having had an upper respiratory tract infection 1 month earlier. Visual acuity was 20/60 OD and 20/30 OS. Ophthalmoscopy showed scattered active foci of whitening of the outer retina in both eyes (Figure 1A and B). Fluorescein angiography showed hypofluorescence during the early phases of the angiogram with late staining of the lesions (Figure 1C and D). A diagnosis of APMPPE was made, but no treatment was given. During the next 3 months, his symptoms gradually resolved and visual acuity returned to 20/20 OU. The patient had no additional eye symptoms until 16 years later, when he noted the onset of a temporal scotoma and peripheral shimmering in the left eye. Best corrected visual acuity was 20/25 OD and 20/30 OS. The vitreous body was clear. The macular area showed the old lesions of APMPPE (largely unchanged and without the development of significant atrophy). However, both eyes showed atrophic punched-out choriotinal scars (Figure 2) in the macula, midperiphery, and far periphery. The appearance of these scars and the patient’s symptoms were consistent with a diagnosis of MFC.

Comment. Gass4 has proposed that many of these white spot syndromes, including MEWDS, punctate inner choroidopathy, and MFC, as well as acute zonal occult outer retinopathy, have common characteristics and may be part of the spectrum of a single disease. He has suggested an infectious cause, in which an unknown virus enters the retina from the peripapillary area or perhaps the ora serrata.5 Jampol and Becker6 have suggested that these entities are distinct inflammatory diseases, although overlapping cases and the occurrence of 2 of the entities in a single patient may occur. They concurred with the common genetic hypothesis that autoimmune inflammatory disease may explain these findings. These patients may have genetic loci that predispose them to immune dysregulation and ocular and systemic autoimmune disease. This may explain the potential for 2 entities occurring in a single patient or overlapping symptoms. The case presented herein is a demonstration of unusual concordance of 2 rare diseases, APMPPE and MFC, in the same patient, separated by 16 years. Although the patient’s initial appearance at age 18 years was highly suggestive of APMPPE, this may have been an atypical presentation of MFC. However, the clinical course was most consistent with APMPPE. The fact that the APMPPE lesions, even 16 years later, did not resemble the MFC lesions supports our conclusion that 2 distinct entities occurred in this patient. This occurrence is consistent with the common genetic hypothesis of a genetic predisposition to autoimmune diseases, which allowed both diseases to occur in a single individual.

Jeevan R. Mathura, Jr, MD
Lee M. Jampol, MD
Mark J. Daily, MD

Financial Disclosure: None.
Correspondence: Dr Jampol, 645 N Michigan Ave, Suite 440, Chicago, IL 60611 (ljampol@northwestern.edu).

Funding/Support: This study was supported in part by an unrestricted grant from Research to Prevent Blindness, Inc, New York, NY.


Normalization of Upper Eyelid Height and Contour After Bony Decompression in Thyroid-Related Ophthalmopathy: A Digital Image Analysis

The treatment of eyelid retraction in Graves disease is one of the most challenging aspects of ophthalmic plastic surgery. There are numerous theories about the etiology of upper eyelid retraction, including overaction of the lid retractors in combination with fibrosis of the inferior rectus muscle, fibrosis of the lacrimal gland and adjacent levator aponeurosis, enlargement of the levator fibers, and increased sympa-
thetic tone in the superior tarsal muscle.\textsuperscript{1-4}

Several anatomical explanations account for the lateral accentuation of upper eyelid retraction that occurs in Graves disease. In 1980, Grove\textsuperscript{2} noted fibrosis in the lacrimal gland and adjacent lateral levator aponeurosis, perhaps indicative of more lateral eyelid retractor shortening. In 1991, Lemke\textsuperscript{3} noted that the forces that affect the upper eyelid are governed by variations in orbital size and shape, globe size and position, and the length-tension characteristics of the eyelid structures. Enhanced lateral upper eyelid retraction occurs in part because the eye in primary position must adduct away from the axial projection of the orbit and exposes more lateral sclera. This retraction is accentuated by axial proptosis.

To further investigate the influence of the anatomical position of the globe on eyelid retraction and lateral accentuation of retraction, we analyzed digital photographs of patients’ eyelids taken before and after bony decompression surgery with no additional lid recession procedures. In this fashion, the effect of reduced proptosis on the contour and lateral accentuation of eyelid retraction could be measured in a consistent patient population.

