Imaging in Neurocysticercosis

Sarun Charumilind, Harvard Medical School, Year III
Gillian Lieberman, MD

September 2004
Index Patient
Index Patient: Mr. CP

- Mr. CP is a previously healthy 25 y.o. man who immigrated from Cape Verde in 1995 and who presents to an outside hospital with:
  - 5-day history of progressively severe headaches and h/o chronic HA x4 yrs, relieved with change in position
  - 2 days of “forgetfulness”
  - Appearance of agitation and nervousness
- No significant PMH, allergies, meds.
- No c/o of N/V, incontinence, visual, speech, personality changes.
- Given Demerol and Phenergan.
- CT and MRI with gadolinium are ordered.
Index Pt.: Imaging at Outside Hospital

- Hydrocephalus
- Third ventricular lesion - ?cyst
- Transferred to BIDMC
- Opening pressure > 30
- L. frontal ventriculostomy and drain placement
- Repeat CT/MRI

Images from PACS, BIDMC
Index Pt: BIDMC Imaging: CT & MRI w/ Gad

- Mild, diffusely enlarged ventricles esp. at third ventricule and temporal horns
- Effaced sulci
- Scattered calcifications along sulci (R pons, R post. frontal lobe)
- Third ventricular cyst with enhancement

Images from PACS, BIDMC
Differential Diagnosis

Obstructive Hydrocephalus
- Tumor: benign or malignant
- Cyst (colloid, arachnoid, PARASITIC)
- Abscess
- Congenital
- Multiple sclerosis
- Tuberous sclerosis

Cyst With Mural Nodule
- Tumor: benign or malignant (ganglioglioma, GBM, astrocytoma)
- Metastasis
- PARASITIC cyst (cysticercus, paragonimus)

Multiple Intracranial Calcifications
- Atherosclerosis
- Idiopathic
- Physiologic (dura, choroid plexi, pineal gland)
- PARASITIC (cysticercosis, paragonimus)

Reeder MM and B Felson, Gamuts in Radiology, 2003.
Hospital Course & Follow-up

- **SURG:** Right transcallosal cyst resection – brown, tan colored cyst removed
- **IMAGING:** resolved hydrocephalus
- **W/U:**
  - ID consult: PPD neg., no growth in serum or CSF, no AFB, positive for *CC Abs*
  - Optho consult: no intraocular Sx
  - Pathology: confirmed NCC
- **TX:** Treated with albendazole x 7d + decadron
- **DISCHARGE DX:** **Neurocysticercosis**
- **F/U:** headaches resolved, discuss PPX if return to Cape Verde

Images from PACS, BIDMC

Index Pt: Resolution of hydrocephalus (CT)
Taenia Solium: Parasite of the Poor

- Most common helminthic infection of CNS
- Based on lifecycle, is facilitated only in poor living conditions
- Endemic to many regions: Mexico, Central and South America, sub-Saharan Africa, Asia
- Higher prevalence in rural areas (10-25%)
- High prevalence in high-pork-consuming/-raising countries
- 50,000 deaths/yr, 20 million infected
- Most common cause of acquired epilepsy worldwide
- United States: ~1000 cases per year

Taenia Solium: Lifecycle

1. Eggs or gravid proglottids in feces and passed into environment
2. Embryonated eggs and/or gravid proglottids ingested by pigs or humans
3. Oncospheres hatch, penetrate intestinal wall, and circulate to musculature in pigs or humans
4. Humans acquire the infection by ingesting raw or undercooked meat from infected animal host
5. Scolex attaches to intestine
6. Adults in small intestine

Oncospheres develop into cysticerci in muscles of pigs or humans

Cysticercosis

Cysticerci may develop in any organ, being more common in subcutaneous tissues as well as in the brain and eyes.

http://www.ucm.es/info/parasito/Taenia%20escolex.jpg
http://www.dpd.cdc.gov/dpdx/HTML/Cysticercosis.htm
Cysticercosis: Clinical Manifestations

Symptomatic Cysticercosis

Neurocysticercosis
- Extraparenchymal
  - Intraventricular
  - Subarachnoid
  - Spinal
- Parenchymal

Extraneural Cysticercosis
- Eye
- Muscle
- Subcutaneous

Pt. #1: Intraocular*
Pt. #2: Muscular (tongue)*
Pt. #3: Parenchymal**
Pt. #4: Subcutaneous**
Pt. #5: Intraocular*

Images from *http://www.facmed.unam.mx/grid/photos.htm and **http://faculty.uaeu.ac.ae/~youssefa/homepage/taenia.htm
Neurocysticercosis

20% Symptomatic
- Mass effects
- Inflammatory response
- Foraminal obstruction
- Ventricular obstruction

80% Asymptomatic

Degree of Sx depends on:
- Lifecycle stage
- Site
- Number of cysts

Tx depends on Sx
1. Meds: antiparasitic, antiinflammatory, anticonvulsant
2. Surgery
3. Nothing!

