Glaucoma drainage devices are currently used to manage high-risk, complicated, adult and pediatric glaucoma when standard filtration surgery with an antimetabolite is unsuccessful.1 The drainage device consists of a plate and a tube. The tube is directly implanted either into the anterior chamber or through the pars plana in eyes undergoing vitrectomy. Tube coverage is imperative to prevent conjunctival erosion, which would lead to tube exposure and pose a risk for the development of endophthalmitis.

Sclera, dura, fascia lata, and pericardium have been employed to cover the tube and fistula sites.2,3 Raviv et al4 recently published safety data in a study of 44 patients (44 eyes) who had a pericardial patch graft placed to cover a glaucoma drainage device. In their retrospective study, they reported 5 cases of asymptomatic thinning of the pericardial patch graft without evidence of tube erosion. No cases of infection were reported. The mean ± SD follow-up was 10.2 ± 4.0 months. Herein, we report 2 cases of tube erosion through the conjunctiva following use of commercially prepared pericardial patch grafts occurring 7 and 8 months postoperatively.

Report of Cases. Case 1. A 74-year-old African American woman with a long-standing history of primary open-angle glaucoma in both eyes underwent an uncomplicated insertion of a 350-mm2 Baerveldt glaucoma implant (Pharmacia Upjohn Co, Kalamazoo, Mich) in the right eye with a pericardial patch graft through a fornix-based conjunctival incision. One month later, because of conjunctival retraction with exposure of the tube and fistula site, she underwent exchange of the original pericardial patch graft for a larger pericardial patch graft as well as a conjunctival autograft. The original pericardial patch graft appeared intact but was replaced to provide better coverage of the insertion site.

Three months later the patient was found to have asymptomatic melting of the pericardial patch graft without conjunctival erosion. Four months later the patient returned complaining of foreign body sensation and mild ocular tenderness. On examination, her visual acuity was 20/40 + 1 OD. Slitlamp examination revealed trace conjunctival hyperemia with a 2-mm area of conjunctival erosion and tube exposure. There was no visible pericardial graft tissue in the subconjunctival space. The cornea was clear. The anterior chamber demonstrated 1+ cells and flare without hypopyon. There were no vitreous cells. Fundus examination findings were unremarkable.

The patient was immediately treated with a topical fluorouracil (ciprofloxacin) hourly for presumed early endophthalmitis. The following day she underwent repositioning of the tube, which was covered with a 5 × 5-mm piece of full-thickness donor sclera. The conjunctiva was mobilized from the superonasal quadrant to cover the scleral patch graft. The patient did well with complete resolution of the anterior chamber reaction. The topical ciprofloxa-cin antibiotics and prednisolone acetate were tapered. All cultures continued to yield no organisms. Fifteen months after surgery, her best corrected visual acuity was 20/25 -1 OD, the overlying conjunctival intact, and the scleral patch graft had no observable thinning.

Case 2. An 89-year-old African American woman with a long-standing history of primary open-angle glaucoma in both eyes and multiple ocular surgeries underwent insertion of an Ahmed glaucoma valve (New World Medical Inc, Rancho Cucamonga, Calif) in the left eye with a 4 × 4-mm pericardial patch graft through a fornix-based conjunctival incision. Three months after surgery the pericardial patch graft was no longer visible. Five months later she came to the office complaining of a 4-day history of pain and blurred vision in the left eye. On examination, her visual acuity had decreased from 20/30 to 20/400 OS. Slitlamp examination revealed diffuse conjunctival hyperemia and a full-thickness conjunctival defect overlying the tube 3 mm posterior to the limbus. No pericardial tissue was visible. There were no sutures at the site of the defect. The anterior chamber demonstrated 3+ cells and flare with a 2-mm layering hypopyon. There were 1+ anterior vitreous cells. Fundus examination findings were unremarkable.

She underwent a vitreous tap and intravitreal injection of vancomycin hydrochloride and ceftazidime as well as topical and intravenous administration of vancomycin and cefazidime for presumed endophthalmitis. Five days later she underwent surgical repair of the conjunctival defect. Intraoperatively, no remnants of the pericardial patch graft were visible. A 5 × 5-mm scleral patch graft was placed over the tube and covered with a conjunctival flap that was mobilized anteriorly and secured to the limbus. Eight months postoperatively, her visual acuity improved to a baseline acuity of 20/30 OS. The conjunctiva was completely intact, and there was no evidence of scleral patch graft thinning.

