Adult Congenital Hydrocephalus

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Objectives

- Discuss normal CSF flow mechanics
- Discuss normal radiographic findings
- Discuss epidemiology of hydrocephalus
- Discuss briefly the etiologies of hydrocephalus
- Discuss pathology and pathophysiology associated with hydrocephalus with emphasis on adult clinical presentation
- Discuss clinical case and radiographic findings
Introduction

“Water on the Brain”

- Abnormal accumulation of CSF in the cranial vault
- CSF generally produced at an average rate of \( \sim 0.3 \text{mL/min} \). To maintain steady state, rate of CSF production must necessarily equal rate of CSF outflow
Two general causes:
- Obstruction of outflow of CSF (common)
- Overproduction of CSF (usually due to choroid plexus papilloma – rare - less than 0.5% of cases)

Two categories of obstructive hydrocephalus
- Communicating
- Noncommunicating

Etiologies can be congenital or acquired
Communicating hydrocephalus – results when the obstruction to outflow is distal to the foramina of Magendie and Luschka

- All ventricles are enlarged
- Obstruction is usually at the level of the arachnoid granulations
- Causes can include intraventricular hemorrhage, meningeal scarring, among others.

Noncommunicating hydrocephalus – results when the obstruction to outflow is proximal to the fourth ventricle foramina

- Ventricle(s) proximal to the obstruction are enlarged
- Ventricle(s) distal to the obstruction are generally normal in size
- Obstruction can be congenital or acquired
Ventricular System

Left lateral phantom view

Frontal (anterior) horn
Central part
Temporal (inferior) horn
Occipital (posterior) horn

Cerebral aqueduct (Sylvius)
4th ventricle
Left lateral aperture (foramen of Luschka)
Left lateral recess
Median aperture (foramen of Magendie)

Central canal of spinal cord

Left interventricular foramen (Monro)
3rd ventricle
Supraoptic recess
Infundibular recess
Pineal recess
Suprapineal recess

CSF Circulation Pathways

Normal MRI Findings in the Brain

Source: http://www.cid.ch/
Normal MRI Findings in the Brain

Sagittal T1-weighted image

Source: http://www.cid.ch/
Normal MRI Findings in the Brain

Source: http://www.cid.ch/
Normal MRI Findings in the Brain

T2-weighted axial image

Source: http://www.cid.ch/
Normal MRI Findings in the Brain

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T2-weighted axial image

Source: http://www.cid.ch/
Normal MRI Findings in the Brain

T2-weighted coronal image

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Normal MRI Findings in the Brain

T2-weighted coronal image

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Normal MRI Findings in the Brain

T2-weighted coronal image

Source: http://www.cid.ch/
Normal MRI Findings in the Brain

T2-weighted coronal image

Source: http://www.cid.ch/
Hydrocephalus
Epidemiology

- Cannot be accurately calculated
- In US there are >125,000 patients with CSF shunts and 50,000 shunt operations performed annually
- The accepted rate of incidence of hydrocephalus is 3 to 4 per 1,000 births; however, even this is considered to be underreporting the actual rate.
- Adult hydrocephalus accounts for greater than 50% of the total diagnoses of hydrocephalus.
- Actual numbers of adults who suffer from congenital hydrocephalus is unknown.
Hydrocephalus Etiology

- Maternal malnutrition (e.g., folic acid deficiency leading to neural tube defects).

- Infectious causes:
  - Bacterial – in rare cases can lead to leptomeningeal scarring and permanent fibrosis of CSF absorptive surfaces:
    - *E. coli*
    - *H. influenzae*
    - *S. pneumoniae*
    - *S. agalactiae*
Hydrocephalus Etiology

- Viral – can lead to aqueductal atresia or stenosis
  - CMV
  - Mumps
  - Varicella
  - Rubella
Hydrocephalus Etiology

- Trauma
- Neoplasms or cysts
- Intraventricular hemorrhage
- Subarachnoid hemorrhage
- Congenital
  - Arnold-Chiari malformations
  - Dandy-Walker malformations
  - Spina bifida (myeloceles and meningomyeloceles)
Hydrocephalus Etiology

- Congenital aqueductal stenosis
  - Idiopathic
  - Congenital malformation
    - Arnold-Chiari I & II
    - Dandy-Walker
    - Klippel-Feil syndrome
    - Agenesis of foramen of Monro
  - X-linked recessive or neurofibromatosis mutation
  - Periaqueductal tumor
  - Abnormal blood vessel
  - Arachnoid cyst
  - Secondary membranous occlusion
Pathophysiologic Findings

- Dilated ventricles
- Periventricular gliosis
- Thinning of corpus callosum and atrophy of the periventricular white matter – hemispheric disconnection can result
- Severe hydrocephalus can cause gross thinning of the cortex
- Basal ganglia atrophy has also been reported in several cases with associated resultant motor pathology
Mechanisms of Injury

