We report 2 separate occurrences of lone-star tick bite to the conjunctiva. Both occurred within a 100-mile radius during the summer of 2000. A search of the literature yielded 2 reports of conjunctival tick bite. In one of these, the tick was removed with difficulty using a cotton-tipped applicator. We propose a simple, yet effective, method of removal.

**Case Reports.**

**Case 1.** On July 9, 2000, a 5-year-old girl was evaluated by her physician for an unidentified “spot” on her right eye. A tick and the surrounding area of erythema were identified in the conjunctiva temporally in the right eye (Figure). The remainder of the ocular examination findings were normal. Following referral to Arkansas Children’s Hospital (Little Rock), conscious sedation with ketamine and midazolam allowed the complete removal of the tick and a small amount of the surrounding conjunctiva with forceps and Westcott scissors. Two weeks later, a follow-up telephone call revealed the patient to be doing well, having been seen twice by her personal ophthalmologist.

**Case 2.** On August 8, 2000, a 2-year-old girl was seen in the emergency department for evaluation of tick bites. An ocular foreign body prompted an ophthalmological consultation and identification of a tick attached to the conjunctiva of her left eye. Conscious sedation with ketamine and midazolam allowed the complete removal of the tick and surrounding conjunctiva with forceps and Westcott scissors. One week later, there was no sign of infection or other abnormality.

**Comment.** The lone-star tick, identified in these 2 cases, is the common name for *Amblyomma americanum*. The life cycle is composed of the egg, larva, nymph, and adult stages of development. The egg hatches into a 6-legged larva (“seed” tick), which attaches to a host and feeds. The larva then drops off the host and metamorphoses into an 8-legged nymph. The nymph reattaches to feed and later metamorphoses into an adult. The adult is differentiated into male and female.

The distinctive morphological features of the species of *Amblyomma* were described by G. Neumann in 1896. The female tick is larger than the male counterpart. On the scutum, or dorsal hard plate, of both the male and female are intermittent white spots, hence the name “lone-star tick.” These spots are typically more prominent on the female than on the male. The female can have red and green markings in addition.

*A. americanum* is known to be a transmitter of diseases to domestic animals and to humans. Published reports by Maria Maver (1911) of Rocky Mountain spotted fever rickettsia transmission by *A. americanum* in guinea pigs led to the hypothesis that spotted fever could be transmitted to humans by this tick vector. In 1943, extraction of spotted fever rickettsia from an *A. americanum* nymph was reported. *Amblyomma* has also been demonstrated to be a carrier of tularemia and an erythema migrans–like rash illness similar to Lyme disease. As a known carrier of a number of diseases, *A. americanum* poses a threat to humans. It probably accounts for most tick infestations in the United States, especially in the south central states. Complete removal is thought to lessen the potential for transmission.

As activities move to the outdoors during the summer months, tick bites, especially on exposed areas of the body, may occur even after a short time in wooded areas. At least 4 hours of tick attachment are thought to be necessary for spotted fever rickettsia transmission in humans. Preventive measures include complete removal of the tick; care must be taken not to leave mouth parts in the skin or to divide the tick’s body. Residual crushed tissue and feces can also transmit disease. In the past, to avoid rupture or incomplete removal of the tick, lindane shampoo, deodorized kero-
sene, ether, or iodine were used.

Since the tick bites we report involved the conjunctiva, mechanical extraction was the procedure of choice. We add our cases of conjunctival tick bite to the literature with a suggested method for removal.

Monica C. Love, BA
Little Rock, Ark
Lucas Platt, MD
Rogers, Ark
Christopher T. Westfall, MD
Little Rock

This work was supported in part by an unrestricted grant from Research to Prevent Blindness, New York, NY.

Corresponding author: Christopher T. Westfall, MD, 4301 W Markham, Mail Slot 523, Little Rock, AR 72205 (e-mail: westfallchristopher@exchange.uams.edu).

4. Kirkland KB, Klimko TB. Erythema migrans.

A Case of Atypical WAGR Syndrome With Anterior Segment Anomaly and Microphthalmos

Wilms tumor, aniridia, genitourinary anomalies, and mental retardation (WAGR syndrome) are caused by the deletion of chromosome 11p13, which includes the Wilms tumor gene (WT1) and the aniridia gene (PAX6) loci (MEM, No. 194070). We report a case of atypical WAGR syndrome with anterior segment anomaly and microphthalmos.