Comment. The use of cadaveric allografts has been gaining wider acceptance for use in ophthalmic surg-
surgery, especially for coverage of glaucoma drainage devices. Pericardial, dura mater, and fascia lata patch grafts have been used. The reported advantages with pericardial patch grafts include uniform size and quality, commercial availability without dependence on an eye bank, potentially lower costs, and a processing method that leads to enhanced immunologic safety and reduced risk of viral transmission.

The dehydration process leaves the graft cell free and without antigenic stimuli. Tissue sterilization with organic solvents as well as low-dose irradiation leads to the inactivation of potential infectious pathogens, including human immunodeficiency virus and Creutzfeldt-Jakob virus. Despite the favorable result reported by Raviv et al., these 2 cases demonstrate that progressive thinning of the pericardial patch graft may occur in patients without predisposing ocular and systemic factors, such as uveitis, or other systemic immunologic disorders. Furthermore, specific antigen-mediated thinning is unlikely to be a major cause of progressive graft resorption owing to the manner in which the pericardial tissue is processed, leaving the tissue virtually antigen free. Surgical factors, such as exposed sutures, tight conjunctiva, or tube malposition, were not contributory in either case. Both cases of erosion occurred at a site over the tube previously covered with a pericardial patch graft. Moreover, in both cases a fornix-based conjunctival flap with superior and temporal relaxing incisions was used at the time of initial surgery to allow tension-free conjunctival apposition to the limbus. Although a conjunctival autograft was used in the first patient owing to retraction, the original pericardial patch graft was still intact. Additionally, the new pericardial patch graft and conjunctival autograft were placed over the tube without tension. Thus, it is unlikely that graft melting and conjunctival erosion, which occurred 7 months later, were related to excessive tension overlaying the tube. Asymptomatic thinning of the graft may eventually lead to tube erosion with subsequent development of intraocular infection. Pericardial patch graft thinning was observed in both cases many months before the acute onset of presumed endophthalmitis.

In addition to these 2 cases, we have observed 7 other eyes with asymptomatic thinning of the pericardial patch graft. Currently, the appropriate mode of action in such cases is unknown, and judgment is reserved to the treating physician on a case-by-case basis.

The long-term safety of pericardial patch grafts for tube coverage is currently unknown. Without a prospective randomized study, the relative safety of pericardial vs other patch graft materials cannot be definitively determined. However, careful clinical observation may help identify potential problems with newer materials.

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Permanent Ligation of Double-Plate Molteno Implant Distal Tube to Control Late Hypotony

Hypotony is an uncommon complication to develop late after placement of a double-plate Molteno implant. Treatment to date for this has been limited to 2 options: permanent reocclusion of the proximal tube or removal of the tube from the anterior chamber. These methods have the disadvantage of eliminating the entire effect of the implant. The double-plate Molteno implant, however, has the potential advantage of reducing, not eliminating, drainage by stopping drainage to the distal plate. In the following case, we used a new technique, permanent ligation of the distal tube, which allowed treatment of late postoperative hypotony with long-term pressure control and minimal surgical morbidity.

To permanently ligate the distal tube, we incised the conjunctiva and Tenon layer in the space between the distal plate and the overlying rectus muscle. We then bluntly dissected under the muscle and located the tube, taking care to spare the fibrous capsule around the distal plate. Next, we tied a clove-hitch knot around the tube previously covered with a double-plate Molteno implant undergoing ligation of distal tube. A, Proximal tube in anterior chamber; B, area over proximal plate; C, incision exposing rectus muscle and underlying distal tube; D, superior or inferior rectus muscle; E, exposed distal tube ligated with 6-0 polypropylene suture; F, area over distal plate.

Report of a Case. We implanted a Molteno double-plate drainage device with adjunctive intraoperative mitomycin C in the left eye of a 17-year-old woman who had a history of congenital ocular tuberous sclerosis (congenital glaucoma, aphakia, and high hyperopia). A previous trabeculectomy with mitomycin C in that eye had failed. To prevent early postoperative hypotony, we placed a 6-0 polypropylene suture in the proximal tube and ligated the tube with a 6-0 polypropylene suture. After
8 weeks, we removed the intraluminal suture, and her intraocular pressure (IOP) decreased from 28 to 7 mm Hg without medication. For the first 4 months after removing the intraluminal suture, her IOP ranged from 14 to 16 mm Hg, and her visual acuity remained at 20/50 OS.

