Review of Choroidal Osteoma: Successful Krypton Red Laser Photocoagulation of an Associated Subretinal Neovascular Membrane Involving the Fovea

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ABSTRACT

Laser treatment of a subretinal neovascular membrane associated with a unilateral choroidal osteoma in a 28-year-old woman is described. To our knowledge, this is the first reported use of krypton red laser photocoagulation for the initial treatment of a subfoveal neovascular membrane in this setting. Early recognition of the membrane allowed treatment prior to involvement of the central fovea. Follow-up has shown obliteration of the membrane without recurrence and 20/20 vision. We emphasize recognition of this unusual tumor, self-monitoring with an Amsler grid, and early treatment of subretinal neovascularization in this high risk group of young patients.

SUBRETINAL NEOVASCULARIZATION IS ASSOCIATED WITH A GROWING LIST OF MACULAR DISEASE PROCESSES IN WHICH BREAKS IN BRUCH'S MEMBRANE ALLOW CAPILLARIES FROM THE CHORIOCAPILLARIS TO PROLIFERATE UNDER THE RETINAL PIGMENT EPITHELIUM (RPE) AND SENSORY RETINA. CHOROIDAL OSTEOMA, A RARE AND OTHERWISE BENIGN CHOROIDAL TUMOR, CAUSES VARYING DEGREES OF DEGENERATION OF THE OVERLYING RPE AND RETINA. PREVIOUSLY, IT IS THIS DISRUPTION OF THE RPE-BRUCH'S MEMBRANE COMPLEX THAT ACCOUNTS FOR THE FREQUENTLY DESCRIBED SUBRETINAL NEOVASCULARIZATION ASSOCIATED WITH THIS TUMOR. SUBRETINAL NEOVASCULAR MEMBRANES (SRNVM) AND THEIR SEQUELAE ARE RESPONSIBLE FOR THE SIGNIFICANT VISUAL LOSS THAT MAY OCCUR EARLY IN THE COURSE OF THIS DISEASE. AS SUCH THEY REPRESENT A POTENTIALLY TREATABLE CAUSE OF BLINDNESS IN THIS YOUNG GROUP OF PATIENTS. IN HIS ORIGINAL SERIES OF PATIENTS WITH CHOROIDAL OSTEOMA, GASS FIRST ENTERTAINED THE POSSIBILITY OF PHOTOCOAGULATION TREATMENT TO HALT THE EXUDATIVE PROCESS ASSOCIATED WITH THE OSTEOMA AND MEMBRANE.

There are now a few reports of extrafoveal SRNVM associated with choroidal osteoma treated with argon

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laser photoocoagulation,\textsuperscript{4-6} and one report of a recurrent membrane threatening the fovea that was secondarily treated with krypton red laser (KRL) photoocoagulation.\textsuperscript{6} Several investigators have demonstrated the theoretical advantage\textsuperscript{7,8} and clinical efficacy\textsuperscript{9,10} of KRL for SRNVM at or near the foveal avascular zone (FAZ). We report an encouraging case of unilateral choroidal osteoma with an associated subfoveal neovascular membrane that was treated primarily with KRL photoocoagulation. To emphasize structure-function relationships, we will refer to the anatomic definition of posterior pole structures when describing the location of lesions.\textsuperscript{11}

CASE REPORT

Our patient is a 28-year-old optometrist's daughter, who was initially referred to the Wills Eye Hospital Retina Vascular Unit (LEM) on November 22, 1985, complaining of two months of decreased visual acuity in the right eye, accompanied by occasional flashes. There was no history of recent ocular trauma. With the exception of oral birth control pills, medical history and family history were unremarkable. The report of a previous examination in August 1985 showed that (with hyperopic correction) her visual acuity measured 20/25 in each eye.

Initial ocular examination revealed a best corrected visual acuity of 20/40 in the right eye and 20/15 in the left eye. A right esotropia was noted. Slit lamp examination of the anterior segments revealed nothing remarkable. Funduscopic examination of the right eye disclosed a slightly elevated yellow choroidal lesion with well demarcated margins involving the superior portion of the optic disc and extending into the macula (Figure 1). A greyish-green subretinal neovascular membrane encroaching upon the fovea was noted at the inferotemporal margin of this lesion. Several retinal folds with subretinal fluid and hemorrhages were present. The left fundus was normal.