Report of a Case. A 1-month-old boy had microphthalmos bilaterally. A microcornea with a corneal cyst in the right eye (axial length, 14.4 mm) (Figure 1A) and corneal opacity and absent anterior chamber in the left eye (axial length, 21.0 mm) (Figure 1B) seemed to be part of an anterior segment anomaly that includes the Peter anomaly. The vitreous cavities and posterior segments were normal. We examined the right eye with a small contact lens and light stimuli and obtained a normal response on the electroretinogram and in the left eye a subnormal response, suggesting retinal dysfunction. Wilms tumors developed bilaterally when the patient was 3 years old (Figure 1C). Because of a large tumor, we resected the right kidney; the left kidney underwent chemotherapy. The resected tumor had predominantly blastemal cells (Figure 1D). The child also had undescended testes and mental retardation. Analysis of G-banded metaphase chromosomes identified deletion of chromosome 11p13-15.1 in 1 allele (Figure 2). Chromosomal analysis and physical findings were compatible with WAGR syndrome, but the ocular findings differed substantially.

Comment. Since the PAX6 gene was identified as a candidate gene for aniridia, numerous mutations of 11p13 have been reported in patients with aniridia. Studies have identified Pax6 mutations in numerous ocular anomalies, including the Peter anomaly, congenital cataract, and lenteal hypoplasia. In situ hybridization and immunohistologic examination identified multiple functions of the gene; the gene moves from the anterior to the posterior segments of the eye throughout development. Therefore, it is not surprising that ocular anomalies other than aniridia result from deletion of 11p13. Two other patients were described...
Previously: a 2-month-old boy with a Peter anomaly and deletion of 11p13 who did not have Wilms tumor and a 6-day-old boy with a Peter anomaly and Wilms tumor in whom chromosomal analysis was not undertaken. A 3-month-old boy with duplication of 11p13 had microphthalmia. Genetically similar mice with multiple copies of Pax6 have a microphthalmic phenotype, indicating gene dosage affects normal function.

PAX6 mutations in aniridia are usually nonsense, frameshift, or splicing errors in 1 allele, which result in a truncated protein, thus, haploinsufficiency of the gene products causes the aniridia phenotype, while few mutations in the Peter anomaly are missense. However, missense mutations recently found in patients with aniridia produce another route of haploinsufficiency. Thus, the relation between PAX6 gene dosage and phenotypic manifestations is still controversial. Affected individuals in a pedigree with aniridia who had the same PAX6 mutations have wide phenotypic variations. Because the PAX6 gene is influential in numerous ocular tissues throughout development, phenotypes may be reflected modifiers unlinked to the PAX6 gene cascade, cofactors of PAX6, or environmental conditions. Although aniridia and anterior segment anomalies including the Peter anomaly are distinct clinical entities, in our patient, aniridia resulting from deletion of one copy of PAX6 may be modified by these other factors and manifest corneal opacity, iridocorneal adhesion, and microphthalmos.

Eriko Kawase, MD
Kiyoshi Tanaka, MD
Toshirou Honna, MD
Noriyuki Azuma, MD
Tokyo, Japan

The authors have no proprietary interest in any aspect of this report.

Corresponding author: Noriyuki Azuma, MD, Department of Ophthalmology, National Children's Hospital, 3-35-31 Taishido, Setagaya-ku Tokyo, 154-8509 Japan (e-mail: nazuma@nch.go.jp).


**Multiple Evanescent White Dot Syndrome Following Hepatitis A Vaccination**

The multiple evanescent white dot syndrome (MEWDS) is an inflam-

![Figure 2. The G-banded prometaphase chromosomes show deletion of chromosome 11p13-15.1. The (p) indicates the short arm of the chromosome; (q), the long arm.](http://archopht.jamanetwork.com/pdfaccess.ashx?url=/data/journals/ophth/6752/)
On examination, best-corrected visual acuity was 20/20 in the right eye and 20/25-2 in the left eye. A relative afferent pupillary defect and decreased color vision were present in the left eye. Examination of the left posterior segment disclosed mild optic disc edema, a shallow serous macular detachment, and multiple, faint, yellow-white lesions that affected the outer retina (Figure, A-B). The results of examination of the right posterior segment were unremarkable. A Humphrey 30-2 visual field test showed marked enlargement of the blind spot (Figure, C) and decreased foveal sensitivity. A fluorescein angiogram showed late leakage from the disc and faint late hyperfluorescence of the outer retinal lesions.

