Neuro-ophthalmology Focus 1982

J. Lawton Smith, M.D.

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PREFACE

Neuro-ophthalmology as a discipline continues to grow by leaps and bounds. It would have been hard to foresee this years ago, for Dr. Walsh really started this field by himself in the United States in the 1930s, and when Dr. Bill Hoyt and I became interested in it in the 1950s, there were only about seven physicians in the world, who to our knowledge, limited their practices to neuro-ophthalmology. At the 1981 Frank B. Walsh Society meeting in Houston, Texas, however, there were more than 100 physicians in attendance. If one stops to consider that about 5% of an ophthalmological practice is surgical—and that is said to be an average figure—that if one stops and adds up all the cases of systemic disease, abnormal pupils, funny looking optic discs, visual field problems, instances of proptosis, and mystery motility cases that go through the average ophthalmologist's office, it could well be 15–20% of the practice. In other words, there is no doubt that the percentage of one's practice that relates to neuro-ophthalmology is at least three to four times as large as the percentage that is strictly surgical.

However, the practitioner today is caught on all hands by many, many pressures. Demands for his or her time continue to challenge all reasonable priorities. This volume is an attempt to not only present some current discussions on timely neuro-ophthalmologic topics, but I have continued the suggestion given by Dr. Costin, which was used in the previous volume. Thus, at the end of each chapter (with the only exceptions being those in which I have helped write the chapter) an editorial comment has been attached. The purpose of this is to help someone, who may have had little previous experience with the topic or who may well have forgotten all they knew about it before, "relate" to the material. In addition to these volumes, other efforts are now being made to help the practitioner. A small quarterly journal, The Journal of Clinical Neuro-ophthalmology, has started in 1981 and is published by Masson Publishing, Inc., who also kindly prints this volume. An audiocassette journal, Neuro-ophthalmology Tapes, is available to subscribers who like to listen to tapes while they ride in the car to work. In these works, we try to delete abbreviations, use prescription names of drugs, use "20/20" visions rather than "6/6," and, in general, attempt to help the practitioner. Many timely subjects are reviewed in this volume—optic nerve sheath meningiomas, the acute retinal necrosis syndrome, mitral valve prolapse and eye manifestations, pituitary apoplexy, and an update on cerebromacular degenerations, are simply a few. I sincerely hope this volume will help you and your patients, and would appreciate getting suggestions and constructive criticisms as to how such a volume could be improved—in case we decide to try again in another two years!

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FOREWORD

Ophthalmologists, neurologists, and neurosurgeons are not excepted from the tremendous pressures and shakings that are occurring in the world today. The literature explosion, rapidity of technologic change, fantastic upheavals in weather, governments, economies, and societies only serve to crowd and press man into the position of trying to cope while in a pressure cooker whose steam intensity becomes greater and greater. At the outset of this book, I should simply like to point out to the reader that I have found by experience that there truly is a way out. It is not a mental mechanism, not a philosophy, not an intellectual experience—it is a *person*. The Lord Jesus Christ said (Matt. 11:28–30), "Come unto me, all ye that labour and are heavy laden, and I will give you rest. Take my yoke unto you, and learn of me; for I am meek and lowly in heart: and ye shall find rest unto your souls. For my yoke is easy, and my burden is light." In 1963, I prayed and sincerely invited the Lord Jesus Christ to come into my heart and to take over my life and to forgive me of my sins. I had already found that in medicine the truth is not defined by the majority vote. That is to say that the truth is not dependent upon the democratic process. There are tremendous swings of opinion in science and medicine, so that if one is not doing the operation in vogue at that time, he is quite out of it. Alas, however, two years later, we often find the pendulum has swung back again quite to the contrary! The Lord Jesus Christ said (John 14:6), ". . . I am the way, the truth, and the life: no man cometh unto the Father but by me." Thus, the Lord Jesus Christ made the bold faced statement that *He* is *the way*—and that *He* is *the truth*—and furthermore, that no man can come unto God the Father but by Jesus Christ. I submit that there are only three possible ways that such a statement can be analyzed. The originator of the statement was either mentally imbalanced, or was the greatest imposter the world has ever known, or was speaking the truth. Twenty centuries of history and personal experience of untold numbers of people have continued to confirm that the Holy Bible is true. I simply want to encourage physicians to spend a few minutes each day reading their Bibles and to find this out for themselves. I believe that reading the Bible will make you a better doctor and will also lead you to have a personal experience with the Lord.

