Retrobulbar Optic Neuropathy: “Neither the Patient nor the Doctor Can See”

Abstract

Retrobulbar optic neuropathy (RBON) is a characterized by normal optic disc appearance and specific visual symptoms. As the injury site of the pathologic process in this optic neuropathy (ON) is behind the optic nerve head, in the other words, behind the lamina cribrosa, or in the intra-orbital or intra-canalicular or intracranial parts of the optic nerve, the optic disc seems normally. RBON may occur as retro-bulbar optic neuritis, traumatic ON, toxic ON, posterior ischemic ON, infiltrative ON, compressive ON, radiation-induced ON or hereditary ON with the classic phrase “neither the patient nor the doctor does not see”. Eventually, OD atrophy may develop following almost all types of optic neuropathy.

Unilateral RBON include RON, CON and infiltrative optic neuropathy related with neoplastic diseases such as leukemia, multiple myeloma or lymphoma while TxONs including tobacco-alcohol induced optic neuropathy, nutritional optic neuropathy (vitamin B12 and folate deficiency), drug-induced optic neuropathy, and Leber’s hereditary optic neuropathy (LHON) can cause the appearance of bilateral normal ODs [3-6].

Retrobulbar Optic Neuropathies

A list of the RBONs and their typical OD appearances were given in Table 1.

Retrobulbar optic neuritis

Retrobulbar optic neuritis (RON) is mainly caused by multiple sclerosis, a common demyelinating disease. The cardinal signs of RON are the loss including visual acuity or/and contrast sensitivity, periorcular pain induced with ocular movements, RAPD and CVD. OD has a normal appearance in 65-75% of cases with RON in initial stages and it is edematous in the rest [8,9].

Traumatic optic neuropathy

Traumatic optic neuropathy (TON) is an important cause severe visual loss occurring following blunt or penetrating head or ocular trauma. TON is usually diagnosed based on the clinical, ophthalmoscopic, radiologic examination findings and trauma story. Ocular examination shows a RAPD except in bilateral symmetric cases, variable loss of visual acuity, CVD, and variable VF defects. TON is classified as direct or indirect. Direct TON is caused by a direct penetrating damage to the optic nerve resulting a severe visual loss. Indirect TON is usually caused by acceleration/deceleration forces due to the blunt head or closed globe trauma. Although TON can occur in any region such as intraocular, intra-orbital, intracranial and chiasmal parts of the optic nerve, it
usually occurs at the intra-canalicul part of the optic nerve (inside sphenoidal optic canal). Ophthalmoscopic examination reveals the OD with a normal appearance at the presentation and initial stages especially in the cases with indirect posterior appearance without edema as the injury is in the retrobulbar optic nerve. However, an OD pallor will develop in about 3–6 weeks following orbital or cranial trauma [10,11].

### Toxic optic neuropathy

Toxic optic neuropathy (TxON) is resulted from toxicity of nutrients, toxins, drugs or chemotherapeutic agents such as ethambutol, ciprofloxacin, isoniazid, amiodarone, lizenzol, methotrexate, sildenafil, oxymetazoline, infliximab, vincristine, carboplatin, paclitaxel, cyclosporine, cisplatin and chloramphenicol, carbon monoxide, ethylene glycol, perchloroethylene, methanol, phosphodiesterase 5 inhibitors, tumor necrosis factor alpha inhibitors and tobacco, and metabolic or nutritional problems (vitamin B12 and folate deficiency). TxON occurs usually as a chronic, slowly progressive optic nerve dysfunction with bilateral and simultaneous involvement resulting early CVD (red-green), the loss of visual acuity and centrocaecal scotoma. Although TxON may cause mild and bilateral OD edema and sometimes OD hemorrhages, ODs usually appear normal in the early stages of TxONs. In the later stages, optic atrophy, most commonly in the temporal OD region develops [12-15].

### Posterior ischemic optic neuropathy

Posterior ischemic optic neuropathy (PION) is a rare form of ischemic optic neuropathies. PION is usually caused by an acute reduction of the blood flow to the retrobulbar optic nerve and acute ischemic damage. It should be considered in the diagnosis in each patient with acute painless vision loss, a RAPD or sluggish pupillary response, and a normal-appearing OD in one eye. PION is classified as surgical (postoperative or perioperative), arteritic (due to temporal arteritis) or non-arteritic. If rarely, bilateral PION occurs, it should be considered that the undergoing cardiac or spinal surgery may be in the etiology. In the etiopathogenesis, it has been considered that some risk factors including anemia, hypotension, hemodialysis, severe blood loss during the surgery, surgeries longer than 6.5 hours such as the spine, cardiac, head–neck surgeries from prone positions, ocular surgery, carotid atherosclerosis, and diabetes. The OD has initially normal appearance without edema as the injury is in the retrobulbar optic nerve. However, an OD pallor will develop within 4–6 weeks of the initial injury [3,4,16,17].

