Blindness from hydrocephalus

General information

Blindness is a rare hydrocephalus complication and/or shunt malfunction. Possible causes include:

1. occlusion of the posterior cerebral artery (PCA caused by downward transtentorial herniation)

2. chronic papilledema causing injury to the optic nerve at the optic disc

3. dilatation of the third ventricle with compression of the optic chiasm.

Ocular motility or visual field defects are more common with shunt malfunction than is blindness.

One series found 34 reported cases of permanent blindness in children attributed to shunt malfunction with concomitant increased ICP (these authors were based in a referral center for visually impaired children, thus incidence was not estimated). Another series of 100 patients with tentorial herniation (most from acute EDH and/or SDH) proven by CT; 48 patients operated; only 19 of 100 survived>1 month (all were in operated group); 9 of 100 developed occipital lobe infarct (2 died, 3 vegetative states, remaining 4 moderate to severe disability) [1)].

Types of visual disturbance

9 of 14 had pregeniculate (anterior visual pathway) blindness with marked optic nerve atrophy (early) and reduced pupillary light reflexes. 5 of 14 had postgeniculate (cortical) blindness with normal light responses and minimal or no optic nerve atrophy (or atrophy late). A few patients had evidence of damage in both sites.

Cortical blindness: due to lesions posterior to lateral geniculate bodies (LGB), it may also be seen with hypoxic injuries or trauma. Occasionally associated with Anton's syndrome (denial of visual deficit) and with Ridoch's phenomenon (appreciation of moving objects without the perception of stationary stimuli).

Pathophysiology

In patients with occipital lobe infarction

Occipital lobe infarctions (OLI) in posterior cerebral artery PCA distribution are seen either bilaterally, or if unilateral are associated with other injuries to optic pathways posterior to the lateral geniculate nucleus. The most often cited mechanism is compression of posterior cerebral artery (PCA) resulting from brain herniating downward through the tentorial notch, where the PCA or its branches lie on the surface of the hippocampal gyrus and tend to cross the free edge of the tentorium (some authors

implicate parahippocampal gyral compression in tentorial notch directly injuring lateral geniculate nucleuss; this may never produce permanent blindness). Alternatively, upward cerebellar herniation (e.g. from a ventricular puncture in face of a p-fossa mass) may impinge on PCA or branches with the same results.

OLIs are more likely with a rapid rise in ICP (doesn't allow compensatory shifts and collateral circulation to develop). Macular sparing is common, possibly due to potential dual blood supply of occipital poles (sometimes filled both by PCA and MCA collaterals); alternatively, the calcarine cortex may be supplied by a distinct branch of the PCA that fortuitously escapes compression. Reported causes of OLI include posttraumatic edema, tumor, abscess, SDH, unshunted hydrocephalus, and shunt malfunction. The occipital poles are also particularly vulnerable to diffuse hypoxia; attested to by cases of cortical blindness after cardiac arrest. Hypotension superimposed on compromised PCA circulation (from herniation or elevated ICP) may thus increase the risk of postgeniculate blindness.

Both coup and contrecoup trauma may produce occipital lobe infarction. Unlike a PCA occlusion infarct, macular sparing is not expected in traumatic occipital lobe injury.

In patients with pregeniculate blindness

see Pregeniculate blindness.

Presentation

These deficits are frequently unsuspected (altered mental state and the youth of many of these patients makes detection difficult); an examiner must persevere to detect homonymous hemianopsias in an obtunded patient.

Pregeniculate blindness is less often associated with depressed sensorium than is postgeniculate (where direct compression and vascular compromise of the midbrain are more likely).

Prognosis

Cortical blindness after diffuse anoxia frequently improves (occasionally to normal); usually slowly (weeks to years quoted; several mos usually adequate). Many reports of blindness after shunt malfunction are pre-CT era; thus, the presence or extent of occipital lobe infarction cannot be ascertained. Some optimistic outcomes are reported; however, permanent blindness or severe visual handicap are also described; no reliable predictor has been identified. As with infarcts elsewhere, younger patients fare better, but extensive calcarine infarcts are probably incompatible with significant visual recovery.

1)

Arroyo HA, Jan JE, McCormick AQ, et al. Permanent Visual Loss After Shunt Malfunction. Neurology. 1985; 35:25–29