In summary, we simply need to remember the following: "For God so loved the world, that He gave His only begotten Son, that whosoever believeth in Him should not perish, but have everlasting life. For God sent not His Son into the world to condemn the world; but that the world through Him might be saved." Therefore, doctor, the best prescription I can offer you, your family, and your patients is this (Rom. 10:9): "That if you shall confess with your mouth the Lord Jesus, and shall believe in your heart that God has raised Him from the dead, you shall be saved." If you will receive that pearl—the pearl of great price—then all the other things offered to you in this volume will fall into proper perspective. May the Lord bless you and yours is my prayer!

J.L.S.

A NEW PEARLS CHECK LIST

Doctor, if you are so busy and so pressured and so tired that you can't read anything—then you might enjoy simply going down the brief check list of references in this chapter. By no means is it all-inclusive, but there are a few points that might help you in your practice. We have attempted to bring you an overview of current information in clinical neuro-ophthalmology in this volume. You also might be interested to know that a new little quarterly journal—The Journal of Clinical Neuro-ophthalmology—just began in 1981. You can get information on that by writing to Masson Publishing Company, 133 East 58th Street, New York, New York 10022. You may also be interested in Neuro-ophthalmology Tapes, a monthly audio cassette teaching tape, and can obtain information on that by writing Neuro-ophthalmology Tapes, 9820 S.W. 62 Court, Miami, Florida 33156. Now, to mention a few pertinent pearls and references!

1. INTRAVENOUS "ARTERIOGRAPHY"!

I consider this the latest red hot pearl you will want to use in your practice! A paper by Dr. Maurice Ducos de Lahitte *et al.* outlined the technique in *Radiology* **137:** 705–711, Dec. 1980. These authors gave *intravenous* injections of contrast dye (using a 14-gauge needle and an automatic pressure injector), and then by using ordinary photographic *subtraction* techniques were able to visualize the great vessels in the neck with good results in 80% of a series of 500 patients. A new digital subtraction computer imaging of the great vessels in the neck is now available in a few centers, but requires about $200,000 worth of additional equipment in addition to your current computed tomography machine. However, after reading this paper, we asked our neuroradiologist, Dr. Quencer, to try this simple technique. We had a woman who had amaurosis fugax attacks in her right eye and also had transient ischemic attacks in her right extremities, who had previously had a normal *left* carotid arteriogram. It was obvious that we needed to see the right sided neck vessels, and she was studied with only an intravenous injection as an outpatient. Dr. Quencer performed double photographic subtraction of the pictures obtained, and they really turned out quite well! We could see that the carotid bifurcation was normal on the right—i.e., there was no surgical lesion there—but that rather high in her neck, the right internal carotid narrowed down to just about a thread. This told us that she was not a candidate for endarterectomy, but might well be considered a candidate for a right superficial temporal-middle cerebral artery bypass—and all that information came from an intravenous study as an outpatient. If that technique continues to be as promising as it looks now, it is really going to put most noninvasive tests (Doppler, oculoplethymosgraphy, infrared circulation studies and the like) out of business, as what we really want to do is actually look at the great vessels in the neck to see if there is surgical atheromatous disease there. This is exactly the test needed by the ophthalmologist—because you see that 60-year-old woman who has had two attacks of "blackouts" in her right eye, has a negative examination otherwise, and you do not want to subject her to an arteriogram. This technique can be done by your radiologist in any hospital that can now do an arteriogram, and you might want to have him or her review the paper cited in *Radiology,* Dec. 1980 and try a few cases for you—until you later have access to digital subtraction angiograms!

