Limited Macular Translocation for Atrophic Maculopathy

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Objectives: To report visual improvement following bilateral limited macular translocation for a patient with atrophic macular disease, and to discuss issues related to the selection of potential candidates for this technique.

Design: Case report.

Results: A 78-year-old woman with bilateral atrophic maculopathy and no choroidal neovascularization had slowly progressive loss of visual acuity for at least 17 months in the right eye and 25 months in the left eye. She underwent bilateral limited macular translocation, using scleral infolding in the right eye and scleral outpouching in the left eye. Following translocation of her maculae, her best-corrected visual acuity improved from 20/200 to 20/30 OD and from 20/180 to 20/100 OS. She remained stable during 30 months of follow-up for the right eye and 22 months of follow-up for the left eye.

Conclusion: Macular translocation may allow visual recovery in selected patients with atrophic maculopathy, even after a prolonged period of poor vision.

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MACULAR translocation was developed as a technique for moving the fovea relative to underlying choroidal neovascularization. This allows both the photocoagulation of formerly subfoveal choroidal neovascularization, while sparing the foveal center, and the repositioning of the fovea over healthier retinal pigment epithelium (RPE). Machemer and Steinhorst first reported success with this technique in 1993 by using a 360° retinotomy to translocate the macula. Later, de Juan et al and Pieramici et al used scleral shortening to perform limited macular translocation without the need for a 360° retinotomy. Others have confirmed this initial success using both techniques. Theoretically, macular translocation may be useful for treating atrophic macular disease by placing the fovea over healthier RPE, which could prevent the loss of photoreceptors and sustain foveal function. This article presents the results of limited macular translocation in both eyes of a patient with atrophic macular disease, presumed to be caused by pattern dystrophy, who had poor visual acuity for a prolonged period of time prior to the procedure.

REPORT OF A CASE

A 78-year-old woman sought care from her ophthalmologist in May 1996 because of decreased vision in her left eye. At that time, her best-corrected visual acuity was 20/50 OD with a refractive error of +1.00 −2.00 × 110° and 20/80 OS with a refractive correction of +2.75 −1.50 × 110°. She had macular changes consistent with atrophic maculopathy. By May 1997, her best-corrected visual acuity had declined to 20/80 OD and 20/100 OS. By March 1998, the visual acuity had further declined to 20/100 OD and 20/200 OS. By July 1998, her visual acuity had declined further to 20/200 OD and 20/400 OS. She was then referred to one of us (J.D.B.) to see if any treatment was available.
On the initial evaluation in August 1998, the patient’s best-corrected visual acuity was 20/200 OD and 20/400 OS. She had bilateral posterior chamber intraocular lenses and a circular area of RPE atrophy and pigmentary change beneath the fovea, with the left eye worse than the right eye (Figure 1A and Figure 2A). In both eyes, she was able to accurately fixate foveally and follow a 200-µm diameter spot projected from the slitlamp (Haag-Streit AG, Koniz, Switzerland). A fluorescein angiogram showed central blocked hypo fluorescence surrounded by a rim of transmission hyperfluorescence within the macula in the right eye, with more prominent transmission hyperfluorescence in the left eye (Figure 1B and Figure 2B). There was no evidence of occult or classic choroidal neovascularization. The patient asked if any treatment would be available to either improve her vision or slow the progression of the macular degeneration she had experienced during the last 4 years. Macu-
lar translocation was discussed because it offered the possibility of relocating the fovea over healthy and viable RPE. After carefully considering all the known associated risks and potential benefits, she elected to undergo macular translocation in her right eye. In October 1998, one of us (J.D.B.) performed a limited macular translocation us-

