Neuro-ophthalmology د.يوسف

The visual pathway from the retina may be divided in to seven levels:
1- Optic nerve: is a tract of the brain surrounded by meninges. It is formed by the axons of the ganglion cells of the retina. Its total length is 5 cm.
2- Optic chiasma: in which there is decussation of the fibers coming from the nasal retinae. Below which lies the pituitary gland.
3- Optic tract: traveling posterolateraly from the angle of the chiasma to reach the lateral geniculate body and then to the visual cortex.
4- Lateral geniculate body: provides a relay station for retinal axons synapsing with neurons of the optic radiation.
5- Optic radiation: arises in the lateral geniculate body and carries visual impulses to the occipital cortex.
6- The striate cortex: lies on the medial aspect of the occipital lobe.
7- The prestriate cortex: the area surrounding the striate cortex. It is concerned with visual memory. Lesion of this area causes visual agnosia (inability to recognize things by sight) and inability to judge distances.

Optic atrophy: may be primary or secondary

1- **Primary optic atrophy**: occurs without antecedent swelling of the optic nerve head e.g. following retrobulbar neuritis, compressive lesions such as tumors or aneurysms, hereditary optic neuropathies, and toxic and nutritional optic neuropathies.

   Signs

   Pale, flat disc with clearly delineated margins, with reduction in the number of small blood vessels on the disc surface.

2- **Secondary optic atrophy:** is preceded by swelling of the optic disc e.g. following papilloedema, anterior ischaemic optic neuropathy and papillitis.

   Signs

   White or dirty grey, slightly elevated disc with poorly delineated margins and reduction in the number of small blood vessels on the disc surface.

   Signs of optic nerve dysfunction

   1- Reduced visual acuity.
   2- Relative afferent pupillary defect.
   3- Dyschromatopsia (impairment of colour vision),mainly affects red and green.
   4- Diminished light brightness sensitivity.
   5- Diminished contrast sensitivity.
   6- Visual field defects.

Optic neuritis

Is an inflammatory, infective or demyelinating process affecting the optic nerve.

Ophthalmoscopical classification:

1-Retrobulbar neuritis: in which the optic disc appearance is normal. It is the most frequent type in adults and is frequently associated with multiple sclerosis.

2-Papillitis: characterized by hyperaemia and oedema of the optic disc, which may be associated with peripapillary flame shaped haemorrhages. It is the most frequent of optic neuritis in children.

3-Neuroretinitis: characterized by papillitis in association with inflammation of the retinal nerve fiber layer. It is the least common type and is only rarely a manifestation of demyelination.

Aetiological classification:

I- Demyelination, the most common.

2- Parainfectious, may follow viral infection or immunization.

3- Infectious, may be sinus-related or associated with cat-scratch fever, syphilis, lyme disease and herpes zoster.

4- Autoimmune associated with autoimmune diseases.

Demyelination:

Is a pathological process by which myelinated nerve fibers lose their myeline sheath. The myelin is phagocytosed by microglia and macrophages and subsequent to which astrocytes lay down fibrous tissue. Demyelination will disrupt nervous conduction within the white matter tracts within the brain, the brain stem, and the spinal cord.

Demyelinating optic neuritis:

Presentation: sub acute monocular visual impairment, discomfort in or around the eye exacerbated by eye movements.

Signs: 1-Visual acuity between 6/18 and 6/60, but may rarely be worse.

2-Other signs of optic nerve dysfunction, particularly impaired colour vision and a relative afferent pupillary defect and Visual field defects.

3- The optic disc is normal in the majority of cases (retrobulbar neuritis), the remainder show papillitis.

4- Temporal disc pallor may be seen in the fellow eye, indicative of previous optic neuritis.

MRI: shows periventricular plaques of demyelination.

Course: Vision worsens over several days to 3 weeks and then begins to improve. Initial recovery is fairly rapid and then slowly improves over 6–12 months.

Prognosis: More than 90% of patients recover visual acuity to 6/9 or better.

Treatment:

Indicated when VA within the first week is worse than 6/12 especially when the disease is bilateral or when the patient has poor vision in the fellow eye.