Six months after removing the intraluminal suture, the patient complained of fluctuating, decreased visual acuity. Her visual acuity was 20/200 OS. Her IOP was 7 mm Hg. Fundus examination revealed horizontal macular folds. We diagnosed hypotony with maculopathy and ligated the distal tube of her Molteno implant. In the next several months, her visual acuity improved to 20/70 OS. Her IOP ranged from 18 to 19 mm Hg with treatment of 1 drop (approximately 20 µL) per day of timolol maleate, and her macular folds substantially decreased.

Comment. To halt and reverse the progression of complications associated with hypotony requires elevation of IOP. Comparison of glaucoma implants in rabbits has shown that the amount of filtration relates to the surface area available for filtration. Consistent with this observation, it has been observed that single-plate Molteno implants result in higher IOP and fewer complications related to hypotony than double-plate implants. Ligation of the distal tube of a double-plate Molteno implant, therefore, raises IOP and treats hypotony because it halves the surface area available for filtration, and yet it still allows filtration through the proximal plate in those situations where some drainage is required.

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Incomplete Spontaneous Regression of Choroidal Melanoma Associated With Inflammation

Spontaneous regression of cancer is a rare occurrence that has been documented with several cancers, including cutaneous melanoma. In cutaneous melanoma the regression is typically partial, occasionally displaying transient dermal inflammation with subsequent pink-gray atrophic changes. Rarely does cutaneous melanoma spontaneously disappear completely, and relapse is common.

Observations of spontaneous regression of choroidal melanoma are exceedingly rare. Of over 8000 patients with choroidal melanoma cared for at the Ocular Oncology Service at Wills Eye Hospital, Philadelphia, Pa, we have personally witnessed and photographically documented spontaneous regression in only 3 patients. We suspect that there may be additional cases of tumor regression that remain subclinical with partial necrosis, not appreciated on ophthalmoscopy or ultrasonography.

Report of Cases. Case 1. A 60-year-old man noted redness, irritation, and pain in his left eye of 1 day’s duration in March 1998 (Figure). Funduscopic examination revealed a superonasal, juxtapapillary pigmented choroidal mass measuring 14.5 mm at the base and 7.6 mm in thickness. Ocular B-scan ultrasonography revealed an acoustically hollow choroidal mass with overlying episcleral edema. A periocular corticosteroid injection was administered for relief of inflammatory pain.

In April 1998, a strikingly smaller choroidal mass measuring 5.3 mm in thickness was noted. In May 1998, the mass continued to decrease to 3.5 mm in thickness and on referral to us in June 1998, it was 3.2 mm thick. Evidence of prior subretinal fluid with retinal pigment epithelial mottling of the inferonasal fundus was found. Our diagnosis was choroidal melanoma with partial spontaneous regression. Options for treatment included continued observation, radiotherapy, or enucleation. The tumor was treated with a notched radioactive plaque and supplemental thermotherapy.

Case 2. A 45-year-old man noted redness, irritation, and pain in his left eye of 3 days in September 1995. Superonasal conjunctival chemosis and episcleral hyperemia were noted. Funduscopic examination revealed a minimally pigmented choroidal mass measuring 6.0 mm at the base and 2.2 mm in thickness, with an overlying serous retinal detachment. On ultrasonography, the mass showed acoustic solidity, choroidal excavation, and echolucency in the overlying episcleral tissue. The patient returned 2 months later without pain or chemosis; the choroidal mass had flattened to 1.2 mm in thickness, and the serous retinal detachment had resolved.

In April 1996, the choroidal mass had enlarged to 2.8 mm in thickness, and the patient was referred to us in September 1996. On examination, the left fundus demonstrated a lightly pigmented choroidal mass measuring 11.0 mm at the base with prominent subretinal fluid. A necrotic central area within the tumor surrounded by a viable peripheral rim was noted. Ultrasonography revealed an excavated, acoustically hollow mass measuring 4.0 mm in thickness. The diagnosis of spontaneously regressed choroidal melanoma with recurrence was made. The tumor was treated with plaque radiotherapy.

