progressive worsening and posterior segment extension. On examination, his visual acuity was light perception OD and 20/40 OS. Examination of the right eye revealed eyelid edema, conjunctival congestion, and a 1-mm hypopyon with fibrinous exudates in the anterior chamber, with no view of the ocular fundus. Results from examination of the left eye were unremarkable. Intraocular pressure was 10 mm Hg OU. He underwent urgent pars plana vitrectomy with injection of intravitreous vancomycin hydrochloride and amikacin sulfate and also received intravenous vancomycin, clindamycin phosphate, and cefotaxime sodium. Vitreous, anterior chamber, and blood cultures grew GBS. A complete blood cell count showed pancytopenia with toxic granulation. Diagnostic inpatient workup including echocardiography revealed no focus of infection. Bone marrow biopsy revealed the diagnosis of precursor T-cell acute lymphoblastic leukemia (Figure). The patient continued to receive parenteral cefotaxime for bacteremia and had systemic chemotherapy initiated by the oncology service, with repeat intravitreous tap and injection of vancomycin 3 days later. Results from the repeat cultures were negative. His visual acuity remained light perception OD 10 days after surgery.

Comment. Endogenous endophthalmitis is a potentially devastating condition characterized by intraocular infection by organisms that access the eye through the bloodstream. Endogenous endophthalmitis accounts for 2% to 6% of all cases of endophthalmitis. In a recent review of cases of endogenous endophthalmitis, GBS was the causative agent in about 5% of cases. Group B Streptococcus most commonly causes infection in neonates and pregnant women, although in the last 2 decades there has been a 2- to 4-fold increase in the incidence of invasive GBS in nonpregnant adults. The most common identifiable sources of GBS are skin or soft-tissue infections, urinary tract infections, pneumonia, bone and joint infections, and endocarditis. In a review of cases of GBS endogenous endophthalmitis, most had ocular involvement within 5 days of onset of sepsis, and ocular infection was not the initial manifestation of sepsis in any patient. The visual prognosis in GBS endophthalmitis is poor, with 76% of cases resulting in visual acuity of light perception or worse.

Our case is unusual because GBS endophthalmitis was the presenting factor leading to the diagnosis of precursor T-cell lymphoblastic leukemia. This case exemplifies the importance of having a high index of suspicion for endogenous endophthalmitis in uveitis cases with a rapidly progressive course that do not respond to standard therapy with corticosteroids. It also shows the important role an ophthalmologist can play in diagnosing serious underlying medical conditions.

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Cystic Solitary Fibrous Tumor of the Orbit

Solitary fibrous tumor (SFT) is a rare spindle cell tumor of mesenchymal origin, most commonly arising from the pleura but also known to occur in extrapleural sites. Orbital SFT was first described in 1994, and since then approximately 70 orbital cases have been published in the literature. Virtually all reported cases of orbital SFT to date were solid tumors. We report the first case to our knowledge of an entirely cystic SFT arising in the orbit.
Report of a Case. A 45-year-old woman had painless right upper eyelid puffiness for 1 year, vision-obstructing ptosis on the right for 6 months, diplopia on left gaze, epiphora, and crusting. On examination, visual acuity was 20/20 OU. There was diplopia at 60° of left gaze. The right eye was dystopic 2 mm laterally and 1 to 2 mm inferiorly with 1 mm of proptosis (Figure 1). An ill-defined softness was palpated superomedially in the right orbit.

Orbital computed tomography revealed a well-defined 2.3 × 1.1 × 1.9-cm cystic mass in the medial aspect of the right orbit with central rim-enhancing fluid hypodensity, central septations, and a 3-mm nodule medially (Figure 1). There was subtle remodeling of the lamina papyracea.

The patient underwent transcaruncular orbitotomy. A 1.8 × 1.1 × 1.5-cm cystic, translucent, blue-gray mass with small amounts of adherent yellow-tan tissue was transected from a thin stalk at its posterior-superior pole. On opening the wall of the mass, prompt deflation with copious straw-colored fluid was noted.

The tumor was pseudoencapsulated, was circumscribed, and had multiple septa (Figure 2). Microscopically, the capsule and septa exhibited features typical of SFT.2,4 There were bland spindle cells haphazardly arranged, without a tendency to form bundles. Some regions were highly cellular, while other regions were hypocellular with abundant matrix and deposition of thick collagen fibers (Figure 2). There was characteristic perivascular fibrosis, particularly in hypocellular areas. Vascularity was rich with dilated vessels and small capillary-sized vessels. Pleomorphism, tumor giant cells, and mitotic activity were absent. The tumor cells formed and lined the septa, indicating that the tumor was really a pseudocyst.