B-scan ultrasonography of the right eye showed a highly reflective peripapillary lesion extending temporally from the optic nerve with acoustic shadowing of the retrobulbar tissues (Figure 2A). This highly reflective lesion persisted despite reduced sensitivities that eliminated all other posterior wall echoes (Figure 2B). These characteristic B-scan findings confirmed the clinical diagnosis of choroidal osteoma.

Fluorescein angiography of the right eye showed early mottled hyperfluorescence over the osteoma with three macular blocking defects corresponding to hemorrhages (Figure 3A). An active subretinal neovascular membrane encroaching on the foveal avascular zone superiorly became evident as the angiogram progressed (Figure 3B). On later phases of the angiogram there was marked leakage, which became obscured by the background hyperfluorescence over the entire mass (Figure 3C).

In an effort to stabilize this process and limit macular damage, the SRNVM was treated directly with krypton red laser photoocoagulation. With the intent of completely obliterating the membrane, a total of 280 burns, 350 mW, 200 μm each for 0.2 to 0.5 seconds were applied.

Three weeks later, on December 12, 1985, the patient

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\caption{Photograph of right eye in November 1985, shows well-demarcated osteoma extending superiorly from the optic disc. There is a blue-green subretinal membrane along the temporal margin of the tumor with serous leakage into the macula and three small hemorrhages.}
\end{figure}

\begin{figure}[h]
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\caption{B-scan ultrasonography of right eye showing acoustic shadowing behind dense tumor (A) and persistent echoes from highly reflective osteoma when sensitivity is lowered so that all other posterior wall echoes are eliminated (B).}
\end{figure}
was re-examined. She reported a definite improvement in vision, which now measured 20/30+ in the right eye. Funduscopic examination revealed no new hemorrhage or exudate (Figure 4). Repeat fluorescein angiography showed complete eradication of the SRNVM (Figure 5).

At the last examination, on March 11, 1986, four months following treatment, visual acuity had returned to 20/20 in the right eye without further evidence of SRNVM or hemorrhage.

**DISCUSSION**

Choroidal osteoma is a benign ossified tumor first described in 1978 by Gass, who published the largest reported series (19 eyes in 15 patients) in 1979. These tumors were observed predominantly in young healthy white women. In 20% of the cases the condition was bilateral. Since that initial series, a number of cases have been reported, making a total of 53 eyes in 39 patients. The majority of patients have been female (82%) ranging in age from five to 69 years, although most patients have been in the second to third decade. One third of all reported cases to date have been bilateral.

Histopathological studies reveal a tumor of mature bone with hypocellular marrow containing...
some dilated thin-walled blood vessels that communicate on the surface of the tumor with a capillary network beneath Bruch's membrane and a degenerated retinal pigment epithelium. There is mild degeneration of the overlying retinal receptors. The pathogenesis of this tumor remains unknown. Originally thought to be a developmental tumor, more recent reports suggest that it is acquired. Osseous proliferation has been observed following trauma or inflammation. Hormones and unidentified environmental toxins may be factors. The tumor has been reported in association with systemic disease and intraocular inflammation. As in our case, this condition can develop de novo in a previously unaffected eye. Hereditary factors may also be involved.

Observed complications of choroidal osteoma include SRNVM and their sequelae: subretinal hemorrhage, serous and hemorrhagic detachment, and disciform scarring. The typically slow growth of the tumor, together with a false-positive 32p uptake (radioactive phosphorus is absorbed by tumor calcium), has prompted enucleation for suspected choroidal melanoma. The differential diagnosis includes amelanotic choroidal melanoma, atypical choroidal hemangioma, and metastatic choroidal tumor. The distinguishing diagnostic findings are on ultrasonography and CT scan which both demonstrate a high density choroidal mass. Although the fluorescein angiographic features are nonspecific, high quality photography is essential in the detection of early SRNVM, when laser treatment is most likely to be beneficial.

Although choroidal osteoma is a benign tumor, it may result in severe visual deficits. In Gass' original series, albeit small, 80% had 20/30 or better vision in the affected eye on initial examination, with less than 10% having 20/200 vision or worse. However, follow-up observation revealed deterioration to worse than 20/200 in one-quarter of the cases. In our review of all reported cases to date, approximately two-thirds of the affected eyes had 20/30 or better vision initially with one-half of the untreated eyes seeing 20/200 or less at final follow-up. Visual loss has been attributed to both RPE and retinal atrophy as well as SRNVM and its sequelae.

