Skull Base Pseudotumor and Other Inflammatory Diseases

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Skull Base Non-infectious Inflammatory Disease: Outline

- Idiopathic Inflammatory Pseudotumor (IP)
- IgG4 Related Disease (IgG4 RD)
- Granulomatosis with Polyangiitis (GPA)
- Sarcoidosis

Take home message:

- Diagnosis of skull base IP or other non-infectious, inflammatory disease should be made only after biopsy excludes malignancy & infectious etiologies

Illustrative Case

37 yo F: H/A, Subacute 12th CN, acute 5th & 6th CN palsies

MRI Report DDx: “Tumor, Infection, Inflammatory Pseudotumor & Granulomatosis w/ Polyangiitis”

Subsequent Work-up

- Labs:
  - CBC, C-reactive protein
  - Serum IgG4 levels
  - C-ANCA
  - ACE
- Imaging:
  - MR → CT, CT → MR
  - PET/CT
- Clinical referrals:
  - Neurology/ Neurosurgery
  - ENT
  - Infectious Disease
  - Rheumatology
- Biopsy: (-) for tumor & infection → repeat Bx. (-)
- Inflammatory Pseudotumor is a Dx. of exclusion → steroids

Inflammatory Pseudotumor (IP)

- Idiopathic, non-neoplastic inflammation:
  - Spindle cells, lymphocyte & plasma cell infiltration
  - Variable fibrosis
- Location:
  - Orbit > Cavernous sinus, Skull base/ Nasopharynx
  - Anywhere in the body
- Clinical presentation of H&N disease:
  - Deep, retro-orbital pain/ headache (~ 60%)
  - Cranial nerve palsies
- Treatment:
  - Steroids
  - Surgical resection: Complete or debulking w/ steroids

Disclosures

No Financial Relations with the Medical Industry to Disclosure

No “off label” application of contrast agents or medical devises
**IP - Imaging**

1. Infiltrative lesion, trans-spatial
2. Mass-like dural thickening
3. T2: iso-hypointense (↑ fibrosis); Homogeneous CE

**IP - Imaging**

1. Marrow infiltration
2. CT: Bone remodeling or erosion
3. Involved ICA may be secondarily narrowed

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**IP - Treatment Response**

1. ~ 80% initially imp. w/ steroids
2. Variable recurrence rate 25-60%
3. Steroids x 4 mos.

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2. T1: isointense; T2: iso-hypointense (↑ fibrosis)
3. Marrow replacement

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**IP - Imaging**

1. Active disease is FDG avid
2. PET-CT may be useful for following Tx. response
3. Image guided core bx. often followed by open biopsy
4. No FNA
IgG4 – Related Disease

- Well defined, solid ST lesions
- Homogeneous CE; T2-Hypointense
- Lesions often extend along 5th CN

IgG4 Related Disease

- Infiltration of IgG (+) plasma cells & lymphocytes → fibrosis & organ dysfunction
- Elevated serum IgG4 levels
- Location:
  - Orbits
  - Lacrimal/ Salivary glands (Dacroadenitis, Mikulicz Dz.)
  - Pituitary/ Cavernous sinus/ Skull base (Autoimmune Hypophysitis)
- Imaging:
  - Solid, well-defined ST lesion
  - T2 Hypointense; Homogeneous CE
  - Pachymeningeal CE/ thickening: Diffuse or localized
  - Perineural spread; V2 > V1; mimics PNT
  - Dramatic improvement w/ steroids

Transsphenoidal Pit Bx.:

↑ IgG4 plasma cells

Complete Sx. resolution following Steroids Tx.

Granulomatosis w/ Polyangiitis (GPA)

- GPA involves H&N in ~ 85% of cases
- Skull base GPA:
  - Usually by spread from sinonasal lesion
  - May occur in isolation
- CT: Moderately CE ST thickening; +/- bone erosion

Granulomatosis w/ Polyangiitis (GPA)

- Secondary CN palsies (~ 6%) predominantly unilateral:
  - CN 1 & 2 most common, but all CN venerable
- MRI:
  - CE infiltrative lesion > discrete mass
  - T1- is to hypointense; T2 - variable

22 yo F: Lt ear pain unresponsive to antibiotics & steroids

Lt. 5, 7, 9, 10 & 12 CN neuropathies noted post-mastoidectomy
**Granulomatosis w/ Polyangiitis (GPA)**

- MRI:
  - CE infiltrative lesion > discrete mass
  - Pachymeningeal thickening/ CE
  - T1- iso to hypointense; T2 - variable
- GPA of NP/ skull base may occur in isolation (“Limited GPA”)

**H&N Bx.**: Only 25-57% (+) for vasculitis & necrotizing granulomas
- (-) C-ANCA does not exclude GPA:
  - Generalized active dz. sensitivity ~ 95%
  - Limited active dz. sensitivity as low as 60%
- Tx: Prednisone & Cyclophosphamide = 95% remission rate
- H&N involvement assoc. w/ imp. survival but ↑ recurrence rate

**Neurosarcoidosis**

- Cranial neuropathies (58%): 2, 5, 7 & 8
- Dural thickening or nodular lesion(s): 17%
  - Avid, homogeneous CE
  - T1: hypo-isointense; T2: variable

**Diagnosis:**
- Elevated ACE: serum 24-76%; CSF < 50%
- Biopsy: non-caseating granulomas & excludes infection
- Treatment: Steroids +/- immunomodulating drugs

**Skull Base Masses- DDx:**

- SCC
- ACC
- Lymphoma
- Osteomyelitis
Skull Base Non-infectious Inflammatory Disease: Conclusions

1. SB lesion: Infiltrative, homogeneous CE & T2 hypointense. Dural thickening/CE → consider Non-infectious Inflam. dz.

2. KSS ("Keep it simple stupid") – Dr. Charles Kerber:
   - Describe lesion & extent (intracranial, perineural)
   - Include a broad Dx. @ initial imaging: "Tumor, Infection, Non-infectious Inflam. dz. (IP, IgG4 RD, GPA, Sarcoid)"

3. Biopsy is required:
   - Tumor & infection must be excluded before treating as a non-infectious inflammatory disease.
   - Endoscopic or CT guided Bx. w/ cutting needle. NO FNA

4. Follow-up is necessary (imaging & laboratory):
   - Lymphoma & Plasmacytoma may improve w/ immunosuppressive TX.
   - Incomplete response → Re-biopsy

Suggested Reading: