

Chronic relapsing inflammatory optic neuropathy (CRION)

Zarvani, Ashraf

inflammatory optic neuropathy

Chronic relapsing inflammatory optic neuropathy (CRION) is a recently described recurrent optic neuropathy which is steroid responsive. Several features distinguish this entity from optic neuritis associated with demyelinating disorders and connective tissue diseases. The severe degree of visual loss, persistence of pain after onset of visual loss, and recurrent episodes are unique to this disorder.

The syndrome comprising subacute visual loss, pain, and a clear and early response to systemic steroids is easily identifiable as an inflammatory optic neuropathy. Chronic relapsing inflammatory optic neuropathy (CRION) is in fact a form of isolated recurrent optic neuropathy.

Diagnosis of CRION requires exclusion of other neurological, ophthalmological, and systemic conditions.

Early recognition of patients suffering from CRION is relevant because of the associated risk for blindness if treated inappropriately.

Diagnostic Criteria:

1. History: ON and at least one relapse
2. Clinical: Objective evidence for loss of visual function
3. Labor: NMO-IgG seronegative
4. Imaging: Contrast enhancement of the acutely inflamed optic nerves.
5. Treatment: Response to immunosuppressive treatment and relapse on withdrawal or dose reduction of immunosuppressive treatment.

The treatment consists of three phases:

- (1) restoring visual function in the acute phase
- (2) finding a strategy to stabilize vision on the interim
- (3) preserving vision on the long term with minimal treatment side effects.

CRION represents a severe optic neuropathy. CRION should be recognized early because there is a considerable risk of severe, potentially blinding loss of vision for the patient.