Methods. Twenty-one patients (5 men and 16 women; age, 27-60 years) with Graves disease were photographed before any orbital or eyelid surgery. A total of 38 eyes were included. Patients were placed in a modified slitlamp headrest. A posterior strap held the head in place with the chin and forehead resting firmly against the appropriate rests. The height of the chin rest was adjusted to place both lateral canthi at the level of a previously marked point on the headrest; this ensured proper alignment of the head both horizontally and vertically. A digital camera (Nikon Coolpix 950 digital camera; Nikon USA, New York, NY) mounted at a fixed distance and height was used to photograph patients. Mounted light fixtures were used to obtain consistent lighting; flash was not used to guard against the blinking reflex.

Opposite eyes were occluded to ensure fixation with the photographed eye. Patients were asked to fixate on a mounted pointer adjacent to the camera lens to obtain axial photographs. Preoperative Hertel measurements were taken by one of the authors (E.L.C.) after photography.

Decompression surgery without lid surgery was performed as an elective procedure and performed by one surgeon (P.A.D.R.). Criteria for surgery included patients with clinically stable, thyroid-related ophthalmopathy, no prior history of any eyelid or orbital surgery, and cosmetically significant proptosis. Stable thyroid-related ophthalmopathy was defined as no detectable changes during serial clinical examinations in the patient’s visual acuity, color-plate measurements, proptosis as measured by a Hertel exophthalmometer, strabismus, or lid retraction for at least 6 months. In 34 eyes, bony decompression of the medial wall and posterior medial floor of the orbit was performed via a transcaruncular approach. The thick bony strut present between the maxillary sinus and ethmoid sinus at the maxillo-ethmoid junction was preserved to prevent hypoglobus. The medial floor of the orbit was removed up to the medial border of the infraorbital neurovascular bundle. Two patients (4 eyes) had bony decompression performed in conjunction with the otolaryngology service via an endoscopic endonasal approach. Only patients with surgery performed via these 2 approaches were included because these approaches avoid scarring and inadvertent injury to the eyelids that might alter their position.

Patients were asked to return at 3 months postoperatively for follow-up clinical examination. At that time, patients were photographed using the previously described protocol, and the same preoperative examiner repeated Hertel measurements.

All acquired images were transferred to a personal computer for digital analysis. Digital images were analyzed using NIH ImageJ 1.27 software.\textsuperscript{6} After threshold and contrast adjustment, digital photographs were calibrated by assuming each cornea to have a horizontal diameter of 12 mm. The horizontal diameter of the cornea was measured using the computer to highlight the cornea on the basis of color contrast differences between the conjunctiva and the cornea and calculating the numerical width of the cornea in pixels. This reference pixel width was then assigned a value of 12 mm for calibration. A reference point was marked at the medial canthal angle. The magic wand tool was then used to create a selection by tracing the junction of the eyelid margin and the conjunctiva. To trace this junction, the pointer was placed on top of the medial canthus reference point, and the magic wand tool was activated. The result is an imaginary turtle that starts moving to the right, looking for an edge. Once the turtle finds the edge, the turtle follows the edge until it returns to the starting point (Figure 1). Using this tracing, the x and y coordinates of the peak of the upper eyelid were calculated. The horizontal distance to the peak of the upper eyelid was measured from the previously placed reference point. In addition, a vertical line through the centroid (ie, the center point of the selection, the average of the x and y coordinates of all pixels in the selection) of the palpebral fissure was used to measure the palpebral fissure height. The reproducibility of the palpebral fissure area measurements resulted in a standard deviation of ±0.03% using this automated tracing technique. Use of the magic wand tracing tool to perform automated tracing of the photographs at the junction of the conjunctiva and the lid margin, based on contrast differ-
ences as measured by the computer, results in minimization of investigator bias and an increase in the reproducibility and repeatability of these measurements.

Results. The average preoperative Hertel measurement was 25.1 mm. The average Hertel measurement 3 months after surgery was 21.6 mm. The average reduction in proptosis after bony decompression surgery was 3.5 mm ($P < .001$). The palpebral fissure height decreased from 12.27 mm to 9.38 mm after decompression.

The average exposed surface area of the palpebral fissure before decompression surgery was 292.92 mm$^2$. Three months after surgery, the average exposed surface area of the palpebral fissure was 236.8 mm$^2$. The average reduction in exposed surface area in the palpebral fissure was 56.12 mm$^2$ ($P < .001$) (Figure 2).

The average height of the palpebral fissure was 14.21 mm before surgery and 12.5 mm after surgery, with an average 1.71-mm reduction in the vertical height of the palpebral fissure after decompression surgery ($P = .002$).

The average distance from the peak of the upper eyelid to the medial canthus reference point was 9.1 mm. The average distance from the peak of the upper eyelid to the medial canthus reference point 3 months after surgery was 7.8 mm.

The net average shift of the upper eyelid peak toward the medial canthus was 1.3 mm ($P = .05$).

