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Cortical blindness

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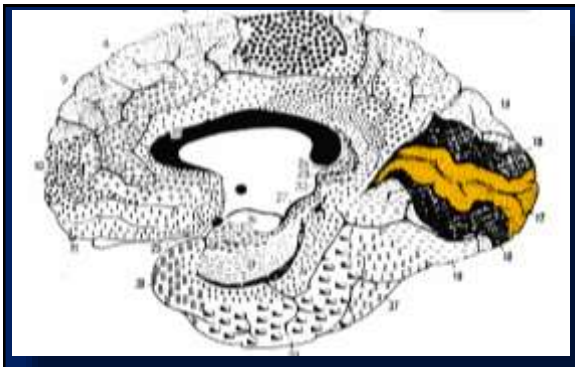
Defintion

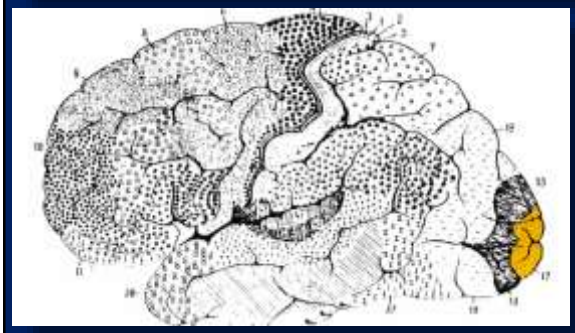
Total blindness due to bilateral occipital lobe lesions

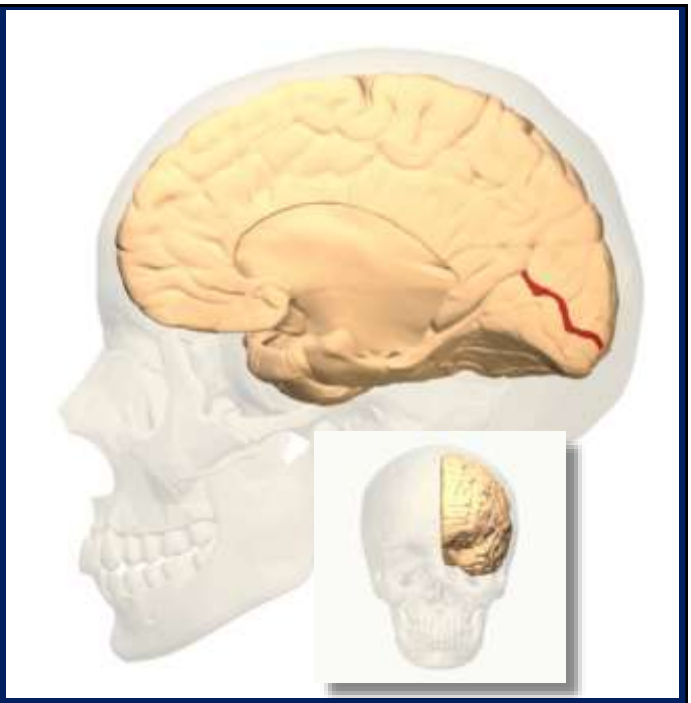
The term **cerebral blindness** is more accurate than cortical blindness, as the lesion may involve both gray and white matters, or even white matter only.

The Visual Cortex

- Located in occipital lobe
- The 1ry visual cortex receives sensory input from the lateral geniculate nucleus and processes visual information (*called also visual area 1 (V1), Brodmann area 17, or the striate cortex*).
- The extra striate areas are the visual areas 2, 3, 4, and 5 (also known as V2, V3, V4, and V5, or Brodmann area 18 and Brodmann area 19).







Criteria

- Total blindness
- Normal pupillary responses
- Normal fundi

Etiology

Vascular causes:

1. Infarction (the most frequent): bilateral cerebrovascular infarction, either simultaneous or consecutive
2. Hemorrhage
3. Hypertensive encephalopathy
4. Eclampsia

Etiology

Vascular causes:

5- Arteriovenous malformation (congenital)

6-Cerebral venous thrombosis

7-Vertebrobasilar arteritis (elderly)

8-Rupture of occipital mycotic aneurysms with endocarditis (rare)

Etiology

Vascular causes:

9 - Vascular complication of cardiac surgery (hemodynamic compromise or emboli)

10- Generalized hypotension, as induced by medications like nifedipine → infarction