### Infiltrative optic neuropathy

Infiltrative optic neuropathy is an optic neuropathy caused by the infiltration of neoplastic or inflammatory cells of the optic nerve in diseases such as leukemia, lymphoma, or any neoplastic disease (multiple myeloma, malignant glioma, breast carcinoma, lung carcinoma), granulomatous inflammation from sarcoidosis, syphilis, TB, and fungal infections. The patients experience gradual and progressive loss of visual acuity in days or weeks. OD may be observed as normal or swollen in appearance at initial presentation. If a retrobulbar involvement is present, OD will have normal appearance. CVD, VF defect and RAPD may be detected or the patient may be asymptomatic [18-24].

### Radiation-induced optic neuropathy

Radiation-induced optic neuropathy (RION) is a late complication due to the necrosis of the anterior visual pathway, the brain or orbit following radiotherapy. RION presents with acute, painless, visual loss (involving visual acuity and VF) usually three years (mean 1-1.5 years) after completion of radiation exposure. It may cause a severe and irreversible visual loss and the second eye may be involved within weeks or months. The diagnosis is performed with the evidence of impaired visual function, a story of the radiation exposure and, the absence of another optic neuropathy causes. As RION is a result of the retrobulbar ischemic process and the endothelial cell injury from radiation, OD usually seems as normal with ophthalmoscopy in the acute phase, however, it can be observed as swollen [3,4,25].

### Leber’s hereditary optic neuropathy

Leber’s hereditary optic neuropathy (LHON) is an inherited optic neuropathy from the mutation in maternal mitochondrial
DNA. LHON affects predominantly affects men and presents with acute, severe, painless and usually irreversible visual loss (including central scotoma and color vision deficiency). The other eye is usually involved within weeks or months after the first eye. Funduscopic examination usually reveals a circum-papillary telangiectasia with OD hyperemia. Approximately 20-33% of the patients with LHON show a normal-appearing disc in initial stage. The classic triad of LHON is circum-papillary telangiectasia, swelling of peripapillary nerve fiber layer and the absence of leakage from the disc on fluorescein angiography. After the telangiectasia and nerve fiber layer swelling disappear, eventually, OD pallor appears in especially in the temporal area with coexistent damage of the papilla-macular nerve fiber layer [3-6,26,27].

Compressive optic neuropathy

Compressive optic neuropathy (CON) is commonly caused by direct compression to the intraorbital, intracranial, or pre-chiasmal optic nerves by a mass such as tumor, aneurysm enlarged extraocular muscles. The common causes of CON are orbital and intracranial meningioma, pituitary adenomas, intracranial aneurysm (involving internal carotid artery or anterior cerebral artery), craniosynostosis, glialoma in the anterior visual pathway, metastatic carcinomas such as glioma, meningioma, astrocytoma, hemangiomas, lymphangioma, teratoma, lymphoma, sarcoma, multiple myeloma, nasopharyngeal carcinoma. However, it may also be caused by sinus mucocoele, sphenoid creid and olfactory groove meningioma, tuberculoma, cryptococcal disease, sarcoidosis, thyroid eye disease and mass lesions in intracranial area or in saddle. Anterior intraorbital lesions often cause OD edema whereas intracranial, intracanalicular, and posterior orbital lesions usually cannot cause OD edema. Especially, a half of the patients with CON caused by thyroid eye disease may have normal appearing ODs. CON usually courses gradual and progressive visual loss. However, the acute and severe visual loss can develop in case of pituitary apoplexy, or ruptured aneurysm. VF defects occur as concordance with the localization of the lesion [1-6,18,28-30].

Stage 0 Papilledema

Papilledema is optic disc swelling due to elevated intracranial pressure. Optic nerve damage develops due to intraneuronal ischemia secondary to the stasis in the axoplasmic flow. Main causes of papilledema include intracerebral tumors, cerebral haemorrhage, meningitis, cranial trauma, hydrocephalus, impairment of the circle of cerebrospinal fluid and idiopathic intracranial hypertension. In the acute stage of this optic neuropathy, in contrast to others which OD edema is present, vision is usually well preserved. Although established papilledema presents with bilateral OD edema, retinal and peripapillary haemorrhages, peripapillary cotton wool spots, vascular alterations in OD, in Stage 0 papilledema OD seems normal or blurring of nasal, superior and inferior poles [7,31,32].

Conflict of Interest

The author declares that there is no conflict of interest regarding the publication of this paper.

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References


