Cranial Nerve II: Optic Nerve

6

One sentence: The optic nerve is considered a part of the brain and serves the special sense of vision and is essential for visual reflexes.

Genetic testing	NCV/ EMG	Laboratory	Imaging	Clinical testing
+	+ EP	+	+ MR, US (partly)_	+

Symptoms

Variable loss of vision, ranging to blindness.

Signs

The major function is the reflex reaction of the pupil, directly and indirectly.

Specific Qualities

Motor:

Sensory:

Autonomic:

Special senses: Information, such as brightness and color perception and contrast (visual acuity). Reflex pathways for lightning and accommodation. Visual field defects.

Other:

Location of Lesions: (Fig. 6.1)

Lesions of the optic nerve can be divided into three categories:

- Anterior to the chiasm (monocular field defect or blindness).
- Medial and temporal compression of the chiasm (hemianopia).
- Posterior to the chiasm.

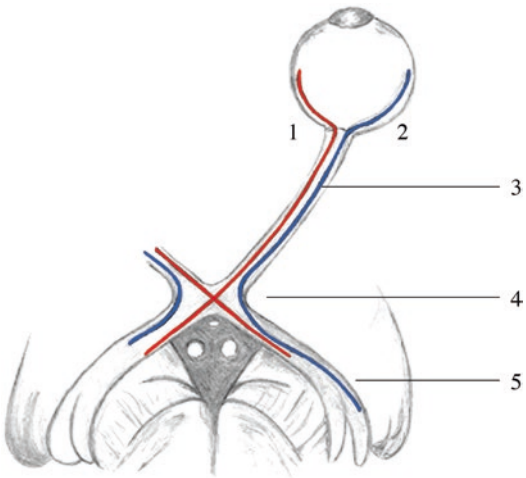

Fig. 6.1 Optic nerve. (*1*) medial fibers, (*2*) lateral fibers, (*3*) optic nerve, (*4*) optic chiasm, (*5*) optic tract

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Central:

Tumors: glioma, infarction; multiple sclerosis and others.

Most axons of the optic nerve terminate at the lateral geniculate body.

From there, they relay to the visual cortex in the occipital lobe. Other fibers relate to the pretectal area (for reflexes) and the suprachiasmatic nucleus.

Intracranial within the skull:

Prechiasmatic lesions.

Chiasmatic lesions [1]: Aneurysm, craniopharyngioma, meningioma, optic glioma, pituitary adenoma, Rathke cleft cyst.

Retrochiasmatic pathways: [2].

Type of lesion: Compression, infiltration, arachnoid thickening (e.g., tabes dorsalis) [3].

Exit of the skull:

Optic canal.

Cavernous hemangioma [4], cysticercosis [5], meningioma [6], mucocele, orbital fasciitis resulting in visual loss, proptosis, and visual defects.

Neoplasm: Optic glioma, cancer [7].

Pressure: e.g., acute in trauma [8].

Outside of the skull: Within the orbit, e.g., neoplasm or other space-occupying lesion.

For practical purposes, the length of the parts of the optic nerve can be assumed as noted in Table 6.1, and the vascular supply of the optic nerve is as noted in Table 6.2.

Table 6.1 Length of optic nerve parts in adults

	Length (mm)	Location
Intracranial	14–16	From the chiasm to the optic nerve canal
Intracanalicular	9	Optic nerve channel
Intraorbital	25	Intraorbital
Intraocular	1	Optic head

Table 6.2 Vascular supply of the optic nerve

Optic nerve	Proximal part: ophthalmic artery Distal part: small branches from internal carotid artery (ICA) and anterior carotid artery (ACA)
Optic chiasm	Superior: perforators from ACA Interior part: ICA, posterior communicating cerebral artery (PCommA), posterior cerebral artery (PCA)
Optic tract	PCommA, PCA, anterior choroidal artery

Combination with Other CN

Frontal tumors, trauma.

Causes and Frequency

Compression: Apoplexy of the pituitary (associated with headache), carotid aneurysm, endocrine orbitopathy. *Tumors in the sella result in visual field defects and a swollen optic disc.* Compression occurs in 50% of pituitary adenomas. Other causes include craniopharyngioma (in childhood), meningioma of the tuberculum sellae, aneurysm, and tumors of the chiasm itself (e.g., meningioma, neurinoma, or retinoblastoma).

Hereditary: Friedreich's ataxia, Leber's hereditary optic neuropathy, lysosomal disease, mitochondrial myopathy, Kearns-Sayre syndrome, neuropathy, ataxia, retinitis pigmentosa (NARP), storage disease (Tay-Sachs disease), optic atrophy 1, spinocerebellar disease. The optic nerve can also be damaged in genetic neuropathies: Autosomal dominant optic atrophy with cataract (ADOAC), cerebral dysgenesis-neuropathy-ichthyosis-keratoderma syndrome (CEDNIK) [9], Charcot-Marie-Tooth disease type 4 (CMT4; HMSN VI), *OPA1* and *OPA3* mutations.