Three days after the onset of symptoms, vision decreased to 20/40 in the affected eye, and the discrete, yellow-white outer retinal spots were more numerous and pronounced. At 10 days, the visual acuity remained 20/40, the afferent pupillary defect had disappeared, the blind spot had decreased in size, and a second posterior segment examination disclosed persistent, although milder, optic disc edema, with partial resolution of the macular serous detachment and disappearance of the outer retinal spots. Six weeks after initial examination, the patient’s vision had returned to 20/20 and the optic disc edema and serous detachment had completely resolved. Subsequent Humphrey visual field examination results were normal. One year after initial examination, vision remained 20/20 for each eye and retinal examination results were normal, although subjective nyctalopia persisted in the affected eye.

Comment. We present the first case, to our knowledge, of MEWDS following a booster HAVV. Our patient sought care 13 days after the vaccine because of unilateral loss of vision, photopsias, an enlarged blind spot, optic disc edema, and multiple yellow-white dots at the level of the outer retina, all findings that are consistent with the diagnosis of MEWDS. Symptoms, visual acuity, visual field changes, and fundus abnormalities all returned to normal within 6 weeks.

The cause of MEWDS is unknown, although both infectious and immune-mediated origins have been proposed. Jampol et al.1 in the original description of this syndrome, reported an antecedent flulike illness in 50% of patients. Others, in contrast, have cited the presence of increased levels of circulating immunoglobulins in the acute phase of MEWDS2 and the occurrence of MEWDS following hepatitis B vaccination3 in support of an immune basis for this disorder. Although it is possible that the occurrence of visual symptoms following HAVV was coincidental, this association observed in our patient would seem to support the role of an immune mechanism in the pathogenesis of MEWDS, particularly since hepatitis A vaccination utilizes inactivated virus. An immune mechanism has also been implicated in the pathogenesis of acute posterior multifocal placoid pigment epitheliopathy,4 which, like MEWDS,5 has been observed following inactivated hepatitis B vaccination.6

Laura Fine, MD
Andrew Fine, MD, MPH
Emmett T. Cunningham, Jr, MD, PhD, MPH
San Francisco, Calif

This work was supported in part by a Career Development Award from Research to Prevent Blindness Inc, New York, NY (Dr Cunningham).

Corresponding author: Emmett T. Cunningham, Jr, MD, PhD, MPH, The Pearl and Samuel J. Kimura Ocular Immunology Laboratory, The Francis I. Proctor Foundation, UCSF Medical Center, San Francisco, CA 94143-0944 (e-mail: emmett_cunningham@yahoo.com).

7. Park D, Schatz H, McDonald HR, Johnson RN. Acute multifocal posterior placoid pigment epi-
0.01% Becaplermin Gel for the Treatment of a Chronic Orbital Ulcer After Exenteration

Chronic orbital epithelial defect is a rare complication that occurs after exenteration. Skin grafts, rotation flaps, or free flaps are the traditional surgical means to correct such abnormalities. We describe a case of a chronic orbital epithelial defect after exenteration refractory to conventional treatment that resolved after being treated with 0.01% topical becaplermin gel (recombinant human platelet-derived growth factor BB [rh-PDGF-BB] or Regranex [Ortho-McNeil Pharmaceuticals Inc, Raritan, NJ]).

