The optic nerve: a clinical perspective

Pasquale Montaleone (Meds 2011)
Faculty reviewer: Dr. Robin Deans, Department of Ophthalmology, UWO

Introduction: optic nerve anatomy

The optic nerve carries approximately 1.2 million afferent nerve fibres which originate in the retinal ganglion cells of the retina. Most of these synapse in the lateral geniculate body, while others are destined for the pre-tectal nuclei in the midbrain and other centres. The optic nerve is approximately 50 mm long from globe to chiasm and can be divided into 4 subdivisions. First is the intraocular segment, which is called the optic disc or optic nerve head. Behind the globe is the intraorbital segment, which is 25-30 mm long. Here the nerve thickens with the addition of myelin sheaths that surround the nerve fibres, a function of oligodendroglia. Interestingly, the optic nerve within the orbit is S-shaped which allows the eye to move without stretching the nerve. The nerve will only become stretched if there is severe proptosis.

After exiting the orbit, the optic nerve enters the optic canal to be directed upward and inward at a 45-degree angle towards its chiasmal destination. This is called the intracanalicular segment, and is approximately 6 mm in length. Last is the intracranial segment, where the two optic nerves meet at the optic chiasm. Here the optic nerve fibres from each eye decussate so that each optic tract carries fibres from the contralateral nasal hemiretina and ipsilateral temporal hemiretina. Based on the arterial supply of the optic nerve, it can alternatively be divided into two segments, anterior and posterior. The anterior segment is simply the optic disc, which is supplied by the posterior ciliary arteries. There is also a minor arterial supply by an anastomotic circle called the circle of Zinn-Haller. The posterior segment consists of the intraorbital, intracanalicular, and intracranial subdivisions and is supplied by a more complex system of peripheral (centripetal) and axial (centrifugal) vessels. Venous drainage of the optic nerve occurs almost exclusively via the central retinal vein.

Examination

Disorders of the optic nerve affect the main components of vision which are contrast, brightness and colour. There are four main tests used to evaluate these components and ultimately optic nerve function.

Visual acuity

This is performed with the aid of a Snellen chart, as the patient reads the smallest line of identifiable letters from a distance of 20 feet (6 metres).

Colour plates

In this test a series of colour plates are used to determine colour recognition in each eye. Loss of signal transmission from cone receptors that are concentrated in the macula through the optic nerve is indicative of optic nerve dysfunction. There can be a marked difference in colour recognition between eyes if there is unilateral optic nerve damage. One should consider that approximately 5% of males have some degree of red-green colour blindness while performing this test.

Visual fields

The simple screening examination is confrontation visual field testing. The examiner stands approximately 1 metre from the patient with each person covering one eye and looking straight ahead. A target finger is moved centrally until seen by the patient and this is repeated in several quadrants. A counting fingers test may be more reliable and automated machine testing is the most sensitive means of detection.

Swinging flashlight test

This is one of the most valuable tests of optic nerve dysfunction available for the general physician. The abnormality detected is a Relative Afferent Pupillary Defect (RAPD), also known as a Marcus Gunn pupil. The key concept is that the optic nerve is responsible for the afferent limb of the pupillary reflex. In a dim room, light shone on a normal eye will result in consensual and contralateral pupillary constriction. The flashlight is then quickly moved to shine on the other eye and if there is optic nerve dysfunction in that eye then both pupils will abnormally dilate.

Optic nerve disorders

Ischemic Optic Neuropathy (ION)

ION is the most common acute optic neuropathy in patients over 50. As the name implies it is caused by optic nerve ischemia, and patients classically present with abrupt onset of painless, unilateral visual loss. It can be subdivided into anterior ION (AION) vs. posterior ION (PION) based on the location of the ischemia, or arteritic vs. non-arteritic based on the etiology of the ischemia.