Larger reductions in proptosis resulted in a larger shift in the peak of the upper eyelid toward the medial canthus reference point.

Comment. Upper eyelid retraction and its lateral accentuation is one of the most common ophthalmic changes caused by Graves disease. The origin of this malposition is multifactorial; it may be caused by overaction of the Müller muscle or the levator muscle, by levator fibrosis and adhesions, or by axial proptosis of the globe.

With axial globe displacement, the lateral sclera projects further beyond the plane of the anterior orbital rim than the medial sclera, accentuating the lateral flare in eyelid retraction. When these anatomical factors are combined with a retracted upper eyelid position due to shortened or fibrotic retractors, the greatest effect is on the portion of the eyelid farthest beyond the plane of the anterior entrance to the orbit. The phenomenon of increased lateral retraction occurs, in part, because the eye in primary position looks medially away from the orbital axis and projects more lateral sclera.

In 1998, van den Bosch et al demonstrated that eyelid position was directly affected by globe position. Eyelid contour was straighter in patients who underwent upper eyelid recession surgery and decompression surgery than in the nondecompressed population.

Several confounding factors from the study by van den Bosch et al were addressed in our study to demonstrate that lid position and lateral flare are a direct function of globe position. To generate an axial reduction in proptosis and avoid hypoglobus, decompression surgery was performed on the medial wall and inferomedial floor of the orbit, with preservation of the thick bony strut between the maxillary sinus and the ethmoid sinus at the maxillo-ethmoid junction. Eyelid measurements were taken before and after decompression surgery, prior to any adjunctive lid recession procedures. Precautions were taken to as-
sure consistency in photography, and automated image analysis software was used to minimize human measurement errors and investigator bias. Even with these precautions, however, it is difficult to address the fact that lid height largely depends on patient effort and that such effort is highly variable.

Reducing axial proptosis of the eye with decompression surgery shifted the peak of the upper eyelid toward the medial canthus an average of 1.3 mm. More significantly, patients demonstrated an average 1.71-mm reduction in the height of the palpebral fissure and a 56.2-mm² reduction in the palpebral fissure surface area. The amount of shift in the lid peak toward the medial canthus is a function of the reduction in proptosis after decompression surgery (Figure 3). Reduction in proptosis after surgery also results in a corresponding decrease in the exposed ocular surface area (Figure 4).

Therefore, performing decompression surgery prior to any adjunctive lid recession procedures may significantly reduce lid retraction, lateral flare, and ocular surface area exposure, depending on the amount of reduction in proptosis. Patients who experience large reductions in proptosis after decompression surgery may attain a more natural and functional eyelid and palpebral fissure appearance without undergoing other adjunctive procedures. Consequently, decompression surgery should be considered before any other lid surgery in the rehabilitation of any patient with Graves disease, not just in patients with optic neuropathy, extreme proptosis, or exposure keratopathy.

Eli L. Chang, MD
C. Robert Bernardino, MD
Peter A. D. Rubin, MD

Financial Disclosure: None.
Correspondence: Dr Chang, Doheny Eye Institute, 1450 San Pablo St, DEI 4705, Los Angeles, CA 90033 (echang@dohenyyeinstein.org).


Cortical Blindness Due to Reversible Posterior Leukoencephalopathy Syndrome in a Patient With Thrombotic Thrombocytopenic Purpura and Preeclampsia

Neurological involvement in thrombotic thrombocytopenic purpura (TTP) is frequent. In one series, magnetic resonance imaging revealed brain lesions in 88% of the patients with TTP. The 2 most common cerebral lesions associated with TTP are edema and infarction. Cerebral edema predominantly affects the white matter, but when it affects gray matter in the territory of the posterior cerebral circulation, it may resemble the radiological findings of reversible posterior leukoencephalopathy syndrome (RPLS). We report a case of reversible cortical blindness caused by RPLS in a patient with TTP exacerbation and preeclampsia. To our knowledge, there have been only 10 previously reported cases of RPLS in the setting of TTP. We also discuss the role of diffusion-weighted imaging (DWI) in differentiating reversible from irreversible ischemic lesions.

Case Report. A 19-year-old, 28-week pregnant white woman came to the hospital with gross hematuria and decreased urine output. Her medical history was significant for recurrent episodes of TTP. Her pregnancy was complicated by 3 prior admissions for TTP exacerbation. Each time she responded to fresh frozen plasma transfusion. Her medications included iron supplements and prenatal vitamins.

At admission her blood pressure was 175/84 mm Hg, and she was afibrile. The remainder of the physical examination results were unremarkable. Laboratory testing re-