

Pituitary Adenomas

Pituitary Adenomas – Hormone Status

- Nonfunctioning/Nonsecretory/Null Cell (60-70%)
- Functioning/Secretory (30-40%)
  - Prolactinoma (70%) (2 - 25 ng/mL)
    - Galactorrhea, amenorrhea
    - Stalk and Hook effect
    - Medically treated with dopamine agonist (bromocriptine, cabergoline, quinagolide)
  - Growth Hormone (20%)
    - Acromegaly (enlargement of extremities, facial features, voice deepening)
    - Hypertension, diabetes mellitus, cardiovascular disease
    - Medically treated with somatostatin (GHIH), Octreotide
  - ACTH (10%)
    - Cushing’s Disease (vs Syndrome)
    - Abdominal striae, moon facies, buffalo hump, acne, hirsutism (women), decreased libido (men)
    - Some studies investigating use of ketoconazole
Treatment Options

• Doing nothing
• Observation with serial imaging (annual MRI)
• Medical Therapy
• Radiation Therapy
• Surgery

Indications for Surgery

• Non-functioning Adenomas
  – Visual/Neural deficits or imaging showing mass effect
  – Radiographic progression

• Functional (Secreting) Adenomas
  – GH, ACTH
  – Prolactinomas that are medically refractory or for patients that cannot tolerate side effects

• Pituitary Apoplexy
• Patient Preference

47 y with vision problems
Serum Endocrine Labs

- Prolactin
- IGF-1
- TSH
- Free T4
- Cortisol
- GH
- LH (female)
- FSH (female)
- Testosterone (male)
- ACTH

Perioperative Considerations

- Endoscopic Endonasal Approach
- Full serum endocrine labs and consultation
- Ophthalmological VF testing
- Stereotactic navigation CT and MRI

- Review our cases at monthly multidisciplinary skull base conference (4th Monday at 4:30pm)

- Patient is consented for lumbar drain (<1%), abdominal fat (3%), and nasoseptal flap to help reconstruct the skull base at the end of surgery
Histology:
Monomorphic cells, loss of acinar structure

Follow Up

• Vision improved, 4 year follow up
• No recurrence

Perioperative Management

• Ceftriaxone perioperative, 50mg
  Hydrocortisone
• Full set of routine labs upon RR arrival
  and then qam
• POD#1 MR imaging
• POD#2 am cortisol (cut off 10)
• DI parameters (>500 cc/hr x 2hr,
  Usp<1.005, Na>145)
• 1 week po abs after discharge
• Routine Na with endocrine follow up
  within 4 weeks
• Evaluated by ENT 1, 3, and 6 weeks
  after surgery
• Evaluated by NSGY 6 weeks after
  surgery
• Surveillance imaging annually
Resection Techniques

• “2-suction” technique (R1)
• Curettes (R2)
• Ultrasonic aspiration/drills/rongeurs (R3)

Must identify the normal pituitary gland!

Complication: Hypopituitarism

• Most common cause: iatrogenic
  – Other causes: trauma, radiation, apoplexy, sarcoid, stroke
• Symptoms vary, depending on hormonal loss
  – Include fatigue, weight loss, intolerance to cold, anemia, decreased appetite, infertility
• Levothyroxine 100mcg qd
• Hydrocortisone 20mg qam, 10mg qpm
• DDAVP (0.1mg po per day usually bid, 10mcg IN, 1mcg IV)
Complication: Diabetes Insipidus

- Various definitions
  - 300cc urine output per hour over two consecutive hours
  - Uspec grav <1.005
  - Serum Na >145

Rare Condition: Pituitary Apoplexy: 51 yo man presenting with 3 day history of bad headache, poor visual acuity OD, right temporal field deficit

Perioperative Management

- Steroid therapy
- Thyroid replacement?
- Other hormonal deficiency?
- Ophthalmological Consultation
- Otolaryngology Consultation
- Endocrine Consultation
- Urgent surgery
Preservation of the posterior gland has led to no patients on permanent Vasopressin therapy.

2. Rathke’s Cleft Cyst
Rathke’s Cleft Cyst

• Mucin containing cyst
• Single columnar/cuboidal ciliated cellular lining (histology)
• Imaging is variable; usually proteinaceous contents and appear hyperintense on MRI Flair imaging
• Surgical goal is fenestration

Follow Up

• Vision improved, 4 year follow up
• No recurrence
3. Infundibular granular cell tumor

33 yo with vision problems
Exam revealed bitemporal field cut

Infundibular (Stalk) Mass
Normal Anatomy
Follow Up

• Vision improved, 4 year follow up
• Hormone Replacement for panhypopituitarism
• No recurrence

4. Craniopharyngioma

46 yo with vision problems and increased urinary frequency
**Craniopharyngioma**

- Bimodal distribution (children 5-14 years and older adults 50-74 years)
- Develops from odontogenic nests of epithelium from Rathke’s pouch (precursor to anterior gland)
- Benign
- Typically grows along stalk

Different types have distinct mutational and transcriptomic profiles

- **Adamantinomatous**
  - Wet keratin
  - Calcification
  - Stellate reticulum
  - Palisading epithelium
  - CTNNB1 mutation

- **Papillary**
  - Better prognosis?
  - Almost exclusively in adults
  - BRAF mutation
  - Medical options?
Follow Up

• Hormone Replacement
• Vision Improved
• 5 years, no recurrence

5. Meningioma

Tubercul Meningioma

preoperative

• 39 year old woman with headaches
• Tubercular mass noted
• Followed with imaging
• Grew over 2 years
• Offered craniotomy at outside institution
Follow up

- Neurologically Intact
- 3 years, no recurrence

Planning for March 2020…
6th Annual Chicagoland Endoscopic Endonasal Skull Base Surgery Course at Loyola University Stritch School of Medicine

Thank you