بِسْمِ اللَّهِ المُحمَد
Neuro-Ophthalmology
Disorders of the Optic Nerve.

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Scenario

• A 30 year old lady is referred to ophthalmic outpatient clinic with gradual decrease of vision in the left eye for 5 days.
• She felt lately numbness of fingers after a hot shower
Examination

• VAR—6/6
• VAL—6/60
• Left— Afferent pupillary defect
• Left—defective color vision
• Other exams.—normal
Questions

• What is your most likely diagnosis?
• How to confirm your diagnosis?
• How to treat?
Applied Anatomy

50mm long from globe (Lamina caribrosa) to chiasma

- Intraocular part: 1 mm long, 1.5 mm thick
  - Prelaminar zone
  - Laminar zone
  - Pot laminar zone

- Intra orbital: 25-30 mm long to the optic foramen
  - 3-4 mm thick, surrounded by annulus of Zinn

- Intra canalicular: 6 mm long, fixed to the canal

- Intra cranial: 5-16 mm long (Av. 10 mm)
Contains 1.2 million axons, most of these synapse in the lateral geniculate body. Some reaches other centres.

- 1/3 of fibres sub serve the central 5 degrees of visual field.
Surrounding sheaths

• Pia mater is the delicate innermost sheath containing blood vessels.
• Subarachnoid space is continuous with the cerebral subarachnoid space and contains CSF.
• Outer sheath comprises the arachnoid mater and the tougher dura mater. The latter is continuous with the sclera.
1. Without systemic associations
   - Tilted optic disc
   - Optic disc drusen
   - Optic disc pit
   - Myelinated nerve fibres

2. With systemic associations
   - Optic disc coloboma
   - Morning glory anomaly
   - Optic nerve hypoplasia
   - Aicardi syndrome
   - Megalopapilla
   - Peripapillary staphyloma
   - Optic disc dysplasia
Tilted disc

- Common, bilateral
- Frequent myopia and astigmatism
- VA - normal
- Small disc, oval or D-shaped
- Axis oblique (most common), horizontal or vertical
- Situs inversus and inferior crescent
- Hypopigmented inferonasal fundus
- Superotemporal field defects not obeying vertical midline
Optic disc drusen

- Uncommon, bilateral and familial
- Associations - RP, angioid streaks and Alagille syndrome
- VA - usually normal

Buried drusen

- Absent optic cup
- Pink or yellow colour
- Indistinct ‘lumpy’ margins
- Anomalous branching patterns with premature branching
- Absent venous engorgement
# Optic disc drusen

<table>
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<tr>
<th>Exposed</th>
<th>Occasional complications</th>
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<tbody>
<tr>
<td><img src="image1.png" alt="Exposed Image" /></td>
<td><img src="image2.png" alt="Exposed Image" /></td>
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</table>
| - Emergence from disc surface during early ‘teens’  
- Waxy pearl-like irregularities | - Choroidal neovascularization  
- Nerve fibre bundle defects |

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**Optic disc drusen** are occasional complications that may occur.

- Emergence from disc surface during early ‘teens’
- Waxy pearl-like irregularities

- Choroidal neovascularization
- Nerve fibre bundle defects
Optic disc pit

- Uncommon, usually unilateral
- VA - normal if uncomplicated
- Large disc containing round or oval pit
- Pit is usually temporal, occasionally central
Myelinated nerve fibres

| Isolated peripheral | Peripapillary | Extensive |
Optic disc coloboma

- Rare, unilateral or bilateral
- Usually sporadic - occasionally dominant
- VA - decreased

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<th>Signs</th>
<th>Ocular associations</th>
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<td>• Large disc with inferior excavation</td>
<td>• May be associated with other colobomata</td>
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<td>• Superior visual field defects</td>
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Signs

Ocular associations
Occasional Systemic Associations of Optic Disc Coloboma

1. **CNS malformation** - basal encephalocele and cysts

2. **Chromosomal anomalies** - Patau syndrome (trisomy 13) and cat-eye syndrome (trisomy 22)

3. **‘CHARGE’** - Coloboma, Heart defects, choanal Atresia, Retarded development, Genital and Ear anomalies

4. **Other syndromes** - Meckel-Gruber, Goltz, Lenz microphthalmos, Walker-Warburg and Goldenhar
Optic nerve hypoplasia