Life Stages of T. Solium

Cysticerci
- Asymptomatic
- Host immune tolerance
- Duration: 3-5 yrs

Degenerating
- Inflammatory response
- Cyst loses ability to moderate immune response
- Active NCC

Inactive
- Calcified
- Asymptomatic
- Potential epileptic/obstructive focus
Stage 1: Vesicular Cysts

**CT:** Small, rounded, well-demarcated, low-density w/o edema.

**MRI:** Signal similar to CSF in T1 and T2; scolex seen in cyst: “hole-with-dot” or “swiss cheese” if severe.

Images from Garica HH and Del OH Bruto, 2003.
Stage 2: Colloidal Cysts

- **CT**: ill-defined with edema, ring pattern with contrast = acute encephalitic phase

- **MRI**: thick hypointense wall + perilesional hyperintense edema

- **Cysticercotic Encephalitis**: diffuse edema + collapsed ventricles + multiple lesions = young cysts attacked by host

Images from Garcia HH and Del OH Bruto, 2003.
Stage 3: Granular Cysts & Stage 4: Calcified Cysts

**Granular**

- **CT**: granulomatous
  - Pt. #11: “Single enhancing lesion” on CT

- **MRI**: signal void + hyper-intense edematous rim

**Calcified**

- **CT**: small, hyperdense nodules w/o edema unless relapse
  - Pt. #12: MRI: Signal void w/ surrounding edema

- **MRI**: edema with high signal surrounding low signal cyst

Images from Garcia HH and Del OH Bruto, 2003.
Other Cyst Locations

- **Subarachnoid**: large, multi-lobulated

- **Ventricular**: best on MRI, isodense on CT

- **Spinal**: nonspecific enlargement, filling defects on myelogram

Images from García HH and Del OH Bruto, 2003.
Comparing Modalities

- **Plain film**: Rarely used (incidental subQ and muscle Ca++)
- **CT v. MRI**: Depends on stage
- **CT**: Cheaper, better at Ca++
- **MRI**:
  - Better sensitivity (esp. active cysts, cysts in post. fossa)
  - Better scolex visualization
  - Not widely available globally
- **Approach**: CT first, then MRI if inconclusive, strong suspicion, or follow-up.

Index Pt: Close-up views of mural nodule cyst: cysticercus with scolex inside (MRI)

Images from PACS, BIDMC
Other Diagnostic Workup

- **Serology**
  - **Targets**: AntiCC antibodies or cysticercal antigens
  - **Substrates**: CSF, saliva, or blood
  - **Methods**: ELISA, complement fixation, immunoblot, etc.
  - **Standard**: Enzyme-linked immunoblot, detect antiCC Abs

- **Pathology**
  - White fluid-filled bladder 5-10 mm diameter w/ solid 2 mm larval tapeworm

Pt. #15: *Taenia solium* egg

Pt. #16: Electron microscopy of an egg

[Links to images and additional resources]
Diagnostic Criteria

1. **Absolute:**
   - Histologic
   - Scolex on imaging
   - Ocular signs

2. **Major:**
   - Lesions “suggestive” on imaging
   - Positive serology for Abs
   - Improvement after treatment
   - Spontaneous resolution of single lesion

3. **Minor:**
   - Lesions “compatible” on imaging
   - Clinical signs “suggestive”
   - Positive serology for Abs or Ags
   - CC outside CNS

4. **Epidemiologic:**
   - Household contact
   - Geographic correlation
   - Travel correlation

**Diagnosis:**

- **Definitive:**
  - 1 absolute + 1 epi
  - 2 major + 1 minor + 1 epi

- **Probably:**
  - 1 major + 2 minor
  - 1 major + 1 minor + 1 epidemiologic
  - 3 minor + 1 epidemiologic
The International Debate over Criteria

In the setting of many countries with endemic NCC...