Arteritic AION is caused by systemic vasculitis that affects the optic disc, most commonly from temporal arteritis (TA). Prompt diagnosis and treatment are imperative as TA can be lead to blindness or death. Headache, often severe, is the most common symptom associated with TA, occurring in approximately 50% of cases. Other
classical symptoms include jaw claudication, scalp tenderness, anorexia, weight loss, anemia, fever and visual loss. Elevated inflammatory markers - C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) - strongly suggest the diagnosis, but temporal artery biopsy remains the gold standard and is recommended in all suspected cases. The hemoglobin level should always be considered when reviewing the ESR as anemia may mask an elevated ESR. CRP, on the other hand, is not affected by hematologic factors or age.

Treatment of TA is with high dose corticosteroids. The vision loss is often more severe than its non-arteritic counterpart, as 75% of patients with arteritic AION demonstrate visual acuity of 20/200 or worse. Transient visual loss, called amaurosis fugax precedes the profound visual loss in up to 30% of cases. Treatment of TA is with high dose corticosteroids. The vision loss is often more severe than its non-arteritic counterpart, as 75% of patients with arteritic AION demonstrate visual acuity of 20/200 or worse. Transient visual loss, called amaurosis fugax precedes the profound visual loss in up to 30% of cases.

Non-arteritic AION is the most common form of ION. Caucasians account for 95% of cases, and individuals typically present in their 60's. The exact pathogenesis is unknown. It is a presumed diagnosis that is made when TA is ruled out. Visual loss may be less profound and partial field loss is common. Contrary to what one might assume it is not triggered by an embolic event as in ischemic stroke. There is no proven treatment for non-arteritic AION. Metabolic issues such as diabetic control and hypertension should be considered and treated appropriately.

PION is defined as infarction of the retrobulbar portion of the optic nerve, and accounts for only 10% of cases of ION. It is characterized by a normal appearing optic disc that later becomes atrophic. TA is the most common cause of PION. It is very important to rule out TA in all older patients who present with an afferent pupillary defect or unexplained visual field defect.

**Optic neuritis (ON)**

ON is characterized by inflammation of the optic nerve. It must be thought of in cases of visual loss in younger patients. Clinical features of ON include visual loss, RAPD, optic disc edema and a classic central scotoma. Most patients improve without treatment, however intravenous steroids may hasten recovery and may reduce the risk of developing multiple sclerosis (MS). An MRI should be done to ascertain the long term risk of developing MS as multiple white matter lesions on an MRI increases the risk of developing MS to over 50%.

**Glaucoma**

Glaucoma is a disease that causes nerve fibre layer loss within the intraocular portion of the optic nerve. Cupping of the disc is apparent before the disc becomes pale. The exact mechanism of glaucoma is not fully understood, but it is highly associated with elevated intraocular pressure (IOP). There are two types of glaucoma, open-angle and closed-angle.

Open-angle glaucoma is a chronic condition account-
ing for the majority of cases. Patients have chronically elevated IOP due to impaired drainage of aqueous humour from trabecular meshwork dysfunction. Visual loss is gradual and is characterized by peripheral visual field contraction and scotomas (blind spots). Generally the patient does not notice the visual field loss until late, which is why screening, especially in those with risk factors is important. Risk factors include increasing age, family history, African-American race and IOP above 22 mm Hg. Treatment is commonly with medications that either decrease aqueous production or increase drainage, and less commonly with surgery.

Angle-closure glaucoma, also known as acute glaucoma, is a medical emergency. It is most often caused by pupillary block between the iris and lens which then bows the iris forward blocking the angle and aqueous drainage through the trabecular meshwork. Complete vision loss can occur within hours. Patients typically present with a red and painful eye, nausea and vomiting, and they see coloured haloes around lights. Examination reveals a fixed, mid-dilated pupil, hazy cornea and firm globe. The iris is mid-dilated because this is where it can be in greatest contact with the lens to cause pupillary blockage and as the pressure rises, it becomes ischemic and remains mid-dilated. Immediate medical treatment with topical pupillary constrictors, intravenous osmotic agents and oral carbonic anhydrase inhibitors is required, followed by a laser iridotomy to allow the aqueous to bypass the pupillary blockage.

Summary

Examination of the optic nerve and understanding its disorders are of great importance to the general physician, as misdiagnosis of optic nerve pathology can lead to permanent visual loss. Table 1 summarizes important differences between the optic nerve disorders that one will encounter as a general physician.

References