CURRENT NEURO-OPHTHALMOLOGY

SIMMONS LESSELL
J. T. W. VAN DALEN

VOLUME 2

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Preface

The purpose of *Current Neuro-ophthalmology* is to provide an in depth, biannual review of the neuro-ophthalmology literature from the past 2 years. Neuro-ophthalmology topics will be reviewed by experts in their fields; each chapter has a wealth of references.

The series started in 1980. Three volumes were published by Elsevier (1980-1984) before publication was taken over by Year Book Medical Publishers. Under their aegis, the first review, *Current Neuro-ophthalmology*, was published in the fall of 1987. The present publication, therefore, is the fifth review of the neuro-ophthalmology world literature. It is remarkable that many of the original contributing reviewers are still willing to take this heavy review task on their shoulders. The proliferation of the neuro-ophthalmology literature is still continually making review an almost impossible task. As editors, we are thankful that we have such an excellent group of experts who have done such a tremendously good job.

One of the most profound changes in the world of neuro-ophthalmology was the advent of neural imaging techniques. Computed tomography (CT) was introduced in 1972 and the clinical use of magnetic resonance imaging (MRI) is spreading rapidly.

Chapters on computed tomography (Drs. M.L. Leib and M.R. Kates) and on neuroradiology (Dr. J.S. Leo et al.) describe threse methods in detail. It should not be surprising however that CT and MRI play a big role in other chapters, too. Other areas that have received much attention in the recent neuro-ophthalmology literature are reviewed, such as Alzheimer's disease (Dr. J.L. Cummings), botulinum toxin therapy (Dr. P.A. Sibony) and stroke-carotid artery endarterectomy (Dr. J.D. Trobe). Also the more classic neuro-ophthalmology entities such as anterior ischemic optic neuropathy (Dr. M.M. Behrens), migraine therapy (Dr. B.T.

Troost) and myasthenia gravis therapy (Dr. D. Schmidt) are discussed at length. Concerning nystagmus-nestor, Dr. L. Dell'Osso has "discovered" two new types of nystagmus. His list now contains 45 different types of nystagmus and 16 saccadic oscillations/intrusions.

These post-publication peer reviews of the neuro-ophthalmology literature give you a current assessment of the present status of neuro-ophthalmology. We hope that this book will be of benefit to you and to your patients.

Simmons Lessell, M.D.
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Contents

PART ONE

THE VISUAL SYSTEM

CHAPTER 1

Optic Neuritis

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DIFFERENTIAL DIAGNOSIS OF OPTIC NEURITIS

Neuroretinitis

One form of optic neuritis (ON) needs to be distinguished from idiopathic or demyelinating ON: neuroretinitis, an inflammatory (vascular?) condition of the optic disk and retina characterized by optic disk swelling and the development of a "macular star figure" in the outer plexiform layer of the retina around the macula by lipid exudates derived from leaking optic disk capillaries. Parmley et al.[1] studied 50 patients with neuroretinitis and followed up 10 prospectively for 1 to 2 years and 13 retrospectively for a mean of 8 years. On follow-up, no patient with neuroretinitis either had developed symptoms or signs of multiple sclerosis (MS) or had oligoclonal bands in the cerebrospinal fluid (CSF). Thus, the clinical sign of a macular star in the presence of ON or disk swelling ("papillitis") distinguishes neuroretinitis from the ON of MS and carries an excellent prognosis in comparison with demyelinating ON. A number of variations occur in the presentation of neuroretinitis: in particular, the macular star may not develop until 1 or 2 weeks after the initial symptoms of visual loss. Therefore, Parmley et al. suggest that patients who have signs and symptoms of ON without a macular star should be reexamined after 2 weeks for the development of such a star, with its consequent improvement in prognosis.

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Infective-Parainfective ON

Jones et al.[2] reported severe ON as the only prominant finding in a 61-year-old man with infectious mononucleosis (Epstein-Barr virus). Visual function recovered nearly completely. The authors also discussed seven other well-documented cases of ON associated with infectious mononucleosis. Purvin et al.[3] described chiasmal ON associated with Epstein-Barr virus infection in a 13-year-old boy, again with good recovery of vision. A 14-year-old girl developed severe bilateral ON, 1 week after a typical varicella vesicular eruption.[4] Visual loss was rapid and accompanied by photopsia and eye pain and proceeded to total blindness. Visual recovery in one eye was very poor, contrary to previous reports,[5, 6] and the authors speculate as to whether treatment with steroids may have *worsened* the visual prognosis. Tunis and Tapert[7] reported a case of unilateral ON occurring 24 days after an episode of herpes zoster ophthalmicus, with the usual poor visual recovery. A useful literature review accompanies this report. Weber and Mikulis[8] reviewed inflammatory disorders of the paranasal sinuses that can cause ON, with emphasis on the diagnostic use of the computed tomographic (CT) scan. Holder et al.[9] described a case of malignant external otitis in a diabetic man that progressed to cause multiple cranial nerve palsies and finally sudden, total monocular visual loss. In a culture from a biopsy of the involved optic nerve, *Pseudomonas aeruginosa* grew.

Margo et al.[10] reported severe unilateral visual loss and neuroretinitis associated with the systemic symptoms of visceral larva migrans. Markedly elevated serum enzyme-linked immunosorbent assay titers for *Toxocara canis* were useful in making the diagnosis, but the visual loss did not respond to treatment with thiabendazole. Farris and Webb[11] reported a classic picture of unilateral ON that developed 15 days after completion of "adequate" antibiotic treatment for Lyme disease *(Borrelia burgdorferi)*. Initial visual loss was severe (light perception), and recovery over several months was incomplete. The patient's initial presentation with Lyme disease had included fever and rash, followed by aseptic meningitis, radicular sensory neuropathy, transverse myelitis, and partial bilateral sixth-nerve palsies.

Pall and Williams[12] reported two complex cases in which ON and hearing loss occurred in association with the Guillain-Barré syndrome, following an otherwise trivial influenzalike illness. In the first patient, ON occurred sequentially in each eye and progressed to total blindness. The second patient had complete unilateral loss of vision. The appearance of the optic disks was normal in the acute stages of visual loss, but optic atrophy later supervened. Auditory function recovered rapidly and completely in both patients. Visual function remained at no light perception for 3 months in the first patient before slowly recovering to 6/9 bilaterally. The second patient remained completely blind in the involved eye. Steroid therapy did not appear to halt progression or accelerate recovery of visual loss in either patient. These two cases are also important in documenting combined involvement of both central and peripheral nervous systems in the Guillain-Barré syndrome.