2. THE BASILAR BALLOON!

Dr. T. M. Sundt, Jr. and his Mayo Clinic colleagues wrote one of the most exciting papers I've seen in years. This was in *Mayo Clin. Proc.* **55:** 673–680, 1980. Two patients with high grade basilar artery stenotic lesions, and whose progressive symptoms did not respond to anticoagulant therapy, were treated by dilating up their basilar arteries by a balloon catheter technique! The results were really

impressive! Now the indications for such a procedure are terribly restricted, but you should know about this, as it offers something for *SOME* patients with basilar disease that we never had before. You can write Dr. T. M. Sundt at the Neurosurgical Section, Mayo Clinic, Rochester, Minnesota and get a reprint of that paper.

3. BACLOFEN IN TRIGEMINAL NEURALGIA

Dr. G. H. Fromm and associates found that baclofen (Lioresal) relieved 10 of 14 patients with refractory tic douloureux in *Arch. Neurol.* **37**(12): 768, Dec. 1980. If your patient with trigeminal neuralgia does not respond to Tegretol, you might consider a trial with Lioresal—or even a combination of the two medications.

4. BACLOFEN for PERIODIC ALTERNATING NYSTAGMUS

Lioresal is now considered the treatment of choice for periodic alternating nystagmus. See: *Ann. Neurol.* **8**(6): 609, Dec. 1980.

5. "ENDEP" FOR CLUSTER HEADACHES

A helpful medicine for cluster headaches is Endep (amitriptyline, Roche). In my experience "Endep" is better for cluster headaches, and "Elavil" is better for depression—even though both are amitriptyline! The name you write on the prescription blank is important.

6. The treatment to try for *subacute sclerosing panencephalitis* (Dawson's encephalitis) is amantadine (Symmetrel) and isoprinosine. See: *Ann. Neurol.* **8:** 422, 1980.

7. When you are doing a Tensilon test, or are thinking of myasthenia gravis, remember to ask if the patient has been taking Penicillamine. The drug is most commonly used for rheumatoid arthritis and Wilson's disease. It can give you a reversible myasthenic syndrome! See: *J.A.M.A.* **244**(16): 1822, Oct. 17, 1980.

8. The next time you see a patient with *Rieger's anomaly* (prominent Schwalbe's line with attached strands of iris, hypoplasia of iris stroma, and predisposition to juvenile glaucoma), you might consider getting a *lateral skull film.* Why? Dr. R. E. Kleinmann and associates reported "Primary empty sella and Rieger's anomaly of the anterior chamber of the eye—a familial syndrome" in *N. Engl. J. Med.* **304**(2): 90, Jan. 8, 1981. This can be an important point!

9. If you see a child with fractures, anemia, and suspected poor vision, order an *electroretinogram* (ERG). This may lead you to make the diagnosis of *osteopetrosis*! See: *Am. J. Dis. Child.* **133:** 955, Sept. 1979—by Dr. Craig Hoyt.

10. INDERAL for *NARCOLEPSY.* This was advocated in a letter to the editor in *Arch. Neurol.* **37:** 735, Nov. 1980.

11. TERSON'S SYNDROME is visual loss due to vitreous bleeding resulting from acute subarachnoid hemorrhage. These patients are ideal candidates for vitrectomy, if the process does not spontaneously clear within six months. You need to see Dr. John Clarkson's paper on this in *Am. J. Ophth.* **90**(4): 549–552, Oct. 1980.

12. Pseudotumor cerebri may rarely be *familial*! The next time you see a case look at the discs of family members, and remember that empty sella can run with this syndrome. See: *Ann. Ophth.* **12:** 1045, Sept. 1980.

13. When you see a patient with a history of *RETINAL EMBOLI,* remember to have them checked for *mitral valve prolapse.* That will require a careful auscultation of the chest for a midsystolic click and also ultrasound or echocardiography. A paper on this was in the *Am. J. Ophth.* **90:** 534, Oct. 1980.

14. Microadenomas were found in 27% of 120 autopsies (32 cases), whereas tomograms were abnormal in only 6 of the 32 subjects. An important paper was "Microadenomas of the pituitary and abnormal sellar tomograms in an unselected autopsy series" by G. N. Burrow, *et al.,* in *N. Engl. J. Med.* **304**(3): 156–158, Jan. 15, 1981.

