

Definition

► Epidemiology

Prevalence of 3:100 000 • First presenting symptom of demyelinating disease in 12–30% of cases • Approximately 50% incidence of bilateral visual impairment.

► Etiology, pathophysiology, pathogenesis

Acute inflammation of the optic nerve (CN II) • Autoimmune diseases (e.g., systemic lupus erythematosus, disseminated encephalomyelitis) • Parainfectious or viral etiology (e.g., cytomegalovirus, rubella, mumps, herpes, toxoplasmosis) • Radiation-induced (exposure of approximately 10 Gy or more).

Imaging Signs

► Modality of choice

Gadolinium-enhanced MRI.

► CT findings

CT often shows no abnormalities • Possible thickening of the optic nerve • Nerve may enhance after contrast administration.

► MRI findings

Intraorbital and intracanalicular thickening of the optic nerve • Mixed punctate and streaky enhancement after gadolinium administration (especially of the intracanalicular nerve) • Increased T2-weighted signal intensity • Sequences with combined fat and water suppression (SPIR FLAIR) are more sensitive for detecting optic nerve lesions.

► Pathognomonic findings

Thickened optic nerve showing enhancement after gadolinium administration on T1-weighted fat-suppressed imaging.

Clinical Aspects

► Typical presentation

Viral: Visual deterioration 10–14 days after underlying disease • Central scotoma • Afferent pupillary defect.

► Treatment options

Steroid therapy • Interferon is given for disseminated encephalomyelitis.

► Course and prognosis

Unilateral optic neuritis has a good prognosis with cortisone therapy • Visual impairment persists in up to 15% of cases, depending on the underlying disease • Recurrence rate approximately 20%.

► What does the clinician want to know?

Diagnosis • Intracerebral foci • Exclusion of a mass.

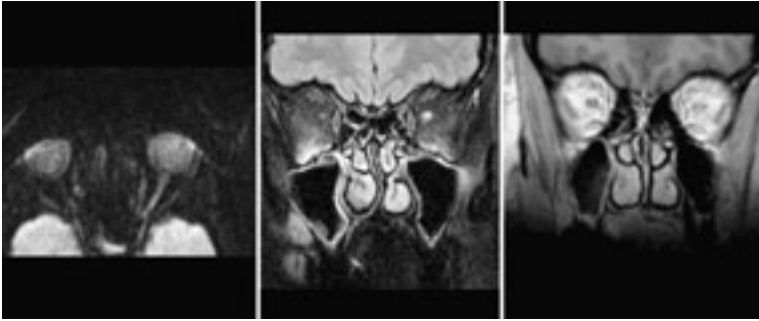


Fig. 3.6 Left-sided optic neuritis as an initial manifestation of multiple sclerosis. Axial diffusion-weighted image (left) and coronal T2-weighted MR image (center) with fat and water suppression (SPIR FLAIR) show increased signal intensity of the optic nerve. Post-contrast coronal T1-weighted image (right) shows marked enhancement.

Differential Diagnosis

<i>Mass (e.g., optic glioma, meningioma)</i>	– Circumscribed optic nerve expansion or mass, enhancing after contrast injection
<i>Orbital pseudotumor</i>	– Pain – May involve all orbital structures
<i>Radiation neuropathy</i>	– Rare – Prior history of radiotherapy

Tips and Pitfalls

Cerebral imaging should be done to exclude a demyelinating disease.

Selected References

- Hickman SJ. Optic nerve imaging in multiple sclerosis. *J Neuroimaging* 2007; 17(S1): 42S–45S
- Jackson A et al. Combined fat- and water-suppressed MR imaging of orbital tumors. *AJNR Am J Neuroradiol* 1999; 20(10): 1963–1969
- Müller-Forell W et al. Entzündliche Orbitaerkrankungen. Teil 2: Bulbus, Extrakonalraum, Glandula lacrimalis, Nervus opticus. *Radiologe* 2003; 43(5): 400–418
- Rocca MA et al. Imaging the optic nerve in multiple sclerosis. *Mult Scler* 2005; 11: 537–541