Figure 2. Left eye: A preoperative fundus photograph (A) shows a central atrophic lesion, within which xanthophyll is visible in the color photograph. A preoperative fluorescein angiogram frame (B) shows patchy transmission hyperfluorescence, consistent with atrophic maculopathy. A preoperative infrared scanning laser ophthalmoscope (SLO) image (C) shows fixation within the atrophic lesion. The cross indicates fixation; shaded circles, stimulus that has been seen; and open triangles, stimulus that has not been seen (scotoma). A number of stimulus intensities were employed, with the brighter symbols indicating a dimmer stimulus. When the cross was fixated within the atrophic lesion, few stimuli were detected near it. When the cross was moved out of the field, the area had a significant relative scotoma (but not a full dense scotoma). A preoperative argon blue SLO image (D) shows the presence of xanthophyll (dark area) within the atrophic lesion. A preoperative autofluorescence SLO image (E) shows a mottled, but not complete, loss of autofluorescence in the atrophic region. An argon blue SLO image (F) from 14 months after macular translocation shows that some of the xanthophyll is still overlying the atrophic lesion. A 14-month postoperative infrared SLO image (G) shows that fixation is inferior to the atrophic region, in the area of xanthophyll seen in F. There is a dense scotoma in the retinal area now overlying the atrophic region.
ing circumferential scleral infolding, as described by de Juan et al.2 The technique included a pars plana vitrectomy and creation of a temporal retinal detachment by infusing fluid into the subretinal space through a 39-gauge subretinal needle. Scleral infolding was performed by tying preplaced 5-0 Mersilene (Ethicon Inc, Somerville, NJ) sutures in the superior temporal quadrant. A gas bubble using sulfur hexafluoride gas was placed in the vitreous cavity, and the patient was positioned nasal side up for 5 minutes. She was then positioned upright to allow the gas bubble to translocate the macula inferiorly and reattach the retina. Postoperatively, her course was uncomplicated, with spontaneous reattachment of the retina and good translocation of the macula (1191 μm).

Two months after surgery, her visual acuity had improved to 20/70-1 OD, but her refractive error had increased dramatically, with a significant amount of induced astigmatism. Her new refractive error in the right eye had changed from her baseline of +1.00 –2.00 × 90° (which had been confirmed on several occasions preoperatively by refraction, keratometry, low vision evaluation, and automated refraction) to +3.00 –6.75 × 80°. Four months after surgery, her visual acuity had improved to 20/50 +2 OD with the same refractive error. To help eliminate the anisometropia caused by the induced astigmatism, she underwent an astigmatic keratotomy in the right eye. Eleven months after translocation surgery, her visual acuity had improved to 20/40 +2 OD with a refractive error of +2.00 –0.75 × 60°. Her acuity had generally been in the 20/40 to 20/50 range, but at her most recent visit, 30 months postoperatively, her best-corrected visual acuity was 20/30 OD.

Given her success with surgery in the right eye, the patient inquired about surgery on the left eye. The patient was assessed at the Wilmer Eye Institute, Baltimore, Md, in June 1999. The patient reported that her left eye had always been her weaker eye. She also reported that she was occasionally aware of a blind spot in her left eye and that she had been aware of a blind spot in the right eye, but that this had resolved following the translocation surgery. The patient reported some difficulty with night vision but normal color vision and peripheral vision. Her best-corrected visual acuity, measured with an ETDRS (Early Treatment of Diabetic Retinopathy Study) chart, was 20/50 OD and 20/180 OS. The patient missed the middle letters of some lines with her left eye. Her Pelli-Robson contrast sensitivity was reduced to 1.35 OD and 1.20 OS (normal, ≥ 1.65). No relative afferent pupillary defect was present. A dilated examination of the right fundus (Figure 1C) revealed an atrophic lesion located superior to the fovea that was not discrete and “punched out” as advanced geographic RPE atrophy generally appears. In the left eye, there was an atrophic lesion of similar character involving the foveal center (Figure 2A). The patient did not have high-risk drusen or the pigmentary alterations typically associated with a pattern dystrophy in either eye.

Scanning laser ophthalmoscope (SLO) macular perimetry showed that fixation was inferior (on the retina) to the atrophic RPE lesion in the right eye and that the area over the atrophic RPE lesion had a dense scotoma (Figure 1D). There was a mild reduction in general retinal sensitivity outside the area of her lesion. With argon blue imaging, the area of xanthophyll was prominent and outside the atrophic lesion (Figure 1C). Autofluorescence imaging showed showed patchy black areas within the atrophic lesion rather than the solid black appearance seen in geographic atrophy with loss of RPE (Figure 1E). In the left eye, the patient fixated within the area of the central atrophic lesion (Figure 2C). In this lesion, only the brightest SLO stimulus (0 dB with a Goldmann III-sized target) could be seen. Peripherally, there was normal sensitivity, except for a mild relative scotoma inferonasally. Argon blue imaging showed xanthophyll present within the atrophic lesion (Figure 2D), and autofluorescence imaging showed patchy black areas within the region of this lesion (Figure 2E).

To better define the cause of her macular lesions, electrophysiological and visual function testing was performed. Electro-oculography was performed to evaluate any evidence of Best disease. The Arden ratio was normal in both eyes (2.64 OD and 2.84 OS; normal, ≥ 1.80). Electoretinography was performed to assess the presence of diffuse retinal disease as a cause of the macular lesions. The scotopic responses were normal. The flicker responses were reduced to approximately 50% of normal in the right eye and 80% of normal in the left eye, and the photopic flash response was reduced to about 80% of normal in the right eye and was normal in amplitude in the left eye, but these responses were somewhat delayed. The Farnsworth D15 panel color vision was normal in the right eye and showed 1 minor error in the left eye.

