pupillary defect is seen in unilateral optic neuritis. Acute demyelinating optic neuritis is often retrobulbar without papillitis. On MRI, the optic nerve can be swollen and enhance after gadolinium contrast. After recovery from the acute episode, the optic disk may appear pale, and the relative afferent pupillary defect may persist. Transient worsening of vision when the body temperature rises due to exercise or fever (Uhthoff's phenomenon) may occur following recovery. The differential diagnosis includes other causes of acute monocular or binocular vision loss, such as Leber's hereditary optic neuropathy, giant cell arteritis, and acute non-arteritic anterior ischemic optic neuropathy.

The Optic Neuritis Treatment Trial studied patients with acute optic neuritis (either idiopathic or due to MS) who were randomized to one of three treatments, intravenous methylprednisolone versus oral prednisone taper versus oral placebo. Visual acuity initially recovered faster in the intravenous methylprednisolone group, but by 6 months later there was no difference among the three groups (level A). Recovery of vision was good. Patients from this trial were examined 10 years later and acuity in the affected eyes was 20/20 or better in 74% and less than 20/200 in only 3%. However, recurrence of optic neuritis was common, and had occurred in either eye in 35% of the patients. Recurrences were more frequent in those who had MS than those with idiopathic optic neuritis ($P < .001$).

CHRONIC RELAPSING INFLAMMATORY OPTIC NEUROPATHY

First described in 2003, chronic relapsing inflammatory optic neuropathy (CRION) is an inflammatory optic neuropathy characterized by acute relapses, but often with more severe visual loss than idiopathic optic neuritis or optic neuritis associated with MS. Patients with CRION can have onset at any age and the entity has been described worldwide. Prevalence rates and epidemiology are still unclear. As in other types of optic neuritis, eye pain at onset is frequent. Return of pain can herald a relapse. Uveitis is present in a small percentage of cases. Five diagnostic criteria for CRION have been suggested: (1) optic neuritis with at least one relapse, (2) objective visual loss, (3) AQP4-IgG seronegative, (4) contrast enhancement on MRI of acutely inflamed optic nerve, and (5) response to immunosuppressive treatment and relapse on withdrawal. Other diseases that might present similarly, such as sarcoidosis and giant cell arteritis, should be ruled out. Treatment for acute CRION is similar to that for other causes of optic neuritis, using intravenous methylprednisolone followed by oral corticosteroids. Relapses are common upon corticosteroid discontinuation. Successful long-term treatment with "steroid sparing" agents such as methotrexate, azathioprine, or mycophenolate mofetil has been reported. The underlying pathology is not yet known, but the disease appears inflammatory based on clinical presentation, imaging, and specific medication response. Eventual visual outcomes are often poor. One report indicated that visual acuity was less than 20/200 in one third of CRION patients.

SUGGESTED READINGS

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