Since its inception as a North American cooperative group in 1972, the Intergroup Rhabdomyosarcoma Study Group has undertaken a series of clinical trials that have employed chemotherapy, surgery, and irradiation for local control. The addition of systemic chemotherapy has markedly improved long-term survival in affected children. These children now have an excellent prognosis with current multimodality therapy. A recent review by Oberlin and associates demonstrates the 10-year overall survival rate to be 87% regardless of initial approach to therapy (chemotherapy alone or chemotherapy with local irradiation). In response to the superior survival rates for patients with isolated orbital rhabdomyosarcoma, achieved during the first 2 generations of North American cooperative studies, subsequent efforts have focused on reducing the short- and long-term morbidity associated with therapy while maintaining a very high cure rate.

Klippel-Trenaunay syndrome is a congenital vascular malformation characterized by 3 features: (1) a cutaneous vascular nevus (capillary malformation), (2) soft tissue or bony hypertrophy in the involved region, and (3) varicose veins or venous malformations. It is frequently associated with deep venous anomalies and lymphatic malformations. Common complications associated with KTS include pain in the affected extremity, thrombophlebitis, and cellulitis. Surgical therapy is ineffective, but pulsed-dye laser, hydrotherapy, and compression treatments can be helpful. The cause of KTS is unknown.

Numerous vascular lesions have been associated with KTS, but malignancies are extremely rare. Lymphatic and venous malformations can be found either ipsilateral or contralateral to the affected site. Orbital lymphatic malformation would not be unexpected in such a setting. In this case, the presence of a URTI at the time of the initial examination gave further support for this diagnosis. Head and neck lymphatic malformations, including orbital lesions, typically enlarge rapidly during respiratory tract infections, presumably owing to infectious stimulation of the aberrant lymphatic tissue. Results of radiographic studies were equally misleading: the lesion molded to the globe, there was no bony erosion, and there were areas of heterogeneity (but no frank cystic or hemorrhagic areas).

Corticosteroids are effective in reducing the size of inflamed lymphatic malformations in cases of spontaneous (nonhemorrhagic) enlargement or postoperative swelling and were, therefore, chosen as the initial therapy in this case. Medical treatment with early clinical reevaluation was chosen to avoid potentially unnecessary surgery.

Four recognized experts in their respective fields (orbital surgery, vascular dermatopathology, pediatric otolaryngology, and head and neck radiology) were misled by these signs and symptoms. In the setting of an expected, benign vascular tumor, one must exclude all potential serious and malignant diagnoses. As demonstrated by our case, rhabdomyosarcoma of the orbit can exhibit clinical and radiographic features that may be misconstrued as a benign, lymphatic malformation.

Aaron Fay, MD
Nicoletta Fynn-Thompson, MD
David Ebb, MD
Boston, Mass

The authors have no relevant financial interest in this article.

Corresponding author and reprints: Aaron Fay, MD, 243 Charles St, Boston, MA 02114 (e-mail: aaron_fay@meei.harvard.edu).


Secondary Chronic Open-Angle Glaucoma After Intravitreal Triamcinolone Acetonide

Intravitreal injections of triamcinolone acetonide have increasingly been used for treatment of various intraocular neovascular, proliferative, or edematous diseases, such as diffuse diabetic macular edema, proliferative diabetic retinopathy, proliferative vitreoretinopathy, chronic uveitis, and persistent pseudophakic cystoid macular edema. View of the widening spectrum of therapeutic indications of intravitreal triamcinolone acetonide, we report the clinical course of a patient who repeatedly received intravitreal injections of triamcinolone acetonide 14 months apart, who showed intravitreal triamcinolone acetonide crystals still present 9 months after the second injection, and who developed secondary open-angle glaucoma uncontrollable by topical antiglaucomatous medication.

Report of a Case. A 79-year-old woman sought treatment for progressive exudative age-related macular degeneration with subfoveal occult neovascularization in her left eye. Snellen chart visual acuity decreased from 0.80 to 0.50 OS with the accompanying complaint of marked metamorphopsia. Intraocular pressure measured 16 mm Hg, and the appearance of the optic nerve head was normal. The right eye demonstrated a large subfoveal disciform scar due to exudative macular degeneration covering the whole macular region be-
between the temporal vascular arcades. Visual acuity was reduced to hand movements OD. The intraocular pressure measurement was within the reference range. In an attempt to reduce subfoveal exudation and to suppress subretinal angiogenesis, the patient received a transconjunctival intravitreal injection of 25 mg of crystalline triamcinolone acetonide in 0.2 mL of Ringer solution in her left eye. The procedure was carried out under topical anesthesia after paracentesis was performed to decrease the volume of the globe. The patient was fully informed about the experimental nature of the therapy. She signed an informed consent form. The ethics committee of the university approved the study, which followed the tenets of the Declaration of Helsinki.