Steroid regimen. Intravenous methylprednisolone sodium succinate 1 g daily for 3 days, followed by oral prednisolone (1 mg/kg daily) for 11 days, subsequently

tapered over 3 days. Oral prednisolone may increase the risk of recurrence of optic neuritis if used without prior intravenous steroid.

Non-arteritic anterior ischaemic optic neuropathy:

Pathogenesis: partial or total infarction of the optic nerve head due to occlusion of the short posterior ciliary arteries.

Predisposing factors: hypertension, diabete mellitus, hypercholesterolaemia, collagen vascular diseases, antiphosphilipid antibody syndrome, sudden hypotensive events and cataract surgery.

Clinical features:

Age of presentation is between 45-65 years with sudden painless monocular visual loss on awakening suggesting that nocturnal hypotension may play an important role.

Signs: 1- V.A. is slightly reduced.

2- Visual field defect is typically inferior.

3- Dyschromatopsia.

4- Diffuse or sectoral hyperaemic disc swelling, often associated with a few peripapillary splinter haemorrhages.

Fluorescein angiography: in late stages shows unequal choroidal filling.

Special Investigations: should include blood pressure, a fasting lipid profile and blood glucose. It is also very important to exclude occult giant cell arteritis

Course: Disc swelling gradually resolves and pallor ensues 3–6 weeks after onset.

Prognosis: About 50% of eyes achieve 6/9 or better although recurrence occurs in about 6%. Involvement of the fellow eye occurs in about 10%.

Treatment: no definitive treatment. Underlying conditions should be treated.

Arteritic anterior Ischaemic optic neuropathy:

It is caused by giant cell arteritis. It typically affects elderly patients (average 70 years) and has a predilection for medium sizes and large arteries. Particularly the superficial temporal, and the posterior ciliary arteries.

The most important diagnostic criteria of giant cell arteritis are;

1- Age (average 70 years).

2- Jaw Claudication.

3- Temporal artery tenderness to palpation or decreased pulsation

4-ESR of 50 mm/hr or greater.

5- Abnormal artery biopsy.

Clinical features: presentation is with sudden, profound unilateral visual loss accompanied by periocular pain. Most cases occur within few weeks of the onset of giant cell arteritis.

Signs: 1- Severe visual loss is the rule, commonly to only perception of light or worse. 2- A pale 'chalky white' oedematous disc.

Fluorescein angiography: shows severe hypoperfusion of the choroid.

Course: Over 1-2 months severe optic atrophy occurs.

Prognosis: very poor, because visual loss is usually permanent.

Treatment

Is to prevent blindness of the fellow eye. Intravenous methylprednisolone 1gm/day daily for 3 days followed by oral prednisolone 1–2 mg/kg/day, then gradually tapered until symptoms resolution and ESR/CRP normalization.

Papilloedema:

Is a swelling of the optic nerve head, secondary to raised intracranial pressure.

Clinical features: Papilloedema is nearly always bilateral, but may be asymmetrical. in early stages there is hyperaemia and mild elevation with indistinct margins of the optic disc but VA is normal. In late stages there is marked elevation and even optic atrophy with severe loss of vision may occur.

Third nerve palsy:

Causes of isolated third nerve palsy:

1-Idiopathic.

2-Vascular disease e.g. hypertension and D.M.

3-Trauma.

4-Aneurysm of the posterior communicating artery.

5-Miscellanous causes: tumours, syphilis and vasculitis.

Signs:

1-Ptosis due to weakness of the levator muscle.

2- Limited adduction due to medial rectus weakness.

3- Limited elevation due to weakness of the superior rectus and inferior oblique.

4- Limited depression due to weakness of the inferior rectus.

5- Abduction and depression in the primary position (down and out) due to unopposed action of the lateral rectus and superior oblique muscles.

6- Dilated pupil and defective accommodation due to parasympathetic palsy.

Treatment:

1- Non-surgical by using prisms, uniocular occlusion to avoid diplopia if ptosis is partial or recovering. And botulinum toxin injection in to the uninvolved lateral rectus to prevent its contracture.

2- Surgical treatment: considered after 6 months.