The macular lesion in the left eye showed evidence of preserved photoreceptor function within the lesion. This conclusion was based on the patient’s ability to fixate centrally in the left eye, the ability to see bright stimuli within the lesion, the presence of xanthophyll within the lesion, as demonstrated by argon blue imaging, and the absence of a completely black appearance on autofluorescence imaging. Although her vision had been decreased to 20/100 or worse in this eye for 25 months, the results of all her measurements suggested that there was a reasonable chance for improvement in foveal function if macular translocation were performed.

The patient therefore underwent limited macular translocation in the left eye by one of us (J.D.B.) in June 1999. This time, a different technique was performed, using radial scleral outpouching.7 The technique was otherwise identical to that performed in the right eye, except that scleral shortening was performed by creating a radial scleral outfold. A scleral folding clamp was used to create a radial scleral outfold, followed by placement of a full-thickness 6-0 Prolene (Ethicon Inc) horizontal mattress suture to secure the radial fold. Postoperatively, her course was uncomplicated, with macular translocation of 832 μm and prompt retinal reattachment. Postoperatively, her visual acuity remained in the 20/200 to 20/400 OS range, although at the Wilmer Eye Institute, she attained 20/100-2 with an ETDRS chart (improved from 20/180 preoperatively at the Wilmer Eye Institute) and 20/100 (Snellen) at 1 other visit. Her refractive error was essentially unchanged from the preoperative level, with no induced astigmatism in the left eye. At a fol-
low-up visit 22 months postoperatively, her best-corrected visual acuity was 20/200.

She was reevaluated at Wilmer Eye Institute by one of us (J.S.S.) in August 2000, which was 22 months postoperatively in the right eye (Figure 1F) and 14 months postoperatively in the left eye (Figure 2F). She reported that the vision in the right eye had gotten better and the vision in her left eye was better than it had been preoperatively. She reported that her night vision continued to be somewhat reduced, as it had been all her life. She denied having diplopia, except when looking at Christmas lights. On this visit, her best-corrected visual acuity was 20/50-1 OD and 20/100-2 OS (ETDRS chart). She was able to read 1 M print with the right eye and 2.5 M print with the left eye at 40 cm. The SLO macular perimetry showed a dense scotoma over the areas of RPE atrophy in each eye (Figure 2G). Argon blue SLO imaging of the left eye demonstrated that the xanthophyll area had been translocated inferior to the atrophic lesion, except for the superior-most portion, which remained partially over the atrophic RPE (Figure 2F). This was in contrast to the right eye, where the entire xanthophyll area had completely cleared the atrophic lesion (Figure 1F).

VISUAL IMPROVEMENT

Several aspects of this case are remarkable. This patient had a significant improvement in vision in her right eye after having had slowly progressive visual acuity loss, with acuity at 20/80 or worse for at least 17 months prior to the procedure in her right eye and for at least 37 months prior to the procedure in her left eye. Her visual acuity improved from 20/200 to 20/30 OD (best acuity, with acuities ranging from 20/30 to 20/50) and from 20/186 to 20/100 OS (best acuity, with acuities ranging from 20/100 to 20/200). The amount of foveal translocation was greater in the right eye (1191 µm) than the left eye (832 µm). As a result, the fovea (as delineated by the xanthophyll) was relocated completely outside the atrophic RPE in the right eye and only partially outside the slightly larger atrophic area in the left eye. The more atrophic appearance of the lesion preoperatively and the longer duration of visual loss may have accounted for the more limited amount of improvement in the left eye. That her vision improved after having been decreased for so long suggests that her foveal photoreceptors had remained viable over the atrophic lesion during this time. After they were relocated to a new site over healthy RPE, the photoreceptors apparently were able to function more effectively, and her vision improved.