Case 3. A 78-year-old woman with Alzheimer disease developed ocular redness, swelling, and extreme pain in her right eye in June 1998. Funduscopic examination revealed a total bullous serosanguineous retinal detachment with shifting subretinal fluid. A large pigmented choroidal mass measuring 20.0 mm at the base and 11.7 mm in thickness was seen. Ocular ultrasonography confirmed a mushroom-shaped ciliochoroidal mass with intrinsic vascular pulsations, consistent with melanoma. Enucleation was advised. At the time
of enucleation, 8 days later, the inflammatory symptoms had completely resolved leaving the patient comfortable, without eyelid or conjunctival edema. On ophthalmoscopy, the tumor was dramatically smaller, measuring 8.5 mm in thickness. We suspected that the melanoma had undergone partial spontaneous regression.

Histopathologically, the enucleated globe demonstrated a highly necrotic ciliochoroidal malignant melanoma. The necrotic cells were rimmed by macrophages, and the viable cells demonstrated features of spindle and epithelioid malignant melanoma cells.

Comment. Spontaneous regression of cancer is a remarkable but rarely observed phenomenon, estimated to occur in 1 in 80,000 to 100,000 cases of cancer. It is defined as the complete or partial disappearance of a neoplasm in the absence of treatment. Spontaneous regression can occur with many systemic cancers, but it has not been correlated with complete tumor cure as most will ultimately recur. In a major review of all of the literature on 504 cases of spontaneously regressed cancer from 1966 to 1987, Challis and Stam found the primary cancer sites to be cutaneous malignant melanoma in 14%, renal cell carcinoma in 13%, lymphoma in 13%, leukemia in 11%, neuroblastoma in 8%, retinoblastoma in 6%, and breast cancer in 4%.

From the standpoint of cutaneous melanoma, it is believed that some degree of spontaneous regression is detected in 10% to 30% of cases. The clinical features suggesting regression are the development of depigmented areas and inflammation. The histopathologic appearance of cutaneous melanoma regression includes degeneration of tumor cells, lymphohistiocytic infiltrate, pigment-laden macrophages, dermal fibrosis, and epidermal atrophy. The prognostic significance of spontaneous regression with cutaneous melanoma is unclear, but some investigators feel it is associated with a worse prognosis. Sondergaard and Hou-Jensen found that stage I cutaneous melanoma has a 95% 10-year survival rate in those without regression and a 79% 10-year survival rate in those with spontaneous regression. In unusual circumstances, the primary site of cutaneous melanoma completely regresses leaving no clinical trace of tumor, but these patients are still at risk for metastatic melanoma.

Spontaneous regression of choroidal melanoma has been recognized, often masquerading as scleritis. Unfortunately, the regressed tumor often relapses. In this report we document photographically the clinical features of 3 patients, all of whom had ocular inflammation. The rate of tumor regression varied from...
Intravascular Papillary Endothelial Hyperplasia With Presumed Bilateral Orbital Varices

Intravascular papillary endothelial hyperplasia (IPEH) is an unusual condition characterized by a benign proliferation of vascular endothelial cells that form papillary projections in the lumen of a blood vessel. It is not a specific neoplasm, but is generally believed to be a reactive response that develops secondarily to a thrombus in vascular lesions such as varices or hemangiomas. It most often occurs as a painless, redish purple lesion in the dermis or subcutis of the head and neck region or extremities. Intravascular papillary endothelial hyperplasia has been recognized to occur rarely in the ocular area, usually in the eyelid and less often in the orbit. In reported ocular cases, IPEH has occurred in association with a unilateral, solitary vascular lesion. We report a case of IPEH that developed in association with multiple bilateral orbital vascular tumors, thus expanding the known ocular spectrum of this condition.

Report of a Case. In August 1991, an 80-year-old woman was seen with a 6-month history of slowly progressive, painless proptosis of the right eye. She had a history of successfully treated uterine cervical cancer many years earlier and a cataract extraction in the right eye 6 years earlier, but had no other systemic or ocular problems. Specifically, she had no history of trauma, ocular inflammation, cutaneous hemangioma, or prior episodes of proptosis.

Her visual acuity was 20/20 OD and 20/30 OS and intraocular pressures were normal. There was bilateral fullness of the upper and lower eyelids and 5 mm of proptosis of the right eye (Figure 1). The proptosis was not exacerbated by head position or Valsalva maneuver. Most of the fullness seemed to be caused by anterior displacement of orbital fat, as there was no distinctly palpable mass. Ocular motility was normal. The remainder of her ocular examination revealed normal findings.