Report of a Case. A 57-year-old African American patient with a history of hypertension, seizures, and diabetes underwent a subtotal exenteration with placement of a full-thickness skin graft for primary orbital melanoma. The orbital defect healed slowly for several weeks. A year later the patient underwent placement of craniofacial orbital implants for retention of an orbital prosthesis. Following this procedure, she developed an ulcer at the apex of the socket that measured 15 x 4 mm with elevated edges and what appeared to be granulation tissue at the base of the ulcer (Figure 1). A biopsy specimen of the area demonstrated chronic inflammatory changes. The ulcer was treated with hydrogen peroxide soaks, wet-to-dry dressings twice a day, and discontinued use of the prosthesis (as allowed). The prosthesis was refitted to minimize the pressure at the orbital apex. A course of topical antibiotic cream was also applied. The ulcer, however, did not resolve for 15 months. Discharge was not present and cultures were not obtained. Treatment was then initiated with 0.01% topical becaplermin gel daily, which was the only modification in the treatment regimen. The ulcer gradually decreased in size and was completely healed after 3 weeks (Figure 2). Use of the gel was discontinued a week later without a tapering dose. No recurrence had occurred with a follow-up period of 12 months.

Comment. Chronic epithelial defects after exenteration have been described after radiation to the orbit,1 after socket infection, and with diabetes (as occurred in our patient). Skin grafts in these conditions have a high chance of failure because of compromised blood supply. A temporalis rotation muscle flap offers a good chance of cure but requires additional surgery and may deform the temporalis fossa region. Free flaps result in scarring and deformity of the donor site and neck area, where the microvascular anastomosis is performed. Local treatment with agents that promote wound healing is therefore the most simple and cost-effective treatment when taking into account the cost of surgery.

Recombinant human platelet-derived growth factor BB is involved in regulation of all phases of normal wound healing. It is either synthesized or released from all cell types involved in the healing process.2,3 Both rh-PDGF-BB and other growth factor levels have been found to be reduced in chronic, nonhealing wounds.2 Exogenous administration of rh-PDGF-BB in the form of 0.01% becaplermin gel promotes wound healing and was found to be effective for the treatment of diabetic foot ulcer and pressure ulcers in multicenter, double-blind, placebo-controlled trials.4 It is currently the only growth factor approved by the Food and Drug Administration for the treatment of chronic diabetic foot ulcers.

This is the first reported case, to our knowledge, of clinical use of 0.01% becaplermin gel (Regranex) in the ophthalmic literature and suggests that this may be a powerful tool in the treatment of chronic epithelial defects in oculoplastic surgery. The Food and Drug Administration, however, has not yet approved becaplermin for treatment of conditions other than diabetic foot ulcers. Appropriate caution must be exercised by physicians who choose to use this medication.

Iftach Yassur, MD
Marc J. Hirschbein, MD
James W. Karesh, MD
Baltimore, Md

The authors have no commercial, proprietary, or financial interest in the product mentioned in the article.

Figure 1. The socket of the left eye before treatment with becaplermin. A 15 x 4-mm ulcer is present at the apex of the orbit. The 2 metal implants that hold the orbital prosthesis are seen at the roof of the orbit.

Figure 2. The same area 3 weeks after treatment with becaplermin.
Corresponding author: James W. Karesh, MD, Department of Ophthalmology, Krieger Eye Institute, Sinai Hospital of Baltimore, 2411 W Belvedere Ave, Baltimore, MD 21215 (e-mail: jkaresh@lifebridgehealth.org).


Progressive Growth of Benign Adenoma of the Pigment Epithelium of the Ciliary Body

A 77-year-old woman underwent iridocyclectomy for a progressively enlarging black mass that apparently originated in the ciliary body and secondarily invaded the iris of her pseudophakic left eye. At the time of surgery, the tumor was adherent to the intraocular lens, which was removed along with the mass. Histopathologically, the tumor was composed of islands and cords of benign pigment epithelial cells that were separated by vascularized fibrous connective tissue. The diagnosis was adenoma of the ciliary body pigment epithelium with secondary invasion of the iris. This case underscores the fact that benign intraocular tumors can show dramatic enlargement without undergoing malignant transformation.

The pigment epithelial layers of the eye often undergo reactive hyperplasia but true neoplasia of ocular pigment epithelia is rare.1-4 On occasion, however, true neoplasms can develop from the pigment epithelia of the iris, ciliary body, and retina.5-7 Tumors of the pigment epithelium generally are benign or low-grade malignancies and have no known tendency to metastasize. However, they can grow slowly and can be locally invasive.5-7 We report a clinicopathologic correlation of an adenoma of the ciliary body pigment epithelium that secondarily invaded the iris and developed into a sizable intraocular mass.