- Rare, unilateral or bilateral
- VA - variable according to severity

- Small disc surrounded by halo (double ring sign)
- Vessel normal calibre but may be tortuous

Occasional association

De Morsier syndrome (septo-optic dysplasia)
Absence of septum pellucidum and corpus callosum
Signs of optic nerve dysfunction

- Reduced visual acuity
- Afferent pupillary conduction defect
- Dyschromatopsia
- Diminished light brightness sensitivity
Applied anatomy of afferent conduction defect

**Anatomical pathway**

**Signs**

- Equal pupil size
- Light reaction
  - ipsilateral direct is absent or diminished
  - consensual is normal
- Near reflex is normal in both eyes
- Total defect (no PL) = amaurotic pupil
- Relative defect = Marcus Gunn pupil
Optic disc changes

- Early compression
- Papillitis and neuroretinitis
- Optic nerve sheath meningioma
- Compression

Normal

Swelling

Optico-ciliar shunts

Atrophy
Visual field defects

Central scotoma  Centrocaecal scotoma
Etiologic Classification of optic nerve disease

• **Optic nerve abnormalities congenital**
  – Hypoplasia
  – Dysplasia
  – Tilted disks
  – Myelinated nerve fibers

• **Hereditary optic atrophy**
  – Laber’s hereditary optic neuropathy

• **Optic neuritis**
  – Demyelinative
  – Immune-mediated
  – Direct infections
  – Granulomatous optic neuropathy
  – Contiguous inflammatory disease
Etiologic Classification of optic nerve disease

- **Vascular (ischemic optic neuropathy)**
  - Nonarteritic anterior ischemic optic neuropathy
  - Giant cell arteritis
  - Systemic vasculitis
  - Migraine
  - Inherited coagulation defects
  - Diabetic papillopathy
  - Radiation optic neuropathy
  - Sudden massive blood loss
Etiologic Classification of optic nerve disease

- Papilloedema
  - Space-occupying lesions
  - Blockage of ventricular system
  - Obstruction of CSF absorption
  - Benign intracranial hypertension (pseudotumour cerebri)
  - Diffuse cerebral oedema
  - Hypersecretion of CSF
Etiologic Classification of optic nerve disease

- **Optic nerve compression**
  - Intracranial disease
  - Orbital disease
  - Optic nerve sheath meningioma
  - Trauma

- **Nutritional and toxic**
  - Vitamin deficiencies
  - Tobacco-alcohol amblyopia
  - Heavy metals
  - Drugs
  - Chemicals (Methanol)
Etiologic Classification of optic nerve disease

• **Optic nerve atrophy**
  – Primary optic atrophy
    • Following retrobulbar neuritis
    • Compressive lesions such as tumours and aneurysms
    • Hereditary optic neuropathies
    • Toxic and nutritional optic neuropathies
    • Degenerative retinal diseases
  – Secondary optic atrophy
    • Chronic papilloedema
    • Anterior ischeamic optic neuropathy
    • Papillitis
Optic Neuritis

Ophthalmoscopic classification

- Retrobulbar neuritis (normal disc)
- Papillitis (hyperaemia and oedema)
- Neuroretinitis (papillitis and macular star)
Optic Neuritis

• **Aetiological Classification**
  – Demyelinating
  – Parainfectious
  – Infectious
  – Autoimmune
Optic Neuritis

- **Demyelinating optic neuritis**
  - **Presentation**
    - Subacute monocular visual impairment, (bilateral)
    - Discomfort around the eyes, (increased with ocular movements)
    - Frontal headaches,
    - Tenderness or the globe
  - **Signs**
    - Visual acuity 6/18 - 6/60
    - Optic disc normal / papillitis
    - Colour vision defects
    - RAPD
    - VEP delayed conduction
    - Diminished light brightness sensitivity
    - Diminished contrast sensitivity
Optic Neuritis

• **Visual field defects**
  – Diffuse depression of sensitivity in the central 30°
  – Central scotoma
  – Ceco-central scotoma

• **Course**
  – Recovery usually within 2-3 weeks maximum at 6 months

• **Prognosis**
  – 75% recover visual acuity better than 6/9
  – 85% recover visual acuity better than 6/12
Optic Neuritis

• **Treatment**
  
  – **Indications**
    • If there is poor vision in the other eye
    • If the visual acuity at the onset is less than 6/12
  
  – **Regimen**
    • I/V methyl prednisone sodium 1g for 3 days
    • Followed by oral prednisolone 1mg/kg daily x 11days
  