Orbital computed tomography disclosed bilateral orbital masses. In the right orbit there was an intraconal mass that measured 17 × 17 × 15 mm (Figure 2) and displaced the optic nerve superiorly. Lesions in the left orbit were not clearly delineated. However, on subsequent computed tomographic examination, a second separate tumor, measuring 12 × 12 × 12 mm, was noted in the region of the superior orbital fissure of the right orbit with posterior extension into the middle cranial fossa (Figure 3). In the left orbit was an irregular mass nasal to the globe with ill-defined extension toward the orbital apex (Figure 3). Our differential diagnosis included metastatic carcinoma, lymphoma, multiple orbital cavernous hemangiomas, and orbital varices.

We elected to perform an excisional biopsy of the larger, symptomatic, intraconal mass in the right orbit by a superolateral orbitotomy through a cutaneous incision with an extraperiosteal approach and lateral osteotomy (Kronlein approach). A reddish blue mass was identified and removed intact. The patient had an uneventful postoperative course.

Pathologic Findings. Grossly, the slightly deflated, irregular, dark-red mass measured 17 × 15 × 10 mm. Microscopically, the lesion was a vascular mass composed of large, thin-walled, blood-filled channels lined by benign endothelial cells. There were localized areas of chronic inflammation. Approximately 70% of the mass was composed of slender fibrous trabeculae lined by benign endothelial cells that projected into the lumen of large vascular channels (Figure 4). The final diagnosis was IPEH, arising in a thomboosed varix.

Three years after surgery, the patient’s visual acuity was 20/40 OD and 20/30 OS. There was 2 mm of right proptosis and slight pallor of the right optic disc. Orbital computed tomography disclosed that all of the remaining lesions were stable.

Comment. Intravascular papillary endothelial hyperplasia usually occurs in the head and neck region and the extremities. Ocular involve-
ment is exceptionally rare. In the series of 44 cases reported by Clear-
klin and Enzinger,4 17 cases reported by Kuo et al,5 and the 91 cases re-
ported by Hashimoto et al,6 there was no mention of specific lesions in the
eyelids or orbit.

To our knowledge, the first re-
ported bona fide case of eyelid IPEH
was by Wolter and Lewis7 in 1974.
Although these authors cited 3 prior
eyelid cases, these subsequently were
not accepted by Font et al,9 who con-
sidered the patient of Wolter and
Lewis7 to be the first acceptable case.
Additional eyelid cases were re-
corded by Sorenson et al8 and Font
compiled the experience of several
surgeons and added 4 eyelid cases,
one of which extended to the lat-
eral orbital rim.

Even fewer cases of deeper or-
bital involvement with IPEH have
been recorded. The first case was re-
ported by Weber and Babel11 in
orbital cases from the files of the
Armed Forces Institute of Pathology.
A similar histopathologic re-
response was also observed in a pa-
tient with a variant of the Sturge-
Weber syndrome.13 Based on the
aforementioned reports, it seems that
there have been approximately 8 re-
ported cases of eyelid IPEH and 5 re-
ported cases of orbital IPEH in the
English-language literature.

Our case of IPEH is unusual
for 2 reasons. First, it occurred in
bilateral orbital vascular lesions,
presumably varices. Second, it was
associated with another lesion that
extended through the superior
orbital fissure into the cranial cav-
ity. Recently, a few cases of IPEH
have been recognized to arise from
intracranial vascular lesions, and
these were summarized by Werner
et al.10 It has also been recognized
in the region of the superior orbital
fissure,14 as occurred in our case.

Based on the histopathologic
findings, the IPEH in our patient
probably arose in a thrombosed
varix. Orbital varices usually are pri-
mary lesions, but they occasionally
can arise secondary to an intracra-
trial arteriovenous communication
that shunts arterial blood to the ve-
nous system, causing secondary di-
lation of orbital veins.7 The fact that
the vascular lesions in our patient
were bilateral and multiple raised the
possibility of secondary orbital vari-
ces. However, our patient had no his-
tory of trauma or clinical signs of ca-
rotid-cavernous fistula. Therefore,
the pathogenesis of the underlying
vascular lesion in our case remains
obscure. It is possible that the pre-
sumed varices in our patient had
been present for many years and that
IPEH was a response to thrombosis
in the larger lesion in the right or-
In summary, this case demonstrates that orbital IPEH can occur in patients with multiple orbital vascular tumors, as well as solitary lesions. Intravascular papillary endothelial hyperplasia should be included in the differential diagnosis of acquired progressive proptosis in adults.

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