15. A rare syndrome, but apparently one you can suspect clinically is as follows: A middle aged patient presents with vision loss mimicking optic neuritis and this rapidly progresses over 5–6 weeks to total blindness. Early symptoms are blurred vision, periorbital pain, unilateral disc edema and venous stasis retinopathy. The

patient dies in 6–9 months of a primary glioblastoma of the optic nerve. Another case was by Spoor *et al.* in *Arch. Neurol.* **38**(3): 196–197, March 1981.

Finally, our experience has continued to lead us to believe that many patients with primary optic nerve sheath meningiomas will definitely respond and be helped with irradiation therapy. In the first part of this volume, you will see papers on radiotherapy of primary optic nerve meningiomas contrasted with another chapter by Dr. Alper emphasizing a different approach to these lesions. These tumors have now been found to be surprisingly common, although we simply missed them for years before computed tomography, and the treatment is now in a state of flux and active investigation. I believe you'll find more and more of them becoming candidates for 5000–6000 rads given over a course of 186–200 rads per day, however. There are many, many other things we could bring out to you in this list. These points were simply to rapidly whet your appetite, and I sincerely hope that perusing this volume will help you in your practice. Thank you!

J. Lawton Smith, M.D.

Contents

1 Radiation Therapy for Primary Optic Nerve Meningiomas

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Basil M. Yates, M.D.
Don C. Bienfang, M.D.

Optic nerve sheath meningiomas, previously considered to be rather rare,[1–4] have been encountered with surprising frequency in neuro-ophthalmologic practice since the widespread use of computed tomography.[5–7] Early diagnosis led to an enthusiastic surgical approach to these lesions,[8] but this has been greatly tempered by the subsequent realization that even in the best of hands and using careful microsurgical technique, postoperative blindness occurred with distressing frequency.[9,10] In the first reported case of visual recovery following surgical removal of such a tumor,[11] the meningioma arose primarily in the orbit and secondarily surrounded the optic nerve sheath. Subsequently there have been a few other cases favorably managed by a surgical approach, but the management of these tumors still constitutes a difficult decision. This is because the small pial blood vessels of the optic nerve are so intimately involved in sheath meningiomas that often it has been impossible to dissect these lesions from the intravaginal space without resultant infarction of the nerve.

Meningiomas have been generally considered to respond poorly to radiation therapy.[12] Many clinicians, therefore, have elected simply to follow these cases. However, there have been sufficient reports of efficacy of radiation therapy for meningiomas[13,14] to warrant consideration of such therapy in selected instances of these tumors. We quite agree with the importance of differentiating primary optic nerve sheath meningiomas from those secondarily extending into the nerve, as noted by Wright et al.,[15] and also in cases with multiple meningiomas.

Radiation therapy has been employed in selected instances of optic nerve sheath meningiomas in our practice since 1975. Documentation of the results in five patients with optic nerve sheath meningiomas is the subject of this report. The results of irradiation therapy in secondary optic nerve sheath meningiomas, or with multiple meningiomas, will be the subject of another report. To our knowledge, the following cases are the first reported instances of radiation therapy for optic nerve sheath meningiomas.

CASE REPORTS

Case 1 (A.B.)

A 61-year-old right-handed white married woman was first seen in 1975 because she was "worried about vision in the left eye." She always thought the left eye a bit weaker than the right eye, but by January 1975 she realized that the vision was definitely less in the left eye, noted a "veil" diffusely over the entire field of that eye, and volunteered that red objects were not as vivid with the left eye. There was no pain on eye motion, no headache, and no other neurologic complaint.

Examination on March 26, 1975, revealed a corrected acuity of 20/15–2 in the right eye and 20/20–1 in the left eye.

