Benign Brain Tumors

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# Overview

- Pituitary Adenoma
- Craniopharyngioma
- Vestibular Schwannoma

- Overview of Anatomy
- Presentation, Diagnosis and Treatment of each
Pituitary Anatomy 101

Patient Profile View (i.e. Sagittal View)
- Showing pituitary gland above the sphenoid sinus

- Adjacent to the optic nerve and Chiasm (crossing of the two optic nerves)

Cross Sectional View (i.e. Coronal View)
- Nearby structures include multiple cranial nerves, and important vessels like carotid artery
Two Parts to the Pituitary Gland

**Anterior Pituitary**
- Adenohypophysis
- Produces and secretes multiple trophic/growth hormones

**Posterior Pituitary**
- Neurohypophysis
- Secretes two hormones (Oxytocin and ADH/Vasopressin) synthesized by the hypothalamus
Function of the Anterior Pituitary

Cell bodies secrete releasing and inhibiting hormones

ANTERIOR LOBE OF PITUITARY

- STH (GH) → growth-promoting effects on most cells
- ACTH
- TSH
- FSH
- LH
- PRL

Produces and secretes its own hormones

- adrenal cortex
- thyroid
- gonads
- mammary glands
Function of Posterior Pituitary

Two Hormones Secreted:

1) ADH/Vasopressin
   - Stimulates Water Retention
   - Raises blood pressure

2) Oxytocin:
   - Uterine Contractions
   - Lactation

Hormones secreted by Hypothalamus
Pituitary Adenoma

- Noncancerous Tumors that occur in the pituitary gland
  - Benign: majority
  - Invasive: 30%
  - Carcinoma: 0.1-0.2%
- If greater than 10 mm = Macroadenoma
- If less than 10 mm = Microadenoma
- Many are incidentally found
Pituitary Adenoma: Presenting Symptoms

- Symptoms from compression as well as from hormone change
- Headache
- Vision Changes
  -- Double Vision
  -- Vision Loss (especially peripheral vision)
- Hormone Abnormalities
  -- over or under secretion
  -- thyroid disease
  -- hypogonadism
  -- acromegaly
Evaluating Pituitary Adenoma

**Imaging:**
- most commonly with MRI
- Contrast Enhancing?
- Size?
- If CT completed: is there calcification?

**Labs:**
- Evaluating hormone function
Pituitary Lesion: Differential Diagnosis

- Pituitary Adenoma
- Pituitary Hyperplasia: secondary overgrowth from other physiologic situation (pregnancy, other endocrine disorder)
- Craniopharyngioma: tumors originating from Rathke’s pouch
- Meningioma: arise from meninges or brain covering
- Pituicytoma: rare benign tumor from posterior pituitary gland
- Hypophysitis: lymphocytic inflammation
Pituitary Adenoma: Hormone Abnormalities

• Many are non-hormone secreting, however when present are most commonly (in order):
  – Prolactinoma (“Lactotrophic”)
  – Growth Hormone Secreting (“Somatotrophic”)
  – ACTH secreting (“Corticotrophic”)

• Hormone Deficiencies Can also arise
  -Gonadotropic Deficiency most common and will result in hypogonadism
  -Diabetes Insipidus indicates a pituitary stalk lesion and undersecretion of ADH
Symptoms in Prolactinoma

- **In Women:**
  - anovulation, cessation of menstruation
  - galactorrhea
  - lowered libido
  - infertility

- **In Men**
  - lowered libido
  - erectile dysfunction
  - galactorrhea and infertility is rare
Growth Hormone Secreting Tumors

Acromegaly = syndrome of excess GH secretion

- skull expansion (frontal)
- soft tissue swelling with thickening of skin in hands, feet, nose, lips and ears
- brow protrusion
- lower jaw protrusion
- hyperpigmentation
ACTH secreting tumors