Despite the predominantly central location of this tumor, many with uncomplicated choroidal osteoma enjoy good visual acuity. It has been suggested, then, that it is the associated SRNVM, with all it entails, rather than the osteoma itself, that presents the greatest threat. Among cases reported to date, more than 50% of affected eyes either had or developed SRNVM and its sequelae during the reported follow-up periods. Excluding the laser treated eyes, 60% of those patients had 20/200 vision or worse at last follow-up. The poor final visual acuity (20/200 or worse) of 70% of all untreated eyes was attributed to SRNVM or its complications.

The histopathologic and clinical progression of subretinal neovascularization has been described as including subretinal hemorrhage with hemorrhagic detachment, serous detachment, cystoid edema with resultant exudate, and ultimately disciform scarring with irreversible visual loss. Laser photocoagulation is the principle therapy used to halt this disciform process. Extrafoveal SRNVM associated with age-related macular degeneration has recently been proven treatable with argon blue-green laser. Recent clinical trials have suggested the efficacy of KRL in the treatment of SRNVM involving the foveal avascular zone.

There are only a few reports of laser treatment of SRNVM associated with choroidal osteoma. Alexander and Hunyor of Australia reported a 32-year-old patient with bilateral disease and extrafoveal SRNVM who was treated with xenon photocoagulation but continued to lose vision to the counting fingers level. In 1983, Burke and Brockhurst reported the first case treated with argon photocoagulation in a 19-year-old patient whose vision dropped to 20/200 when a SRNVM with hemorrhage and serous detachment occurred. Treatment eradicated membrane and resolved the fluid and hemorrhage. Although vision did not improve, it was stabilized without further loss. Avila reported a case of bilateral choroidal osteoma in which the first eye that developed SRNVM was untreated and went on to lose vision because of subretinal hemorrhage. The fellow eye was treated with argon green photocoagulation when vision was threatened by an extrafoveal SRNVM with serous leakage and hemorrhage. A second treatment was required to obtain closure of neovascularization. Vision was stabilized at 20/60.

Grand and his associates reported three cases. The first case was a bilateral one in which the first eye demonstrated the progression of SRNVM to end stage disciform scarring and finger counting vision. The fellow eye was treated with argon green laser for extrafoveal subretinal neovascularization. Recurrences required multiple retreatments with KRL as the SRNVM approached and finally involved the fovea completely; final vision was 20/60. Their second case of SRNVM encroaching on the fovea was treated with AGL with excellent results; eight years later there was no recurrence of membrane, and vision was 20/25-. Their third case of extrafoveal SRNVM with serous detachment and hemorrhage was treated with AGL and resulted in 20/20 vision prior to the development of an internal limiting membrane contracture that decreased vision to 20/100.

Our report illustrates a case of unilateral choroidal osteoma in a young woman who had minimal but noticeable visual loss caused by a subfoveal SRNVM. Immediate KRL photocoagulation yielded excellent results: complete obliteration of the membrane and 20/20 vision. This case demonstrates the value of recognition and therapy while the visual deficit is still small and before extensive hemorrhage or scarring has
developed. Thus we need to diagnose choroidal osteoma and monitor those with this condition, as well as others at risk of developing SRNVM, with daily Amsler grid testing. Although not required in this case, others have cited the need for multiple photocoagulation treatment of membranes associated with choroidal osteoma because of what appears to be poor laser absorption by the atrophic RPE and hypomelanotic tumor. Thus post-treatment angiography should be used to detect persistent neovascularization as well as recurrent neovascularization in other sites. Since we are limited to treatment aimed at stopping disease progression, rather than halting the underlying etiology, continued monitoring of the central vision to detect membrane formation is extremely important.

Recently a case of subretinal neovascularization associated with choroidal nonperfusion and retinal ischemia has been reported, suggesting ischemia as the stimulus of neovascularization. In the case of choroidal osteoma, perhaps the inadequate “trans-tumoral blood supply” results in retinal ischemia. The enlarging tumor itself may be responsible because by demanding additional blood flow and oxygen it renders the overlying retina hypoxic. Preliminary results of treating SRNVM associated with choroidal osteoma with AGL and now KRL are encouraging.

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REFERENCES