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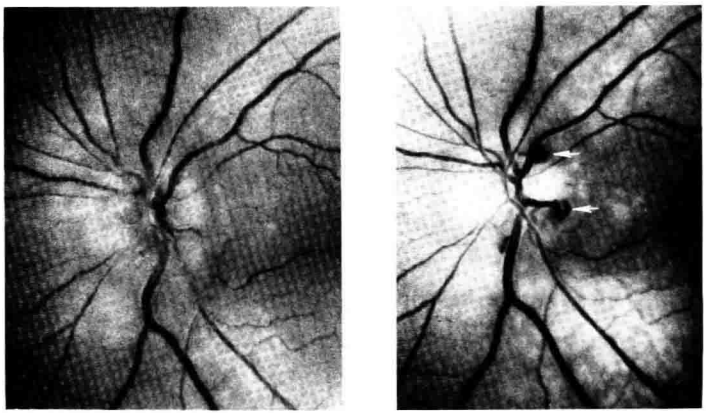

A B

Fig. 1. A. Left optic disc of case 1 in March 1975, before development of optociliary veins. B. Left optic disc of case 1 in April 1979; note optociliary veins at 2 and 4 o'clock.

The palpebral fissures measured 10 mm on the right and 11 mm on the left. The pupils were 4 mm on the right and 5 mm on the left. There was a 3+ Marcus Gunn afferent pupillary reaction in the left eye. Exopthalmometry revealed 2-mm proptosis of the left eye. Visual field examination was normal in the right eye and revealed a very slight lower nasal contraction in the left eye. The right optic disc was normal, but the left disc was blurred and had elevated margins (Fig. 1A). The arterioles were narrow but no hemorrhages were seen.

Because of the slight asymmetry of her eyes, old photographs were reviewed. A picture made in 1942 substantiated the fact that her appearance had not subsequently changed. Because of this photograph, x-rays were not ordered at that time. The clinical impression was between atypical ischemic optic neuropathy, thyroid exopthalmos, and a left orbital meningioma. She was found to have mild diabetes mellitus, and was sent back to her private ophthalmologist with careful follow-up advised.

Four years later, the patient was referred back because of a further drop in left eye vision. She had noted a slow, steady drop of acuity in the eye during the preceding 6–8 months, and could no longer tell colors with the eye. She was taking 16 units of NPH insulin daily, was under good control, and had no other complaints.

Examination on April 16, 1979, revealed a corrected acuity of 20/15−2 in the right eye and 20/80+1 in the left eye. A profound loss of visual field had occurred in the left eye (Fig. 2). There was now a 3-mm proptosis of the left eye. One look at the fundi established the diagnosis for classic optociliary veins[5] were evident on the left disc (Fig. 1B). It was apparent that the patient had an optic nerve meningioma on the left. Roentgenograms of skull, optic canals, and computed tomography (Fig. 3) confirmed this diagnosis. After discussing the matter with the patient, it was elected to treat the left optic nerve with radiation therapy.

Supervoltage radiotherapy treatments to the posterior one-third of the left optic nerve were given between May 7 and June 25, 1979.

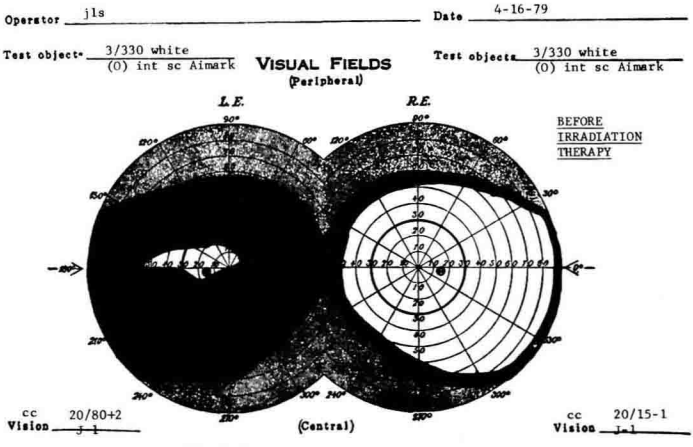

Fig. 2. Visual fields of case 1 before radiotherapy.