Iatrogenic: Pressure on the eye bulb caused by anesthesia (ischemic optic nerve neuropathy), blepharoplasty, fractures of the orbit, or surgery of the nasal sinus.

Infectious: Meningitis, sarcoid, syphilis, tuberculosis. *Focal*: Granulomatous disease, orbital tumors, sinusitis. Chronic sinusitis [10].

Inflammatory: Optochiasmatic arachnoiditis.

Immune mediated: Optic neuritis in Devic's syndrome and multiple sclerosis, aquaporin 4 [11].

Metabolic: Diabetes, thyrotoxicosis, uremia.

Nutritive: Alcohol ingestion, B1 deficiency, B12 anemia, Cuban neuropathy, folic acid, methylol toxicity, Strachan's syndrome.

Paraneoplastic: Rarely involved in paraneoplastic dysfunction – carcinomatous retinopathy (CRMP5 and CAR) antibodies [12].

Radiation: Radiation therapy of brain tumors, pituitary tumors, metastases, or ENT tumors can cause unilateral or bilateral loss of vision with long latencies. Progressive optic nerve atrophy is seen within 6 weeks of exposure to 70 Gy.

Toxic optic neuropathy*:*

Alcohol: Methyl alcohol [13].

Drugs: Table 6.3.

Other causes: Heavy metals (arsenic, lead, mercury, thallium), aniline dye, carbon monoxide, carbon tetrachloride, tobacco [14], nitrous oxide.

Ethambutol: Color perception [15].

Trauma: Several mechanisms have been implicated:

- "Blowout" fractures, gunshot wounds, penetrating trauma, trauma of the orbit, traumatic optic neuropathy. Directly by objects: Penetrating trauma, transaction, avulsion, bleeding, air and gases.
- Indirect trauma (blow combined with commotion or concussion).
- Lesion in the intracanalicular part: Contusion of nerve axons and edema.

Tumors: Metastasis, melanocytoma, meningeal carcinomatosis (Fig. 6.2), nasopharyngeal tumor compresses the nerve and chiasm, neurofibromatosis (NF1, NF2), orbital tumors, optic nerve glioma, retinal infiltration (leukemia).

*Vascular:*Vascular diseases of the optic nerve: [16].

Aneurysms, giant cell arteritis, herpes zoster, ischemic optic neuropathy retrobulbar optic neuropathy, systematic lupus, temporal arteritis.

Table 6.3 Drugs causing optic nerve toxicity (see also toxic)

Drug class	Drugs
Antibiotics	Chloramphenicol, ethambutol, isoniazid, linezolid, streptomycin sulfonamides
Anticancer drugs	Chlorambucil, methotrexate, tamoxifen vincristine
Antimalarial drugs	Chloroquine, hydroxychloroquine, quinine
Antiarrhythmics	Amiodarone, digitalis
Phosphodiesterase inhibitors	Sildenafil

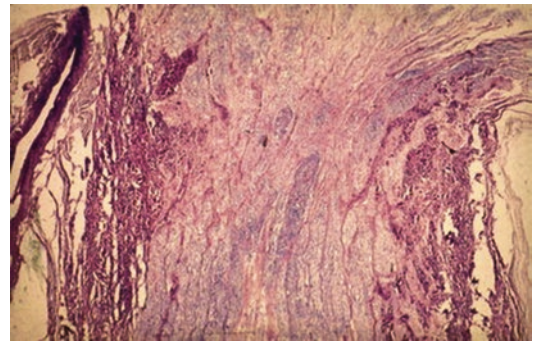

Fig. 6.2 Optic neuropathy. A photomicrograph of an optic nerve that is compressed by tumor cells ("cuffed") in meningeal carcinomatosis, resulting in blindness of the patient

Ischemia: Anterior ischemic optic neuropathy (AION), nonarteritic (NAION) and arteritic (AAION) forms.

Main Investigations

Diagnosis is based on clinical test, on X-ray, CT, or MR imaging, optical coherence tomography, visual function and color discrimination tests, ophthalmoscopic exam, visual evoked potentials, and electroretinogram.

Special ultrasound techniques also allow a partial identification of the infraorbital optic nerve.

Differential diagnosis: Other causes of papilledema need to be considered, including increased intracranial pressure and pseudotumor cerebri.

Therapy

Treatment depends upon the cause of the lesion.

Prognosis is presently subject to research [17].

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