- ACTH over secretion leads to downstream over secretion of cortisol
- Cortisol is a steroid hormone produced in the adrenal gland
- Normally released to increase blood sugar, decrease inflammatory response
- “Stress Hormone”
Cushing’s Syndrome

- Inappropriately high levels of cortisol secretion
- Weight gain
- High blood pressure
- High blood sugars/insulin resistance
- Depression
- Skin changes
- Osteoporosis
Symptoms of Hypogonadism

- Low levels of sex hormones: testosterone (androgens) and estrogen and progesterone
- Impaired Spermatogenesis and Ovulation
- **In Women:**
  - anovulation, cessation of menstruation
  - lowered libido
  - loss of body hair and hot flashes
- **In Men**
  - lowered libido
  - loss of body hair, loss of muscle mass
  - gynecomastia
Symptoms of Diabetes Insipidus

- From a decrease in vasopressin aka Antidiuretic Hormone (ADH)
- Excessive Urination
- Excessive thirst
- Dehydration
- Loss of Potassium
Hormonal Evaluation

- Serum Prolactin
- Insulin Like Growth Factor (IGF-1) surrogate for measuring GH activity
- 24 hour Urine Cortisol with high ACTH level
- TSH/T4/T3 and assessment for thyroid goiter
Management of Pituitary Adenoma

If 10 mm or less

“Incidentaloma”

2-5 mm
No follow up indicated

5-9 mm
Check prolactin
Serial MRI scans

With Hormone Abnormalities

Medical Treatment
Management of Pituitary Adenoma

If more than 10

- With hormonal abnormalities:
  - Surgery
  - Hormonal therapy

- Non-functioning or gonadotroph causing vision symptoms:
  - Transphenoidal surgery
Medical Management of Hormone Secreting Pituitary Adenomas

- Generally indicated when large sized tumor cause local symptoms OR if hormone abnormalities are causing systemic symptoms such as:
  1) Galactorrhea or Gonadotropin Deficiencies
  2) Acromegaly
  3) Cushing’s Disease
Treatment of Hormonal Abnormalities

**Treatment of Hyperprolactinemia**
- generally serum prolactin >200 ng/mL
- treatment with dopamine agonists (cabergoline, bromocriptine, pergolide)

**Treatment of Acromegaly**
- high serum IGF-1
- Somatostatin analogues: octreotide and lanreotide

**Treatment of Cushing’s Disease/Corticotrophic Tumor**
- Dopamine agonist: cabergoline
- Somatostatin analogue: pasireotide
- Glucocorticoid antagonist: mifepristone
Transsphenoidal Surgical Approach

- Either sublabial or often endonasal approach approach

- Usually with endoscope (sometimes microscope)

- Reaches the pituitary gland via the sphenoid sinus

- Complications include bleeding, infection, or hormone insufficiencies
Radiation in Pituitary Tumors

- Used predominantly for tumors that are not amenable to medical treatment

- Types of Radiation used:
  - External Beam/Intensity Modulated Radiotherapy
  - Stereotactic Radiosurgery (gamma knife, cyberknife)
  - Proton Beam therapy
Prognosis and Follow Up

- Serial MRI scans following treatment (regardless of modality)
- Nonfunctioning tumors are not likely to experience growth if less than 10 mm
- For tumors greater than 10 mm, 4 four likely to grow in follow up
- Post surgically, tumor progression is approximately 10-19% in 6-10 year follow up (variable statistics)
<table>
<thead>
<tr>
<th>Craniopharyngioma</th>
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<td>• Solid/mixed solid-cystic tumors arising from Rathke's pouch remnants anywhere along a line from the nasopharynx to the diencephalon (thalamus/hypothalamus)</td>
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<td>• Typically slow growing, benign tumors that grow from pituitary stalk</td>
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<td>• Bimodal age peaks</td>
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<td>-children between 5 and 14 years (adenomatous)</td>
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<td>-adults between 50 and 75 years of age (papillary)</td>
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Clinical Presentation