Two parallel coplanar opposing portals were used to give 200-rad increments for each treatment session over the 5-week course. A total tumor dose of 5300 rads was given to the left posterior orbital and intracanalicular optic nerve. Within 10 days of beginning the therapy, vision began improving in her left eye. The day after finishing the course of radiotherapy, acuity was 20/50+3 in the left eye and the

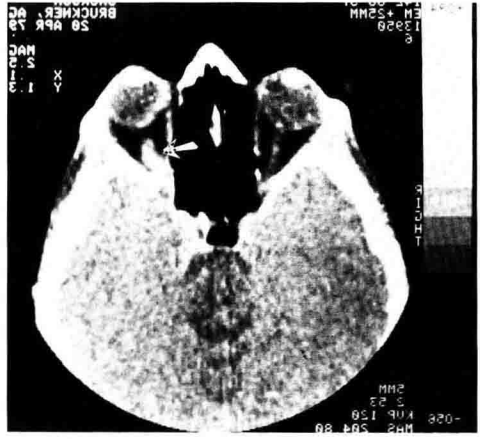

Fig. 3. Computed tomographic scan (Case 1) showing left optic nerve sheath meningioma.

visual field was over three times as large as prior to treatment (Fig. 4). Interestingly enough, the shunt vessels in the left optic disc were smaller the day after radiotherapy had been concluded (Fig. 5). On July 12, 1979, she had 20/40+1 acuity in the left eye and volunteered that whereas she had only been able to see in "black and white" before radiotherapy, she could now detect "blue and yellow" colors in the left eye, but still could not appreciate red and green in the eye. Four months after treatment, vision was 20/50+2 in the left eye, and the field was unchanged from that seen in Figure 4.

The patient returned on October 7, 1980, with no complaints and considered that her vision was stable. Dr. James Mitchell examined her in our office and found the corrected acuity to be 20/20+3 in the right eye and 20/50+2 in the left eye. A careful visual field examination revealed that the inferonasal arcuate scotoma breaking out of the blind spot appeared to be smaller and less dense than had been noted 10 months earlier. Exophthalmometry revealed 2-mm proptosis of the left eye. Ophthalmoscopy revealed that the left optic disc was markedly pale with attenuated vessels on the surface, but the optociliary shunt vessels had completely disappeared (Fig. 5C).

Comment

A 61-year-old woman presented with a swollen left optic disc, 20/20−1 vision, and a modest lower nasal field defect in 1975. Four years later, she returned with acuity down to 20/80 and a profound loss of visual field in the involved eye. Classic optociliary veins (the Hoyt-Spencer sign)[5] were now present. A course of radiation therapy was given to the left optic nerve. Within 2 months, acuity improved from 20/80 to 20/40, the visual field enlarged to at least three times its former size, and the shunt vessels became much smaller. A comparison of the fundus photographs confirmed that the latter was not due to a general decrease of disc vascularity, for whereas the optociliary veins became notably smaller, the other vessels traversing and surrounding the optic disc remained unchanged in size (Fig. 5). She

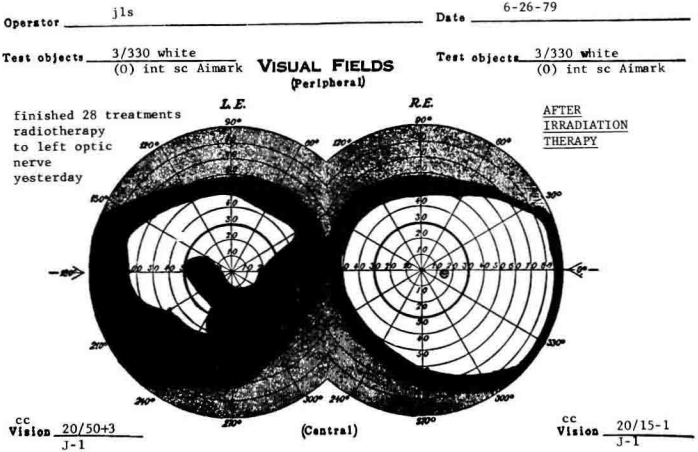

Fig. 4. Visual fields of Case 1, immediately after radiotherapy to left optic nerve.