- Physical/mechanical distortion and parenchymal injury – compression

- Altered extracellular environment – which can lead to altered neuronal function
  - Impaired diffusion
  - Areas of stagnation
  - Improper accumulation and clearance of potentially toxic metabolites, neurotransmitters, and other substances.
  - Blood brain barrier is mildly altered
Mechanisms of Injury

- **Vascular – postulated mechanisms**
  - Doppler blood flow studies, SPECT, MRI and CT have all been used to show decrements in blood flow in white matter – ischemic changes (various animal models have also shown this)
  - Changes in cerebral blood flow and oxidative metabolism
  - Consequence of diminished blood flow is injury to oligodendrocytes and axons in the white matter
  - Mechanisms and morphological characteristics of axonal damage in rats have similarities to those detected after ischemic insult
  - HTN and atherosclerosis may aggravate situation in adult humans
Adult Onset Hydrocephalus

- Includes hydrocephalus caused by
  - Tumor, hemorrhage, trauma, infection, or other brain pathology
  - Congenital hydrocephalus
  - Primary hydrocephalus occurring in older adults such as that seen with idiopathic normal pressure hydrocephalus (NPH)
Adult-onset Hydrocephalus

Almost always caused by a CSF outflow tract obstruction

Etiology of adult onset hydrocephalus. Shown here are percent distribution of 468 cases tried at the Cleveland Clinic between 1994 and 2000. Sx = surgery; ICH = intracranial hemorrhage; NPH = normal pressure hydrocephalus; SAH = subarachnoid hemorrhage.

Table 1. Katzman’s Table of the Etiology of Hydrocephalus in 914 Adult Patients

<table>
<thead>
<tr>
<th>Cause</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Subarachnoid hemorrhage</td>
<td>315</td>
</tr>
<tr>
<td>Head injury</td>
<td>102</td>
</tr>
<tr>
<td>Aqueductal stenosis</td>
<td>34</td>
</tr>
<tr>
<td>Meningitis</td>
<td>34</td>
</tr>
<tr>
<td>After craniotomy</td>
<td>43</td>
</tr>
<tr>
<td>Obstructive lesions in third ventricle</td>
<td>26</td>
</tr>
<tr>
<td>Tumors in third ventricle</td>
<td>21</td>
</tr>
<tr>
<td>Tumors in other locations</td>
<td>11</td>
</tr>
<tr>
<td>Basilar impression</td>
<td>4</td>
</tr>
<tr>
<td>Syringomyelia</td>
<td>1</td>
</tr>
<tr>
<td>Encephalopathy/encephalitis</td>
<td>2</td>
</tr>
<tr>
<td>Idiopathic*</td>
<td>31</td>
</tr>
</tbody>
</table>

Adult-onset Hydrocephalus

- Little understanding of the adaptive brain response to hydrocephalus. It may be that this adaptive response is what makes the brain tolerant of a slowly evolving ventriculomegaly when similar ventriculomegaly is devastating, and often fatal, when it is more acute.

- Adaptive responses may include:
  - Hydrodynamic responses
  - Brain compliance changes
  - Vascular adaptations
  - Etc?

- While the exact process is unknown, the above might explain why patients with severe ventriculomegaly (congenital or otherwise) may do well for decades and then present in adulthood with symptoms of hydrocephaly.
Clinical Presentation – Adult Onset

- May be acute, subacute, or chronic and insidious
- Acute and subacute forms present with symptoms of increased ICP:
  - Acute – stupor and coma – most often seen with SAH, exudative meningitis, meningeal neoplastic infiltration and fourth ventricle tumors.
  - Subacute – develops over a few days or weeks and causes progressive drowsiness or abulia with incontinence

- Symptoms of a more gradually evolving case include (patient needn’t have all symptoms)
  - Headache
  - Nausea (position independent)
  - Vomiting
  - Ataxia
  - Visual disturbances
Clinical Presentation cont.

- Symptoms often evolve over years in a patient with aqueductal stenosis. Symptoms include:
  - Ataxia (generally truncal)
  - Slowed mentation
  - Seizures
  - Urinary incontinence

- Symptoms resulting from aqueductal stenosis in adults are somewhat dependent on age and the degree of ventriculomegaly. Fukuhara et al. reviewed features of late onset idiopathic stenosis and concluded:
  - Younger adults present with symptoms of increased ICP such as headaches and nausea
  - Older adults with larger ventricles present with symptoms similar to NPH
 Hydrocephalus evolves after SAH, meningitis, and severe trauma frequently enough that it should be suspected with any delayed deterioration over weeks or months after the original insult. Symptoms of congenital hydrocephalus can evolve insidiously over years (as in the case of this patient)

- MR is the diagnostic tool of choice as it is better able to identify areas of obstruction than CT, ultrasound, or plain films. MR can also allows direct measurement of CSF flow.
69 year old Caucasian man

- PMHx:
  1. Complex partial seizures
  2. Chronic atrial fibrillation
  3. S/P surgery for melanoma on scalp
  4. HTN
  5. CHF
  6. Obesity
  7. S/P bilateral total knee arthroplasties
  8. Benign prostatic hyperplasia
  9. Urinary incontinence
  10. Questionable dementia
  11. Congenital hydrocephalus
Clinical Case cont.