11- Complications of cerebral angiography

Etiology

- **Demyelinating disease**
- **Alzheimer's disease**
- **Seizures (ictal or postictal)**
- **Hydrocephalus**

Etiology

- **Tumor:** (meningioma, metastasis, glioma)
- **Tentorial herniation:** from cerebral mass
- **Toxins:** (e.g., lead, mercury, ethanol, carbon monoxide)
- **Trauma**
- **Radiation encephalopathy**

Etiology

- **Infection:** progressive multifocal leukoencephalopathy,, subacute sclerosing panencephalitis, HIV encephalitis, syphilis, encephalitis, abscess, Jakob-Creutzfeldt disease
- **Inflammation**: sarcoidosis

Etiology

- **Metabolic:** adrenoleukodystrophy, hypoglycemia, hepatic encephalopathy, acute intermittent porphyria, mitochondrial encephalopathies
- **Medications:** Iodinated contrast agents, amphetamine, interleukin-2, tacrolimus, chemotherapy (Cisplatin, cyclosporine, vincristine, FK506)

❖Cerebral blindness can be divided into ***persistent*** and ***transient*** forms (e.g., vasospasm, postictal, drugs).

❖Infarction causes permanent blindness in 25% and residual visual field defects in the rest.

Clinically

- Bilateral complete or severe **hemianopia**
- Visual acuity at **light perception or worse**
- **No** detectable peripheral vision

Clinically

- **Normal** pupillary light responses
- **Normal** fundus
- **Normal** EOM movement, but no reflex lid closure in response to light or danger

Clinically

- **Cerebral (central) achromatopsia:** complete loss of color perception
- **Cerebral dyschromatopsia:** some residual color perception (more common) however both are rare.
- **Hemi-achromatopsia** and **hemi-dyschromatopsia**: color loss restricted to the contralateral hemifield

Clinically

- **Visual agnosia:** Patients no longer recognize previously familiar objects nor learn to identify new objects by sight alone.
- **Prosopagnosia:** Patients no longer recognize the faces of previously familiar persons nor learn newly encountered faces.

Clinically

- **Acquired alexia:** loss of reading ability in previously literate persons.
- **Alexia with Agraphia**
- **Cogan's rule**: a patient with homonymous hemianopia but otherwise neurologically intact is caused by a lesion in the occipital lobe.

Clinically

Anton's syndrome:

- In about of 10% of cases
- Denial of blindness and fabrication of an imaginary visual environment
- Patients are not aware of their visual deficit and insist that they can see

Investigations

- Visual evoked potentials can be normal in patients with striate lesions.
- Absent alpha rhythm on electroencephalography is a more sensitive diagnostic sign.

Investigations

- CT scans can be normal
- MRI with coronal images through the occipital lobe demonstrates most lesions.
- SPECT (Single photon emission computed tomography scans) may reveal bilateral functional defects in cases with only unilateral structural lesions on MRI.

General criteria of occipital lesions:

- **Extremely congruous homonymous** visual field defects from whether they are hemianopic or involve a lesser area of field.
- Lack of congruity rules out an occipital lobe defect
- The contrary is not true, i.e., congruous homonymous visual field defects may occur from lesions anywhere along the retro-chiasmal pathways.

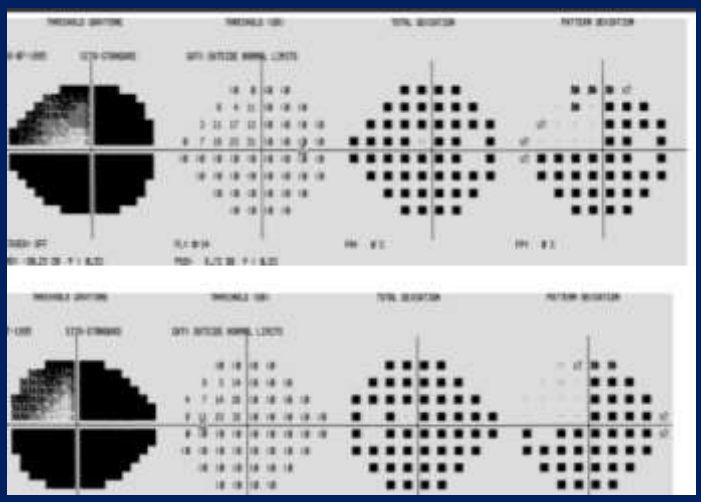
General criteria of occipital lesions:

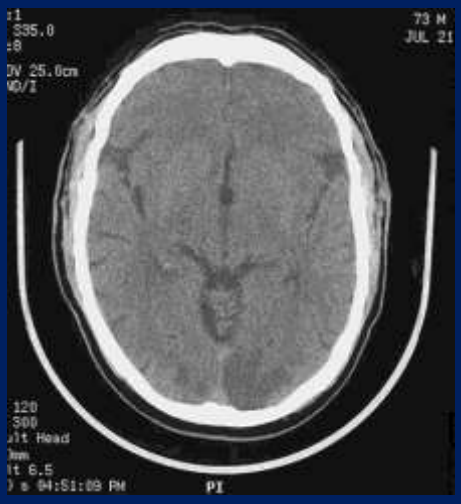
Macular sparing: occipital hemianopias preserve an area around central fixation (Bilateral homonymous hemianopia with macular sparing → key hole field).

Mechanisms:

1-Dual vascular supply (watershed zone between branches of the middle and posterior cerebral a.)

2-Dual nerve supply to both sides of the occipital cortex???



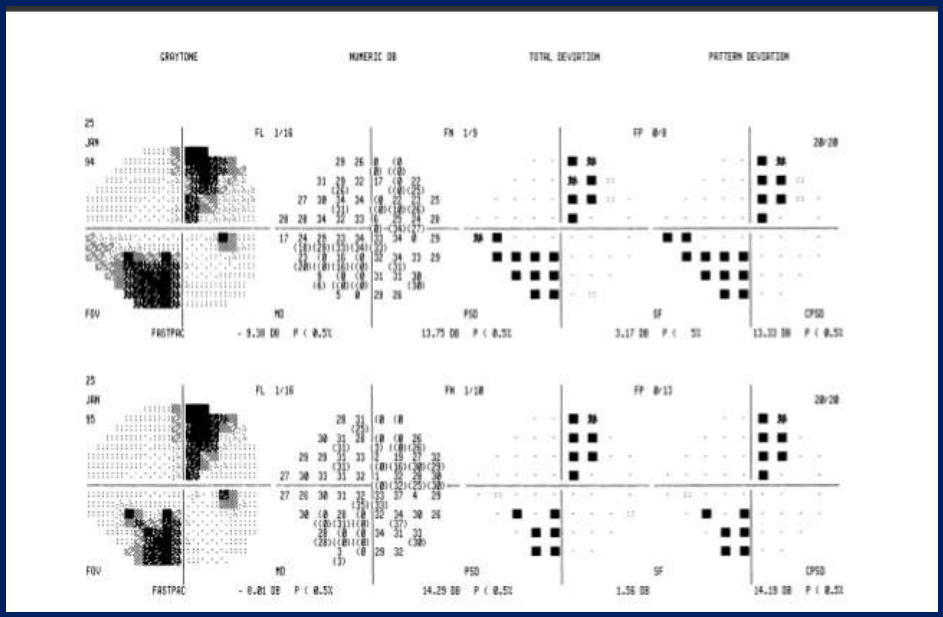


Vascular infarct. On neuroimaging there is a low attenuation signal in the occipital lobe greater on the left side than the right. (b)
Visual field assessment there is a complete congruous right hemianopia and incomplete left hemianopia (bilateral hemianopia). Central vision is spared – note the decibel values.

General criteria of occipital lesions:

Checkerboard field:

- Bilateral ***sectorinopias***, superior on one side and inferior on the contralateral side
- For example, a right superior quadrantopia combined with a left inferior quadrantopia
- The upper and lower calcarine lips can be involved separately by ischemia because the lips have separate blood supplies.



A congruous right superior quadrantanopia and congruous left inferior quadrantanopia

General criteria of occipital lesions:

Temporal crescent syndrome:

- Fibers representing the nasal retina synapse more anteriorly along the interhemispheric fissure, and can be spared in certain occipital strokes, causing preservation of a temporal crescent of field.
- Less frequently, this is the only portion of the visual cortex damaged, resulting in loss of this temporal crescent.

General criteria of occipital lesions:

Riddoch phenomenon: in retro-geniculate lesions= the perception of moving but not static targets.

Unformed visual hallucinations: Unlike the formed hallucinations associated with temporal lobe disease, lesions of the visual cortex usually cause unformed visual hallucinations (paroxysmal scintillating scotomas and light flashes).

Thank You