Report of a Case. In 1993, a 70-year-old woman was found on routine ocular examination to have an early cataract in her left eye. At that time, her irides were normal and no pigmented lesions were noted. In 1995, the cataract had progressed and a small pigmented lesion was first noticed in her left iris. This was interpreted as a melanocytic nevus. In 1996, the cataract had shown further progression and successful cataract extraction with placement of a posterior chamber intraocular lens was performed. Two years later, the iris lesion had shown no convincing change, but suspicion of possible melanoma prompted referral to the oncology service.

At the time of our initial evaluation in 1997, the corrected visual acuity was 20/20 OD (phakic eye) and 20/20 OS (pseudophakic eye). Intraocular pressure readings were normal. In the nasal aspect of the left iris was a black lesion that spared the pupillary margin and extended to involve the base of the iris between the 8:30- and 10-o’clock meridians (Figure 1). The mass appeared to be pushing anteriorly through the iris stroma from the ciliary body but it did not seem to arise from or infiltrate the stroma. Gonioscopy showed that the lesion obscured a view of the angle structures and there was fine pigment dusting in the angle inferiorly. Ultrasound biomicroscopy results revealed ciliary body thickening by the mass and transillumination disclosed a shadow that extended across the pars plicata for 1 mm into the pars plana. The favored diagnosis was adenoma of the ciliary body pigment epithelium with secondary extension through the iris. Because of her age and good visual acuity, it was elected to continue to follow the patient conservatively.

In 1998, the lesion was first noted to be displacing the intraocular lens. In 1999, the patient’s visual acuity had decreased to 20/40 OS and the intraocular lens appeared to be covered by fibrous tissue in the quadrant of the tumor. An episcleral sentinel blood vessel had developed over the tumor and the angle pigmentation inferiorly had become denser. By 2000, the iris component of the lesion had become larger (Figure 2) and ultrasonography showed the ciliary body component of the lesion to be 6 mm thick. With transillumination, the lesion measured 7 × 7 mm in diameter and extended more pos-
teriorly into the pars plana, almost reaching the ora serrata. Because of the progressive enlargement, the mass was resected by a partial lamellar iridogoniocyclochordopectomy. Firm adherence of the mass to the intraocular lens made them inseparable, necessitating removal of the lens along with the tumor. Postoperatively, there was diffuse vitreous hemorrhage, which slowly resolved during 3 months without the need for a vitrectomy.

Grossly, the black lesion had clear scleral margins. The mass partially encompassed the lens capsular bag, which contained an intraocular lens (Figure 3). Microscopically, the specimen consisted of a lamella of sclera on which rested a pigmented tumor that was passing through the thinned iris (Figure 4). The mass was composed of nests, cords, and islands of intensely pigmented epithelial cells that were separated by prominent septa of vascularized fibrous connective tissue (Figure 5). The cords were solid without evidence of duct formation. The melanin granules in the cytoplasm were large and generally spherical in configuration, resembling those seen in the normal pigment epithelium. Bleached sections showed that the tumor nuclei were generally round or oval with relatively well-dispersed chromatin and small to mildly conspicuous nucleoli (Figure 6). The nuclei were only mildly pleomorphic and only 2 mitotic figures were counted in 40 high-power fields. The posterior surgical margin was free of tumor. Fibrous tissue encompassed a groove in the tumor that contained lens cortical and capsular remnants and a space that was occupied by the intraocular lens haptic in vivo. This was surrounded by capsular fibrosis. There was mild invasion of the ciliary body stroma by the tumor. The final diagnosis was adenoma of the ciliary body pigment epithelium with secondary iris invasion.

Comment. Tumors of the ciliary body pigment epithelium are rare. Until recently, they were often misdiagnosed clinically as malignant melanoma. In our case, the diagnosis was suspected clinically because of the black color of the lesion. Tumors of the pigment epithelium traditionally have been considered to be relatively dormant but a recent study revealed that those in the ciliary body can cause vitreous hemorrhage, subluxation of the lens, and secondary cataract, and that those of the retinal pigment epithelium can be locally invasive and cause exudative retinal detachment. Conversely, tumors of the iris pigment epithelium tend to be more stationary and are less likely to exhibit local invasion. In a clinical series of 20 consecutive cases of adenoma of the iris pigment epithelium, only 2 required surgical removal and 18 remained stationary during the course of follow up. The tumor in our patient was initially suspected to be an iris lesion but detailed examination suggested that it probably originated in the pigment epithelium of the ciliary body. Docu-
mented growth of the tumor prompted removal and the diagnosis was confirmed histopathologically.

