Silent Brain Syndrome

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Chief Complaint: Excess tearing

History of Present Illness:
A 32 year-old gentleman presents with a 10-year history of excess tearing in both eyes. There has also been some mucous-like yellowish discharge. He states that the excess tearing hinders his vision.

Past Ocular History:
None

Past Medical History:
- Choroid plexus papilloma and resultant hydrocephalus after a resection and shunt placement at the age of 15. Subsequently, multiple shunt revisions were performed.
- Seizure disorder

Medications:
- Tegretol
- Phenobarbitol

Allergies: No known drug allergies

Family History: Non-contributory

Social History: Non-contributory

Review of systems: Negative on 12 -point review of systems

Ocular Exam:

Visual acuity:
- 20/30-2 PH 20/25-1 OD
- 20/25-2 PH 20/20-2 OS

Pupils: 4mm → 2mm, brisk, equal OU, no RAPD OU

Extraocular movements: Full, orthotropic OU

Confrontation visual field: Decreased peripheral visual field superiorly OU

External:
- Increased head circumference, enophthalmos OU
- Palpebral fissure: 8mm OD, 8mm OS
- Margin reflex distance 1: 4mmOD, 4mm OS
- Levator function 12mm OD, 12mm OS

Hertel exophthalmometry: 6mm OD, 5mm OS, base 97mm

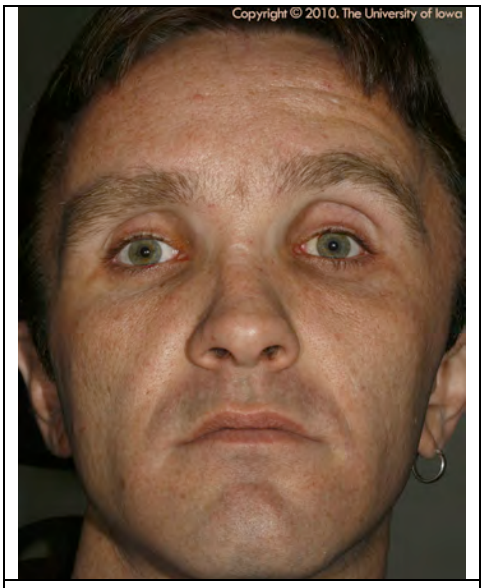

Figure 1. Full face photograph demonstrating bilateral enophthalmos

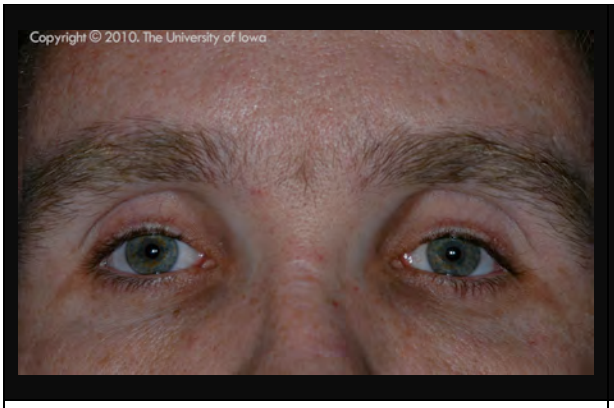

Figure 2. Close up view of bilateral enophthalmos

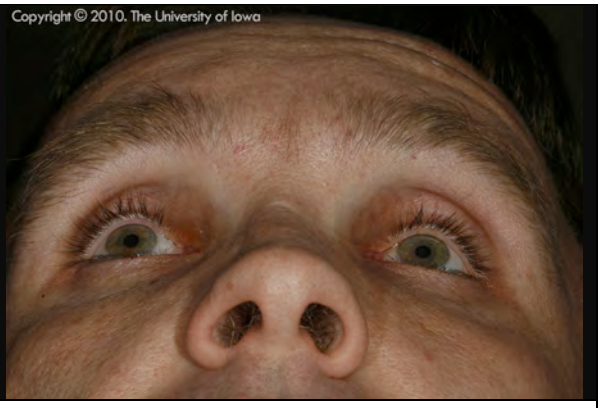

Figure 3. Bilateral enophthalmos with lack of apposition of all four eyelids from the ocular surface (also see Figure 4)