- Medication list
  - Lanoxin
  - Atenolol
  - Paroxetine
  - ASA
  - Phenytoin
  - Ditropan
  - Plendil
Physical exam is remarkable for no neurologic abnormalities except for slightly ataxic gait, inability to perform finger-nose-finger test satisfactorily, and inability to touch heel to shin.

- Patient is slightly macrocephalic
- Patient has never had a craniotomy (i.e. – ventriculoperitoneal or ventriculoatrial shunt never placed)
No complaints of diplopia, blurred vision, or tinnitus

Serial MRI’s of brain were done between 1998 and 2002 show marked, but stable, enlargement of lateral and third ventricles with a normal fourth ventricle, likely due to chronic congenital aqueductal stenosis, however, the aqueduct of Sylvius appears patent on imaging studies. There is compression of the cortices along the occipital, temporal, and parietal regions. There is a small amount of preserved cortex within bilateral frontal lobes.
Clinical Case MRI

T1-weighted sagittal image

Source: University of Illinois Hospital PACS
Clinical Case MRI

T1-weighted sagittal image

Source: University of Illinois Hospital PACS
Clinical Case MRI

T1-weighted sagittal image

Source: University of Illinois Hospital PACS
Clinical Case MRI

Source: University of Illinois Hospital PACS
Clinical Case MRI

Source: University of Illinois Hospital PACS

Cerebral aqueduct of Sylvius

T1-weighted sagittal image
Clinical Case MRI

T2-weighted axial image

Source: University of Illinois Hospital PACS
Clinical Case MRI

T2-weighted axial image

Source: University of Illinois Hospital PACS
Clinical Case MRI

T2-weighted axial image

Source: University of Illinois Hospital PACS
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T2-weighted axial image

Source: University of Illinois Hospital PACS
Clinical Case MRI

Source: University of Illinois Hospital PACS
Clinical Case MRI

T2-weighted axial image

Source: University of Illinois Hospital PACS
Clinical Case MRI

Source: University of Illinois Hospital PACS

T2-weighted axial images
Clinical Case MRI

T1-weighted coronal image

Source: University of Illinois Hospital PACS
Clinical Case MRI

T1-weighted coronal image

Source: University of Illinois Hospital PACS
Clinical Case MRI

Source: University of Illinois Hospital PACS
Clinical Case MRI

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Source: University of Illinois Hospital PACS

T1-weighted coronal images
Clinical Case MRI

Source: University of Illinois Hospital PACS

T1-weighted coronal images
Clinical Case MRI - 2000

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T1 sagittal, T2 axial, and T1 coronal images respectively from 5/2000 demonstrating stability of patient’s hydrocephalus

Source: University of Illinois Hospital PACS
Unique features of this case include

- Patient has never had a shunting procedure done to remove the excess CSF
- Patient has apparently been relatively stable for many years with little or no neurological degradation

Some of patient’s symptoms fit the symptomatology associated with Normal Pressure Hydrocephalus (NPH)

- Urinary incontinence (wet)
- Ataxia (wobbly)
- Decreased mentation (weird)
**Clinical Case - Discussion**

- **Reasons NPH is not a likely diagnosis**
  - Chronicity of hydrocephalus (patient has studies dating back to 1998 but apparently had been followed at an outside hospital for many years). Is much more likely that patient has hydrocephalus associated with increased intracranial pressure.
  - Decreased mentation of NPH is qualitatively different from the dementia this patient suffers from.

- **Please see handout for a more complete discussion of NPH**

- **Further imaging studies might involve performing MR quantification of CSF flow through the aqueduct of Sylvius.**
Treatment

- Generally involves diversionary shunting procedure to relieve the pressure caused by the excess CSF in the calvarium.

- It is not a perfect solution
  - Ependymal lining is generally not restored
  - Large blood vessels can resume their normal configuration but capillaries do not, or at least not quickly.
  - Once axons have been destroyed it is unlikely that they can be restored

- Early shunting is better than late shunting.

- In this patient since he has been hydrocephalic for many years (perhaps his whole life) it is unlikely shunting will prove efficacious. Observation is probably best.


http://www.cid.ch
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