A remarkable histopathologic feature of the tumor reported here is that it was composed predominantly of solid cords and tubules of tumor cells. These findings are more consistent with reported cases of adenomas of the iris pigment epithelium. In contrast, tumors that arise from the ciliary body pigment epithelium are usually characterized by numerous clear vacuoles and have less connective tissue stroma. The tumor in our patient lacked such vacuoles. From a clinical standpoint, however, the tumor appeared to originate in the ciliary body and to secondarily invade the iris.

The tumor reported here showed progressive enlargement, secondarily invaded the iris, induced the development of a sentinel blood vessel, and displaced and partially adhered to the intracocular lens. Physicians should be aware that progressive growth of a ciliary body or iris mass does not necessarily imply malignant transformation. Ancillary measures such as gonioscopy, transillumination, and ultrasound biomicroscopy are often necessary to determine the size and extent of a ciliary body mass. Recognition of the characteristic features of adenoma of the ciliary body pigment epithelium should facilitate the diagnosis of this unusual tumor.

Jerry A. Shields, MD
Ralph C. Eagle, Jr, MD
Carol L. Shields, MD
Arun D. Singh, MD
Philadelphia, Pa
Paul F. Torrisi, MD
Syracuse, NY

This research was supported by the Eye Tumor Research Foundation, Philadelphia, the Award of Merit in Retina Research, Houston, Tex (Dr J. A. Shields), the Macula Foundation, New York, NY (Dr C. L. Shields), and by the Noel T. and Sara L. Simmonds Endowment for Ophthalmic Pathology, Wills Eye Hospital, Philadelphia (Dr Eagle).

We thank Anthony LaTessa, MD, and Richard A. Frio, OD, for their help in the management of the patient.

Corresponding author and reprints: Jerry A. Shields, MD, Oncology Service, Wills Eye Hospital, 900 Walnut St, Philadelphia, PA 19107.


Extramedullary Myeloid Cell Tumor in an Elderly Man

Extramedullary myeloid cell tumor (granulocytic sarcoma, chloroma) is a rare cause of proptosis that can masquerade as lymphoma. Proper distinction between the two allows appropriate radiation dosing. We demonstrate in a 72-year-old man we saw with epiphora, proptosis, binocular diplopia, and pain in the right eye that extramedullary myeloid cell tumor may be treated with radiation alone using as little as 450-rad (4.5 Gy) applied in small fractions. This approach spares the elderly patient from the general myelosuppression of chemotherapy.

Report of a Case. An otherwise healthy 72-year-old man developed epiphora, proptosis, binocular diplopia, and pain in his right eye over an 8-month period. Ocular and medical histories were noncontributory. Visual acuity was 20/50 OD and 20/40 OS. External examination revealed right-sided hypoglobus, exophthalmos, and a ruddy, red bulbar conjunctiva associated with a large orbital mass that was firm to the touch.

Figure 1. When first seen for evaluation, the patient had right-sided hypoglobus, exophthalmos, and a ruddy, red bulbar conjunctiva.
and the mass did not perform retro-pulsion, instead when pushed it was firm and moved back (Figure 1). Hertel exophthalmometry measurements were 23 mm OD and 13 mm OS with a base of 125 mm. Confrontational fields were full. Ocular motility was restricted in all fields of gaze in the right eye. Intraocular pressure was 21 mm Hg OD compared with 15 mm Hg OS. Slitlamp biomicroscopy revealed the conjunctiva of the right eye was severely congested and expanded by a solid, salmon-colored mass (Figure 2).