**Visual Symptoms**
- Classically “bitemporal hemianopia” (peripheral field vision loss)
- Unilateral vision loss

![Visual Field Defects Diagram](image)
Clinical Presentation: Continued

**Endocrine Abnormalities**
- In kids: classically growth failure
- decreased GH, decreased LH/FSH/ACTH or hypothyroidism

**Signs of Increased Pressure**
- Headache
- Nausea/Vomiting
Craniopharyngioma: Imaging

• On CT, often calcified lesion

• On MRI, contrast enhancing
Craniopharyngioma Treatment: Surgery

- In most cases, Surgery is first step
- Surgery established diagnosis, alleviates mass related symptoms and cyst drainage if indicated
- Most commonly, transsphenoidal approach, others include pterional craniotomy
Craniopharyngioma Treatment: Radiation

- May be indicated in patients without total resection or after recurrence

1) Stereotactic Radiosurgery
   - single fraction of radiation

2) Intensity Modulated Radiation Therapy
   - optimizes the delivery of radiation to irregularly shaped volumes

3) Proton Beam Radiation Therapy
   - heavier particle to limit scatter to normal surrounding structures
Acoustic Neuroma

- Many synonymous terms
  - Acoustic Schwannoma
  - Vestibular Schwannoma
- 1 in 100,000
- Median age is 50 years old
- Unilateral in 90%
- If bilateral primarily due to Neurofibromatosis Type 2
Acoustic Neuroma: Anatomy

Nearby Structures:
- Vestibular Nerve
- Cochlear Nerve
- Trigeminal Nerve
- Facial Nerve
- Cerebellum
- Brainstem
### Presenting Symptoms

**Cranial nerve involvement**
- Vestibular Nerve
- Cochlear Nerve
- Trigeminal Nerve
- Facial Nerve

**Cerebellar compression**
- uncoordinated, walking/gait issues

**Brainstem Compression**
- brain swelling, herniation, lower cranial nerve deficits
Acoustic Neuroma: Diagnosis

- MRI imaging: contrast enhancing lesions in the area of the internal auditory canal

- Physical Exam: abnormal hearing and other ipsilateral cranial nerve defenses

- Audiometry: only 5% of patients have normal audiometry testing and hearing loss does not necessarily correlate with tumor size
Differential Diagnosis

- Meningioma (4-10% of cases)
- Lipoma
- Glioma
- Cholesterol Cyst/Cholesteoma
- Hemangioma
- Metastatic Tumor
Acoustic Neuroma Treatment: Surgery

- Three approaches which may result in long term control and establishes diagnosis
1) Retromastoid suboccipital
2) Translabyrinthine - recommended for acoustic tumors larger than 3 cm and for smaller tumors when hearing preservation is not an issue
3) Middle fossa - Suitable for small (<1.5 cm) tumors
Acoustic Neuroma Treatment: Radiation

1) Stereotactic Radiosurgery
2) Proton Beam Irradiation

-both allow local key structures to be spared
-long term follow up data is limited, however studies have shown acceptable control with these modalities
• For older patients and patients small tumors and limited hearing loss, often observation with serial imaging and audiometry is suggested
• Patients should be monitored at least annually for evidence of tumor progression
• Rapid tumor growth (>2.5 mm/year) rather than absolute tumor size may be the most useful indicator for therapy
Neurofibromatosis 2

- Bilateral Vestibular Schwannoma characteristic clinical feature

- Mutation in Neurofibromin/Merlin Gene on chromosome 22

- Often Associated with positive family history (autosomal dominant) but may be sporadic mutation
NF2: Vestibular Schwannoma

- Typically bilateral
- Typically benign in NF2, however given their location can invade brainstem, cause hydrocephalus
- In NF2, poor correlation between lesion size and hearing loss
- Patients also have other types of tumors: meningiomas, retinal hamartomas etc
Thank You! Questions?