AMERICAN ACADEMY OF OPHTHALMOLOGY
The Eye M.D. Association

OPTIC NEURITIS

I. Describe the approach to establishing the diagnosis

A. Describe the etiology of this disease
1. Inflammatory demyelination of the optic nerve, either idiopathic or related to multiple sclerosis (MS)

B. Define the relevant aspects of epidemiology of the disease
1. Typically age 20-50 years, but cases may occur outside this range
2. Approximately 75% female (parallels MS epidemiology)

C. List the pertinent elements of the history
1. Acute visual loss (typically progresses over less than 7 days)
2. Typically monocular
3. Majority have peri-orbital pain initially, often increased with eye movement (resolves in week 1-2)
4. May have history of demyelinating symptoms or known diagnosis of multiple sclerosis

D. Describe pertinent clinical features
1. Decreased visual acuity
2. Decreased color vision (often out of proportion to acuity loss)
3. Visual field defect (diffuse, altitudinal, cecocentral or other)
4. Relative afferent pupillary defect (if unilateral or asymmetric)
5. 2/3 normal disc appearance (retrobulbar), 1/3 disc edema (bulbar or papillitis); if present, disc edema is typically mild without hemorrhage or exudates

E. Describe appropriate diagnostic/laboratory testing
1. In typical case of optic neuritis with a known diagnosis of MS, no further testing may be required
2. In monosymptomatic or clinically isolated syndrome cases (1st episode of demyelination), MRI provides prognostic information
    a. MRI T2 hyperintensities suggestive of demyelination stratifies to higher MS risk (approximate risk at 5, 10, and 15 years is 50%, 60%, and 70%, respectively)
    b. Normal MRI stratifies to low MS risk (Approximate risk at 5, 10, and 15 years is 15%, 20%, and 25%, respectively)
        1) Normal MRI <u>with</u> the following features define extremely low MS-risk cohort (no MS cases at 15 years): painless optic neuritis, no light perception vision at onset, severe disc edema or disc hemorrhage, or macular star figure exudate

II. Define the risk factors

A. Female and Caucasian predominance
B. History of MS
C. Age range 20-50 years

III. List the differential diagnosis

A. Anterior ischemic optic neuropathy
B. Maculopathy with a normal appearing fundus
C. Infiltrative optic neuropathy
D. Compressive optic neuropathy
E. Other inflammatory optic neuropathy (e.g., sarcoid)
F. Infectious optic neuropathy (e.g., CMV, Lyme)
G. Neuroretinitis (e.g., Bartonella henselae 'cat scratch disease')
H. Hereditary (e.g., Leber hereditary optic neuropathy)
I. Miscellaneous other optic neuropathies

IV. Describe patient management in terms of treatment and follow-up

A. Acute treatment options: high dose corticosteroids (e.g., intravenous methylprednisolone), or no treatment depending upon individual risk-benefit ratio
    1. Optic Neuritis Treatment Trial* protocol: methylprednisolone 250 mg iv q 6 hours x 3 days, then prednisone 1 mg/kg/day po qd x 11 days, then 20 mg po on day 15 and 10 mg on days 16 and 18 then discontinue prednisone; many centers now administer methylprednisolone 1000 mg iv q day x 3 days for outpatient convenience with or without corticosteroid taper
B. Chronic treatment options: referral to a multiple sclerosis specialist for management and consideration of MS immunomodulatory therapy if abnormal MRI scan or other neurological abnormalities are suggested by examination or history
C. Oral prednisone 1 mg/kg/day alone <u>contraindicated</u> due to increased risk of recurrent optic neuritis (and no more effective than placebo concerning visual function)

V. List the complications of treatment, their prevention and management

A. Complications of corticosteroids
    1. Mood alteration including rare psychosis
    2. Hyperglycemia
    3. Insomnia
    4. Gastrointestinal irritation
    5. Osteoporosis or avascular necrosis (rare with short steroid courses)
    6. Weight gain

VI. Describe disease-related complications

A. Persistent decreased visual acuity (only 5% of patients have visual acuity worse than 20/40 at 6 months)
B. Persistent dyschromatopsia, decreased contrast sensitivity and impaired depth or motion perception
C. Progression to MS

VII. Describe appropriate patient instructions

A. Discuss risk, benefits, side effects and alternatives of high dose corticosteroid course
    1. Institute and monitor therapy if appropriate

B. Obtain MRI of brain (consider orbital images) with and without gadolinium in appropriate cases
    1. If MRI reveals T2 hyperintensities consistent with MS
        a. Discuss potential relationship to MS, possible MS therapies, or refer to MS neurologist

C. Monitor for atypical course or features
    1. Report new neurological symptoms
    2. Macular star figure emergence
    3. Failure to improve
    4. Treatment side effects

*** Results of the Optic Neuritis Treatment Trial:** High dose IV methylprednisolone given to patients first seen within 8 days of symptom onset with unilateral vision loss in an eye that had not had optic neuritis had quicker visual recovery but at one month, had no significant difference in VA, visual fields, color vision, or contrast sensitivity. Oral prednisone treatment alone did not improve visual outcomes and was associated with a higher rate of recurrence.

Additional Resources

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