Orbital computed tomography (Figure 3) revealed a homogeneously enhancing mass in the region of the right upper eyelid. The mass molded to the lateral aspect of the globe, involved the tendinous insertion of the lateral rectus muscle, and extended to the intraconal space. A transconjunctival biopsy of the lesion was performed. Intraoperative touch preparations demonstrated a malignant hematolymphoid neoplasm. Portions of the tissue were placed in Zenker fixative and 10% buffered formalin for routine light microscopy. Additional specimens were submitted fresh for flow cytometry and cytogenetics. Histopathological examination findings demonstrated a solid sheet of monomorphous, intermediate to large cells filling the conjunctival stroma. Individual cells showed a high nuclear-cytoplasmic ratio, with only a scant rim of eosinophilic cytoplasm. Nuclei were hyperchromatic, irregularly contoured, and contained central nucleoli and dispersed chromatin (Figure 4). On immunohistochemical staining, the neoplastic cells were reactive to CD45 (leukocyte common antigen), CD43, and lysozyme (Figure 5) but did not react with CD3 (pan T-cell) and CD20 (pan B-cell) markers. Flow cytometry confirmed the myeloid nature of the neoplasm, as demonstrated by expression of CD33 tumor cells and a subset of CD34 tumor cells (Figure 6A). There was an absence of CD5 and CD20 cells (Figure 6B). The pathological diagnosis was extramedullary myeloid cell tumor.

Systemic evaluation showed no lymphadenopathy and findings from the patient’s peripheral blood smear were normal. No other extramyelogenous deposits were found on meta-
static workup. Therapy with 30 rad (0.3 Gy) of orbital irradiation in 15 fractions was accomplished over a 3-week period. All signs and symptoms were eliminated by 1 month (Figure 7). After 13 and again at 21 months, he reported no symptoms by telephone but declined to return for evaluation.

Comment. Extramedullary myeloid cell tumor (granulocytic sarcoma, chloroma) is a rare tumor composed of immature granulocytes. The disease may manifest in several different clinical settings. Most commonly it occurs in childhood, and most often in combination with pre-existing acute myelogenous leukemia. It also occurs as a harbinger of acute myelogenous leukemia in non-leukemic patients. Less often, it has been reported in conjunction with myelodysplastic disorders or chronic myelogenous leukemia. The majority of nonleukemic patients with extramedullary myeloid cell tumor will develop leukemia within a matter of months, but a delay of 16 years following initial evaluation has been observed. The tumor is exceedingly rare in the elderly.

Extramedullary sites of involvement are usually in extraocular tissues. Most commonly involved are bone, lymph nodes, periosteum, and skin. When ophthalmic manifestations occur, they are usually orbital. Signs and symptoms frequently include pain, proptosis, chemosis, and epiphora. Computed tomography frequently reveals a homogeneous, well-defined tumor that molds to surrounding bone.

Histopathological diagnosis of extramedullary myeloid cell tumor may be difficult to ascertain on routinely processed tissue sections, and it is often confused with malignant lymphoma. The distinction is important because successful treatment of lymphoma of the orbit and ocular adnexa requires 2400 to 4000 rad (24-40 Gy) of total irradiation. Immunophenotyping tumor cells either by flow cytometry or immunohistochemistry is efficacious in differentiating these 2 en-
Extramedullary myeloid cell tumors are positive for CD45 and CD43 and negative for other B-cell and T-cell markers. In addition, they usually show some degree of reactivity with antibodies to lysozyme or myeloperoxidase. By flow cytometry, tumor cells are CD33 positive and variably CD34 positive.

Prognosis and treatment of extramedullary myeloid cell tumor is dependent on the presence or lack of an associated leukemia or dysplasia, but in the absence of an associated systemic malignancy, the prognosis is indeterminate. In our case there was no identified associated malignancy. Treatment in such cases is not standardized. Frequently, chemotherapy and irradiation are used together and result in a rapid response. Irradiation alone, as in this case, may be efficacious in the treatment of a systemically non-leukemic patient with isolated orbital extramedullary myeloid cell tumor, and it will spare the elderly patient from the general myelosuppression that often accompanies chemotherapy.

Wade D. Brock, MD
Harry H. Brown, MD
Christopher T. Westfall, MD
Little Rock, Ark

This work was supported in part by an unrestricted grant from Research to Prevent Blindness, Inc, New York, NY.

Corresponding author and reprints: Christopher T. Westfall, MD, Harvey and Bernice Jones Eye Institute, Department of Ophthalmology, University of Arkansas for Medical Sciences, 4301 W Markham, Slot 523, Little Rock, AR 72205-7199.