PATIENT SELECTION

This case illustrates the use of macular translocation for patients with atrophic macular disease. Preoperative evaluation of visual function may provide insight into which eyes are likely to benefit from translocation. The most critical element is residual photoreceptor function and foveal fixation within the atrophic region prior to the procedure. In this patient, the SLO evaluation was used to make this determination in the left eye, and a 200-µm diameter slit-lamp spot was used to assess fixation in both eyes. Another indirect measure of viability and residual photoreceptor function is the presence of xanthophyll within the atrophic lesion. Xanthophyll may be visualized by ophthalmoscopic examination and standard fundus photography, but its visualization is enhanced using either argon blue SLO imaging or fundus photography with an appropriate filter for detecting xanthophyll. Finally, the SLO can be used to identify residual RPE within the atrophic area. The SLO macular perimetry preoperatively showed fixation and residual function within the atrophic lesion (although postoperatively there was a dense scotoma of the retina now overlying the atrophic lesion). Our patient had mottled, rather than totally black, autofluorescence in the atrophic lesion. This provides further suggestive evidence that the photoreceptors are viable and complements macular perimetry over the atrophic lesion.

The exact cause of this patient's atrophic maculopathy is unclear. Her progressive visual loss in her late 70s and the well-circumscribed area of subfoveal RPE atrophy are consistent with age-related macular degeneration. Yet, the absence of drusen, the blocked hypofluorescence on the fluorescein angiogram, and the retention of fixation within the lesion are more consistent with a pattern dystrophy of the macula. Additionally, the pattern of a patchy decrease in autofluorescence seen with the SLO in this patient was different from the homogeneously black area (loss of autofluorescence) seen in patients with geographic atrophy from age-related macular degeneration. The electro-oculogram was normal, so Best disease was not the cause. The electroretinogram showed a mild reduction in cone amplitudes and some delay in cone responses, suggesting a cone dystrophy, but the color vision was essentially normal. The patient did not have a dark choroid or flecks that would suggest Stargardt disease. The most likely diagnosis is RPE atrophy, resulting from pattern dystrophy. Early RPE changes and atrophy, from age-related macular degeneration and related conditions, may be associated with decreased retinal function even before frank geographic atrophy occurs; this decreased function may reverse with translocation to a healthier area of RPE.

Other forms of atrophic macular disease may not benefit from translocation surgery, so careful consideration must be given to the likelihood of visual improvement. Geographic atrophy that already involves the fovea, with a dense central scotoma, will likely not benefit from translocation. Translocation that will move the fovea to an area that will likely develop frank RPE atrophy in the future is not likely to benefit the patient. For example, in a patient with a bull's-eye scotoma surrounding the foveal center, macular translocation might relocate the fovea to a site with a higher risk of becoming atrophic than the foveal center itself. Because geographic atrophy often develops in the parafoveal region first, thought must be given to the health of the RPE in the area to which translocation is contemplated. The patient with geographic atrophy from age-related macular degeneration most likely to benefit from macular translocation is one with recent visual loss who has an isolated atrophic lesion just superior to the fovea that is threatening to spread into the foveal center. In this case, macular translocation would
move the foveal center away from the atrophic RPE, reducing the likelihood that atrophy would spread to involve the new foveal center.

This case suggests that limited macular translocation may be helpful for patients with certain other atrophic maculopathies. However, the disease process causing the atrophy may well be a factor in determining whether surgery would be likely to be beneficial. If enlargement of atrophy over time is anticipated, one might have to contemplate performing further translocation at a later time. In limited macular translocation, the fovea is generally translocated inferiorly because of anatomical and surgical limitations. In terms of visual function, when the fovea is moved inferiorly, the atrophic lesion is then positioned beneath a focal area of retina superior to the fovea. The patient will then have a scotoma inferior to fixation (corresponding to the superior retina that overlies the atrophic lesion), which is a disadvantageous position.8,15 (Presumably, the recovery of central vision outweighs the disadvantage of having an inferior scotoma.) One possibility for avoiding an inferior scotoma is to perform a 360° retinotomy to allow enough superior movement of the fovea to be beneficial. Whether the additional risk associated with this technique is warranted remains to be seen.

POSTOPERATIVE LOSS OF FUNCTION OF RETINA OVERLAPPING THE ATROPHIC LESION

An interesting observation was the loss of functioning retina in the region that came to reside over the RPE atrophy in each eye after surgery. This was demonstrated at 8 months postoperatively in this case. The loss of function of the previously normal retina overlapping the RPE atrophy following macular translocation has been shown to develop as early as 1 week postoperatively (G. Y. Fujii, MD, and J.S.S., unpublished data, 2000). Prior to translocation, our patient had a relative scotoma in the fovea overlapping the RPE atrophy, but she could still see the brightest stimulus. Following the translocation surgery, the retina that was relocated over the RPE atrophy, and had previously tested normal with the SLO, now had a dense scotoma. The fact that function remained in the foveal retinal area overlapping the RPE prior to translocation could be due to specific protection conferred on the foveal photoreceptors, or it may reflect additional damage to the RPE-photoreceptor interface following translocation.

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