- **Biopsy**: not possible to perform in all suspected pts.
- **Ocular**: direct visualization rare, hardly reported
- **Scolex**: uncommon on CT/MRI
- **Epi**: majority of Indians have single enhancing, yet this is only a “major” criteria
- **Different DDX**: CNS tuberculosis, with also single/multiple tuberculoma that show as multiple enhancing CT/MRI lesions, similar to NCC; also multiple tuberculoma, fungal granuloma, primary or secondary malignancies, and multiple pyogenic abscesses
- **Serology**: EITB (Ab immunoblot) not available, not sensitive
- **Tx**: albendazole/praziquantel may not be effective, and tx role is unclear
- “Minor” and “Epi” Criteria: not specific enough?

Current criteria more suitable for the patients in regions where both NCC and other CNS infections are uncommon, as opposed to India or other developing countries, which have high prevalence of these.
A Patient with MVA 2° to Seizure

- **Mr. EM**: previously healthy 33 yo man from Cape Verde, struck another vehicle at 80 mph.
- Seized at scene of MVA and outside the ER.
- **Imaging**: Trauma, Spine, Head series: No fractures
- **NCHCT**: 8 small calcifications without edema
- **TX**: Seizure control. No surgery.

Images from PACS, BIDMC

- Pt.#17: Calcification near cranial vertex (CT)
- Pt.#17: T2 MR of same lesion
- Pt.#17: MR, susceptibility of same lesion
- Pt.#17: Calcification in posterior fossa (CT)
A Patient with MVA 2° to Seizure (cont.)

**Remember!**

In developing regions of the world, seizure is a common first presentation of NCC. NCC is the most common cause of adult-onset acquired epilepsy worldwide.
A Patient with Incidental NCC?

- **Mr. RB**: 38 yo Nepali man with seizure and head trauma
- **NCHCT**: Ventricular and parenchymal calcifications
- **DDX**: EtOH withdrawal v. NCC

Pt #18: Multiple parenchymal and ventricular calcifications (noncontrast CT)
A Patient with Incidental NCC?
(cont.)

Remember!
Though calcifications may cause obstruction or serve as seizure foci, more often than not, they cause no pathology. 80% of persons with NCC are asymptomatic.
A Patient with Arachnoiditis 2° to NCC

- **Mr. MB**: 52 yo Cape Verdean man with previous Dx of NCC with 4th ventricular cyst removal
- **Presents**: severe HA, fever, and LLE radicular pain, neck stiffness, blurry vision
- **Imaging**: Obstructive hydrocephalus + spinal cord inflammation
- **TX**: Anti-TB for meningitis without success (misdiagnosis of TB)

Images from PACS, BIDMC

Pt.#19: T1-T2 enhancement (T1 MR)
Pt.#19: Cervicomedullary enhancement (T1 MR)
Arachnoiditis 2° to NCC (cont.)

- **Arachnoiditis**: spinal cord inflammation evidenced by nerve root clumping and adhesions to thecal sac
- **DX**: Hydrocephalus 2° to impaired CSF absorption from CSF inflammatory process $\rightarrow$ Neurocysticercosis

Pt.#19: Lumbosacral involvement: clumping and adhesions (T1 MR, post gad)

Images from PACS, BIDMC
Remember!

NCC may have spinal manifestations. Despite this patient's h/o cyst removal, other cysts may have been present in various life stages. Albendazole is only 75-90% effective.* Up to 41% of patients post-treatment may exhibit noninflamed but viable cysts.**

A Patient with Toxoplasmosis v. NCC

- **Mr. HD**: 33 yo Haitian man who on his 1-year anniversary of being diagnosed with HIV/AIDS presents with severe headaches, photophobia, blurred vision, dizziness, NVx1, neck pain on movement, fevers, chills, sweats

- **Imaging**: Multiple enhancing lesions with surrounding edema

- **DX**: DISSEMINATED CEREBRAL TOXOPLASMOSIS

Images from PACS, BIDMC
A Patient with Toxoplasmosis v. NCC (cont.)

Remember!

The differential for multiple intracranial calcifications is wide. In addition, settings where NCC is highly prevalent are also settings for other infectious pathologies of the brain: toxoplasmosis, lymphoma in HIV patients, TB granulomas, etc.
Conclusion

- Imaging findings depend on modality, cysticercus stage, and location.
- Both CT and MRI have a role, but CT is the best screening tool.
- Debatable whether diagnostic criteria work for all global settings (differences in prevalence and technology).
- Diagnosis often requires serologic, clinical, pathologic, and epidemiologic correlation.
References

Acknowledgements

- Joe Barry, MD
- Steve Reddy, MD
- Barbara Appignani, MD
- Andrew Tarulli, MD
- David Hackney, MD
- Gillian Lieberman, MD
- Pamela Lepkowski
- Larry Barbaras, Webmaster