  – **Benefits**
    • Delays further neurological events with MS by 2 years
    • Hastens visual recovery from optic neuritis but does not appear to have any long term benefit on final visual acuity
TREATMENT

Acute Relapses of MS

• For clinically significant symptoms/lesions, consider methylprednisolone (Solumedrol), 250 mg IV q6h x 3-5 days followed by an oral prednisolone (Deltacortil) in tapering dose

• Steroids hasten the rate of recovery from acute exacerbations but have never been proved to improve overall outcome

• Plasmapheresis, ACTH, and cyclophosphamide are options for patients who fail methylprednisolone therapy
Interferon Therapy

- Interferon (INF) therapy has produced almost equivalent levels of excitement and disagreement among neurologists regarding effectiveness and indications.

- It is generally well tolerated, with flulike symptoms and injection site reactions being the most common reactions.
TREATMENT

• Other agents include mitoxantrone, IV immuno-globulin, cyclophosphamide, cladribine and methotrexate
Papilloedema

1. Introduction

- Circulation of cerebrospinal fluid
- Causes of raised intracranial pressure
- Hydrocephalus

2. Classification of papilloedema

- Early
- Established (acute)
- Longstanding (chronic)
- Atrophic (secondary optic atrophy)
Circulation of cerebrospinal fluid

(a) Subarachnoid space
(b) Lateral ventricle
(c) Third ventricle
(d) Aqueduct
(e) Fourth ventricle
Causes of Raised Intra-cranial Pressure

- Space-occupying lesions
- Blockage of ventricular system
- Obstruction of CSF absorption
- Benign intracranial hypertension (pseudotumour cerebri)
- Diffuse cerebral oedema
- Hypersecretion of CSF
Hydrocephalus

Dilated cerebral ventricles

**Communicating** – obstruction to CSF flow in basilar cisterns or cerebral subarachnoid space

**Non-communicating** – obstruction to CSF flow in ventricular system or at exit of foramina of fourth ventricle
Early papilloedema

- VA - normal
- Mild disc hyperaemia
- Indistinct disc margins – initially nasal
- Mild venous engorgement
- Normal optic cup
- Spontaneous venous pulsation - absent
  (also absent in 20% of normal)
Established papilloedema (acute)

- VA - usually normal
- Severe disc elevation and hyperaemia
- Very indistinct disc margins
- Obscuration of small vessels on disc
- Marked venous engorgement
- Reduced or absent optic cup
- Haemorrhages + cotton-wool spots
- Macular star
Longstanding papilloedema (chronic)

- VA - variable
- Marked disc elevation but less hyperaemia
- Disc margins - indistinct
- Variable venous engorgement
- Absent optic cup
Atrophic papilloedema (secondary optic atrophy)

- VA - severely decreased
- Mild disc elevation
- Indistinct disc margins
- Disc pallor with few crossing vessels
- Absent optic cup
Disorders of the chiasm

Anatomy of chiasm and pituitary gland

Size 8x12x4mm
Cross roads of 2.4 M of axons
More than 50% fibres cross to the opposite side
10mm above the diaphragm sellae
Surrounded by internal carotid and communicating arteries
Disorders of the chiasm

Normal anatomical variations

Central - 80%

Prefixed - 10%

Postfixed - 10%
Pituitary adenomas

Cushing syndrome

ACTH

Basophil

Acidophil

Chromophobe

PROLACTIN

Growth hormone

Acromegaly

Gigantism

Amenorrhoea

Infertility

Galactorrhoea

Hypoglandism

Impotence

Infertility

Gynaecomastia

Galactorrhoea
Acromegaly

Enlargement of hands and feet

Enlargement of lower jaw
Acromegaly

- Facial coarseness
- Hypertension, diabetes and gonadal dysfunction

- Organomegaly
- Carpal tunnel syndrome and cardiomyopathy
Visual field defects in pituitary adenomas

Decussating fibres are most vulnerable
MRI of pituitary adenoma

Sagittal

Axial

Coronal
Treatment options for pituitary adenomas

- Surgery
- Radiotherapy
- Trans-sphenoidal
- Bromocriptine
Craniopharyngioma

Presents
- In children with endocrine dysfunction
- In adults with visual field defects

The posteriorly crossing fibres are most vulnerable
Meningioma

Typically affect middle-aged women
THANK YOU
THANK YOU