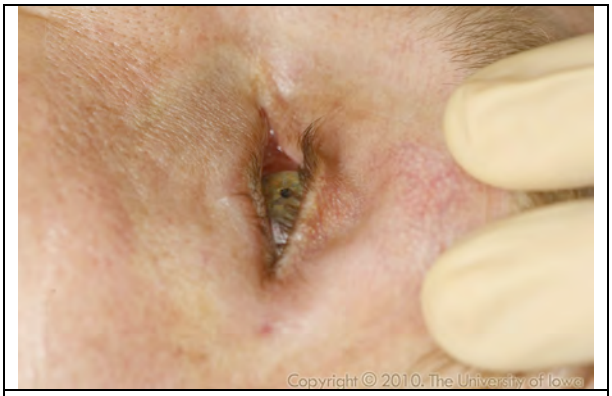

Figure 4. Side view demonstrating the lack of apposition of the eyelids to the globe

Slit Lamp Exam:

- Lids/Lashes: upper and lower puncta displaced from the globe OU. Very small upper and lower punta but patent, OU.
- Conjunctiva/Sclera: 1+ papillary reaction on upper and lower palpebral conjunctiva, trace subconjunctival hemorrhages superiorly and temporally OU
- Cornea: good tear film with tear break up time (TBUT) > 10 sec OU, dense staining inferiorly with Rose Bengal stain OU
- Anterior Chamber: deep and quiet OU
- Iris: Normal OU
- Lens: Clear OU
- Vitreous: Clear OU

Dilated Fundus Exam: Normal disc, macula, vasculature and periphery OU

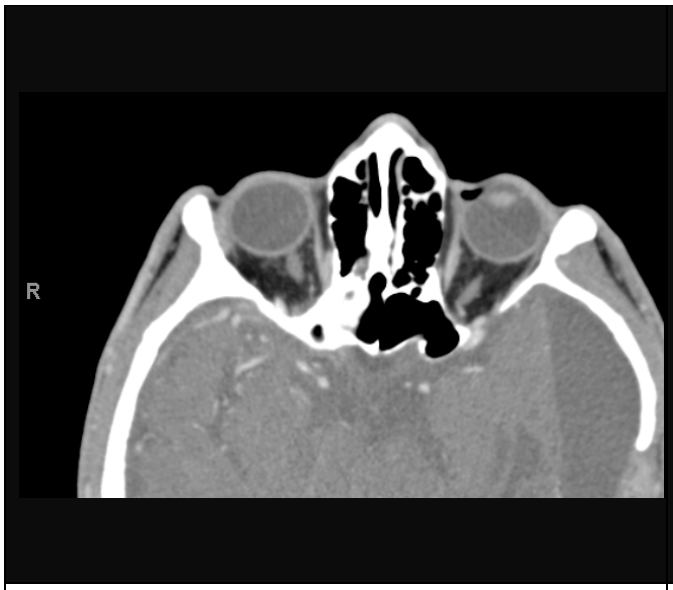

Figure 5. CT scan, axial view, confirming the bilateral enophthalmos

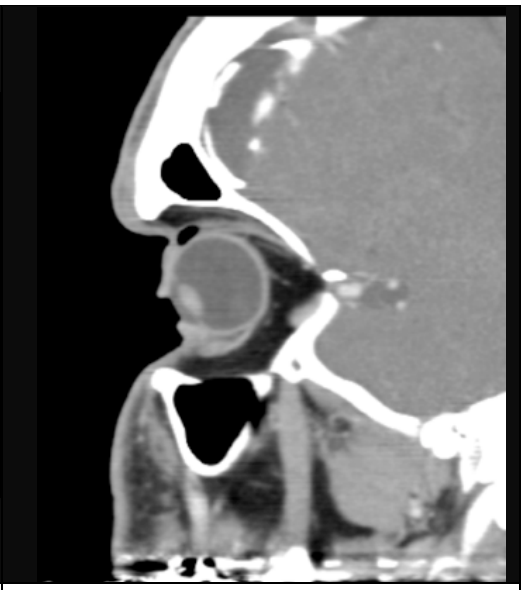

Figure 6. CT scan, sagittal view, demonstrating the upward bowing of the orbital roof and air entrapment under the upper eyelid

Diagnosis: Acquired enophthalmos consistent with silent brain syndrome

Discussion:

Bilateral secondary enophthalmos after ventriculo-peritoneal shunting (VPS) for hydrocephalus at a young age was first described in literature in 1996 by Meyer and colleagues. They described three patients with progressive severe bilateral enophthalmos who each had congenital hydrocephalus treated with VP shunts (Meyer, 1996).