The tumor cells also invaded through the pseudocapsule, reaching the resection margin multifocally. The tumor cells stained positively for immunoreactive vimentin, CD99, CD34, and Bcl-2 (Figure 2). Fibroblasts, including those forming the pseudocapsule, did not stain with CD99, CD34, or Bcl-2. Tumor cells showed no S-100 protein, glial fibrillary acidic protein, epithelial membrane antigen, neurofilament, smooth muscle–specific actin, or desmin immunoreactivity.

Postoperatively, the patient experienced complete resolution of her eyelid swelling, ptosis, diplopia, dystopia, epiphora, and crusting.
Comment. Orbital SFT typically causes insidious, painless proptosis developing over an average of 2 years in patients averaging 40-years-old (age range, 9–76 years). Treatment consists of excision of the circumscribed growth, but local recurrences due to incomplete excision can occur. In our case, microscopic invasion through the pseudocapsule may theoretically permit recurrence. A case of malignant orbital SFT also has been reported. There is a single report of an orbital SFT that was mostly solid with some cystic components. Entirely cystic SFT has been described in other extrapleural locations but never in the orbit. To our knowledge, this case is the first reported case of an entirely cystic SFT arising from the orbit. The fact that tumor cells rather than endothelium lined the septa probably contributed to the pseudocystic fluid accumulation.

The main differential diagnosis in this case was cystic schwannoma and giant cell angiofibroma. The tumor was negative for S-100 protein and neurofilament stains, and there were no giant cells. These findings support a diagnosis of SFT. We recommend that SFT be considered in the differential diagnosis of cystic orbital lesions.

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COMMENTS AND OPINIONS

Ophthalmic Pathology and Ophthalmology

I congratulate Drs Albert and Chévez-Barrios for the August 2009 joint issues of Archives of Ophthalmology and Archives of Pathology and Laboratory Medicine that focused on ophthalmic pathology. Having donated to ophthalmic pathology, including integrating it into clinical ophthalmology practice, I am very supportive of their endeavor to emphasize the importance of this discipline to ophthalmologic teaching, research, and patient care. Ophthalmic pathology distinguishes ophthalmology as a medical specialty because we, as physicians, base our medical and surgical treatments of ophthalmic conditions on understanding the pathobiology of ophthalmic disease. Ophthalmic pathology and ophthalmology are inexorably linked; to diminish or dilute one diminishes or dilutes the other.

Regarding the importance of ophthalmic pathology in the education of ophthalmologists, I agree with the 3 suggestions raised by Clarkson in his editorial: regional ophthalmic pathology centers, enhancing the relationship with departments of pathology, and teaching clinicopathologic correlation through advanced imaging techniques. I would suggest the importance of ophthalmic pathology remaining a vibrant component of ophthalmology departments and ophthalmic practice as well. One example of how this may be accomplished is by linking ophthalmic pathology with a specific clinical, patient-based area of practice.

One such patient-based area of practice is ocular oncology. Ten of the 12 original articles in the Archives of Ophthalmology issue (83%) and 14 of 16 original articles (93%) in the Archives of Pathology and Laboratory Medicine issue were directly related to ocular oncology, many of which were submitted by ocular oncologists. This is an important consideration as ophthalmic pathology moves forward. Therefore, a fourth suggestion for Clarkson’s list is for ophthalmic pathology to become more aligned with specific patient-based ophthalmology areas, such as ocular oncology, in departments of ophthalmology and cancer centers.

This is not to diminish in any way the importance of ophthalmic pathology to other areas of ophthalmology, as there are ophthalmic pathology educational, research, and clinical components in all areas of ophthalmology. Alignment with other clinical disciplines such as cornea, retina, glaucoma, oculoplastics, and others may depend on geographic variations, prevalence of ophthalmic disease, and the particular emphasis of a given ophthalmology department. The important point is alignment with at least one area in an ophthalmology department or region.

For the field of ophthalmic pathology to advance, we as ophthalmic pathologists must not rest on past laurels; we must forge ahead by incorporating modern techniques and technologies into our practice. These entities include, but are not limited to, molecular biology, nanotechnology, and digital technology. We must remain engaged with ophthalmologists and within ophthalmology departments. As in the past, our success will be measured as deliverable accomplishments and contributions. These contributions have been, and only can be, to ophthalmology.

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