After the injection, the patient’s vision remained stable for 4 months. On indirect ophthalmoscopy as well as Goldmann contact lens ophthalmoscopy, the triamcinolone acetonide crystals were localized in the preretal vitreous cortex at the 6-o'clock position in the periphery of the fundus. Two months after the injection, intraocular pressure increased to as much as 32 mm Hg, necessitating antiglaucomatous topical treatment with latanoprost, timolol maleate, and dorzolamide hydrochloride, under which intraocular pressure normalized. Five months after the injection, visual acuity started to decrease. Simultaneously, the triamcinolone acetonide crystals were no longer detectable on indirect ophthalmoscopy and Goldmann contact lens ophthalmoscopy. Eight months after the injection, uncomplicated phacoemulsification was performed with implantation of a foldable posterior chamber lens. Fourteen months after the injection, visual acuity was 0.30 OS with enlargement of the subfoveal neovascular membrane and an increase of the subretinal exudation. In view of the stabilization of visual acuity after the first injection, a second intravitreal injection of 25 mg of triamcinolone acetonide was performed. For 6 months after the second injection, visual acuity remained at 0.30 OS (Snellen charts). Despite maximally tolerable antiglaucomatous topical treatment, intraocular pressure ranged between 20 mm Hg and 30 mm Hg with the eventual development of glaucomatous changes in the appearance of the optic nerve head. Because intraocular pressure remained uncontrolled and triamcinolone acetonide crystals were still detectable in the vitreous cavity on indirect ophthalmoscopy, although 9 months had passed since the last triamcinolone acetonide injection, a standard trabeculectomy was carried out using mitomycin C intraoperatively in a concentration of 0.25 mg/mL. Postoperatively, the anterior chamber was well formed, there was no leakage at the limbal wound edge, and intraocular pressure ranged between 5 mm Hg and 10 mm Hg in the first postoperative weeks. Concentration of triamcinolone in the aqueous humor sample obtained at the start of the trabeculectomy by anterior chamber fluid aspiration was 11.2 µg/L.

Comment. The clinical course of this patient demonstrates that, in contrast to previous reports on an intraocular presence of triamcinolone for as long as 6 months after the injection, triamcinolone acetonide crystals can be present in the vitreous and soluble triamcinolone can be detected in the aqueous humor for 9 months or longer after intravitreal injection of 25 mg of triamcinolone acetonide. This long intraocular presence of triamcinolone acetonide, especially in patients receiving repeated intravitreal injections of triamcinolone, may not only lead to secondary ocular hypertension but also to steroid-induced secondary open-angle glaucoma necessitating trabeculectomy. This case may also demonstrate that intraocular concentrations of triamcinolone can be achieved for 9 months or longer by a single injection of 25 mg of triamcinolone acetonide. This finding may be interesting for the discussion of when to repeat intravitreal injections and when to use an intraocular slow-release device for corticosteroids.

Jost B. Jonas, MD
Ingrid Kreissig, MD
Robert Degenring, MD
Mannheim-Heidelberg, Germany

The authors have no relevant financial interest in this article.

Bone Located Centrally Within a Dermolipoma

Elsas and Green defined a dermolipoma as “a congenital, chorioretinal tumor containing dermis-like connective tissue and adipose tissue; it was usually covered by non-keratinized, or less commonly by keratinized squamous epithelium.” On pathologic study, bone has been found adjacent to excised dermolipomas in several reports. In an article by Eijpe et al on the characteristic appearance of dermolipomas on computed tomography (CT) scan, the authors did not find calcification in any of their 10 reported cases. We report the clinical, CT scan, and pathologic findings of a patient with bone located centrally within, rather than adjacent to, a pendunculated dermolipoma.

Report of a Case. At birth, a mass was noted at the left lateral canthal area in an otherwise healthy girl. The mass reportedly remained the same size relative to her eyelid and facial structures as she grew. When she was 6 months old, her pediatrician obtained a CT scan of the orbits. The CT scan showed a cystic area with low density similar to fat at the left lateral canthal angle. Centrally within the cystic area was a high-density region that was consistent with bone located centrally within the dermolipoma.

Corresponding author: Jost B. Jonas, MD, Universitäts-Augenklinik, Theodor-Kutzer-Ufer 1-3, 68167 Mannheim, Germany (e-mail: jost.jonas@maaugen.uni-heidelberg.de).