Due to the severe lack of lid apposition to the globe, these patients, initially, present to ophthalmologists seeking treatment for dry, irritative ocular symptoms. This may include a foreign body sensation, tearing, epiphora, or less likely, decreased vision. Some patients do state a chief complaint of their eyes "sinking into the sockets." In addition, there can be entrapment of air underneath the eyelids (Figure 6). Most patients have suffered from

progressive worsening of these symptoms for multiple years prior to presenting to an ophthalmologist.

These patients, like our patient, usually have a previous history of hydrocephalus requiring a VPS early in life.

In 2008, Cruz *et al.* were the first to propose that VP shunting early in life can cause a reduction in size of the anterior cranial fossa, which, in turn, alters the structure of the orbit and leads to the development of enophthalmos. In young children, the thin floor of the frontal cranial fossa or the orbital roof is thought to be less calcified and, therefore, more susceptible to remodeling. These bones become progressively bowed superiorly over time as the intracranial pressure decreases (Cruz, 2008).

Bernadini *et al*. reported two patients with similar findings and labeled the disease as the "silent brain syndrome" (Bernadini, 2009). The silent brain syndrome is the counterpart to the "silent sinus syndrome," (Please see the "Silent sinus syndrome" EyeRounds case at http://www.eyerounds.org/cases/102-enophthalmos-silent-sinus-syn.htm)

Patients with silent brain syndrome can be treated with orbital volume augmentation since the enlarged orbital cavity is responsible for the enophthalmos, as well as the consequent absence of lid apposition and air entrapment. Surgically, an appropriately-sized implant can be placed to bridge the bowed orbital roof. If an implant is not used, lid malposition repair can be performed. However, this option is usually not as successful in resolving the symptoms as orbital roof implants. Post-operatively, the patients may need to be evaluated for strabismus surgery. Our patient underwent bilateral orbital implants which improved his enophthalmos and his presenting ocular symptoms.

Figure 7. Photograph after bilateral orbital volume augmentation with Medpor plates.

Differential Diagnosis of acquired enophthalmos:

- Silent sinus syndrome
- Orbital fractures
- Silent brain syndrome (CSF shunt surgery)
- Orbital varix
- Phthisis bulbi
- Chronic sinusitis
- Parry Romberg syndrome
- Osteomyelitis
- Atrophy of orbital fat
- Contraction of orbital fat (most commonly, breast cancer)
- Malignant infiltration
- Anophthalmic enophthalmos
- Pseudoenophthalmos
    - Contralateral exophthalmos
    - Horner's syndrome

Epidemiology:	**Signs:**

• Diagnosed with hydrocephalus at a young age with history of VP shunt.
  • Mean age of CSF shunting: 8.3 years.
  • Mean age of ocular symptoms: 20.4 years.
• Mean age at presentation to an ophthalmologist: 27.5 years (Bernardini, 2009)
  • No gender predilection
  • No racial predilection
	• Bilateral enophthalmos
	• Superficial exposure keratopathy
	• Epiphora
	• Trichiasis ± erosive keratopathy
	• Entropion
	• Lagophthalmos
	• Lid retraction

Symptoms:	**Treatment:**
• Tearing • Foreign body sensation • Decreased vision	• Orbital volume augmentation • Repositioning of eyelids • Possible strabismus surgery

References:

1. Bernadini FP, Rose GE, Cruz AAV, Priolo E. Gross Enophthalmos After Cerebrospinal Fluid Shunting for Childhood Hydrocephalus: The "Silent Brain Syndrome". Ophthal Plast Reconstr Surg 2009;25:434-436
2. Cruz AAV, Mesquita IG, Santos de Oliveira R. Progressive Bilateral Enophthalmos Associated with Cerebrospinal Shunting. Ophthal Plast Reconstr Surg 2008;24:152-154
3. Hardy TG, McNab AA. Bilateral Enophthalmos Associated with Paget Disease of the Skull. Ophthal Plast Reconstr Surg 2002;18:388-390
4. Meyer DR, Nerad JA, Newman NJ, Lin JC. Bilateral Enophthalmos Assocaited with Hydrocephalus and Ventriculoperitoneal Shunting. Arch Ophthalmol 1996;114:1206-1209

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