spontaneous normal amplitude. Oscillatory potentials were reduced, and flicker responses were of marginal amplitude with a delayed time to peak.

Eight months after initially developing these symptoms, the patient’s mental status had returned to baseline. The visual acuity remained hand motions in the right eye and light perception in the left eye. The fundus appearance was unchanged.

Comment. The chorioretinal lesions seen in our patient are similar to, but much more extensive than, those described in previous articles.1,2 We presume that the severity of both the neurological and the chorioretinal involvement in our patient is associated with immune compromise related to senescence and diabetes mellitus. In contrast to our patient, previously described patients with ocular involvement by West Nile virus have suffered less initial visual loss and have usually enjoyed visual improvement during disease resolution.3,4 The severity of the visual loss in the present case is clearly related in part to the degree of chorioretinal damage seen clinically and angiographically. However, the mild to moderate changes in electoretinography responses do not support visual loss to the light-perception level. In the context of such profound visual loss, the electoretinography findings, reactive pupils, and healthy-appearing optic discs suggest that retrogenticulate damage is likely a contributing morbidity. Because routine magnetic resonance imaging has failed to show structural damage in this area, functional magnetic resonance imaging studies are planned.

John P. Myers, MD
Thellea K. Leveque, MD, MPH
Mark W. Johnson, MD

Correspondence: Dr Johnson, Department of Ophthalmology and Visual Sciences, W. K. Kellogg Eye Center, 1000 Wall St, Ann Arbor, MI 48105-0714 (markwj@med.umich.edu).

Financial Disclosure: None.


Retinal Folds as Initial Manifestation of Orbital Lymphangioma

Orbital lymphangioma is a rare, benign tumor that typically develops in childhood as proptosis or an orbital mass. These highly vascularized lesions, which also contain various amounts of lymphoid tissue, may demonstrate an acute increase in size concomitant with a viral illness or spontaneous hemorrhage. In this report, we describe an unusual manifestation of orbital lymphangioma in which an acute decrease in vision was the sole complaint and retinal folds the initial objective clinical finding.

Report of a Case. A 15-year-old girl was referred for the evaluation of transient decreased vision in both eyes and persistent decreased vision in the right eye 1 week after being diagnosed as having mononucleosis. The patient was evaluated by an outside ophthalmologist who noted an acuity of 20/50 OD and 20/25 OS. A funduscopic examination of the right eye revealed a star-shaped wrinkling of the macula. She was referred to our clinic for the evaluation of an epiretinal membrane with cystoid macular edema.

On initial examination at our clinic, her uncorrected acuity was 20/80 OD and 20/20 OS. The refraction was +4.50 sph minus 0.75 at 135 yielding 20/20 OS. External and anterior segment examinations were normal in both eyes (Figure 1). Extra ocular movements were full. Contact lens biomicroscopy of the right eye revealed retinal folds radiating outward from the center of the macula (Figure 2). The left eye was normal. To evaluate for retinal edema, epiretinal membrane, or central serous retinopathy, fluorescein angiogram and optical coherence tomography examinations were performed. The fluorescein angiogram transiting the right eye revealed retinal folds and rare chorioretinal folds in the macula with no evidence of leakage (Figure 3). Optical coherence tomography revealed chorioretinal folds in the right eye (Figure 4). No epiretinal membrane, macular edema, or serous retinopathy was present. To evaluate for posterior orbital masses, B-scan ultrasonography of the right eye was obtained. It demonstrated a flattened globe with a cystic, loculated mass in the retro-orbital space (Figure 5). An orbital computed tomographic scan with contrast demonstrated a heterogeneously enhanc-
ing intraconal and extraconal mass on the right side of the eye that distorted the posterior aspect of the globe. Prone and supine images demonstrated no difference in the size of the mass, and no calcifications were seen (Figure 6).

The cystic nature of this patient’s mass was felt to be most consistent with a lymphangioma or an orbital varix. However, because a malignant tumor with a cystic component, such as rhabdomyosarcoma, could not be excluded, an incisional biopsy was performed. During the biopsy, serosanguinous fluid drained from the lesion. The pathologic specimen demonstrated dense fibrous tissue mixed with adipose tissue, blood vessels, and a thick-walled vascular channel with a lumen that was devoid of identifiable cells or other material. The histopathologic results were consistent with lymphangioma, and observation was recommended (Figure 7). At the 6-month follow-up visit, the patient’s examination findings were unchanged.

Comment. Lymphangioma is a rare, benign tumor of the lymph-vascular system that typically develops early in life. Orbital lymphangioma, which accounts for approximately 25% of cases, usually progresses slowly during childhood and stabilizes by late adolescence. In a review of 158 cases of orbital lymphangioma and orbital varices, Wright et al found the most common initial findings to be pain (43%), mass (42%), hemorrhage (37%), and proptosis (15%). Females are affected 2 to 3 times as often as males. It is well documented that lymphangiomas, which contain various amounts of lymphoid tissue, may increase in size during a viral illness or secondary to an intrinsic hemorrhage.

Although dramatic visual loss has been described, most patients with orbital lymphangioma do not exhibit a primary complaint of decreased vision. It has been presumed that lymphangiomas are often too soft to cause significant visual impairment, although compressive optic neuropathy and visual loss can occur with acute hemorrhagic expansion of these lesions. This softness may also be a reason for the infrequent description of retinal folds as an initial characteristic in these cases.

The management approach to orbital lymphangioma is controversial. The diffuse weblike pattern, with interdigitation of adjacent orbital structures, often makes complete resection difficult. In the past, multiple subtotal excisions have been recommended. However, surgical excision can be complicated by new hemorrhagic cysts that can increase the risk of vision loss and motility impairment. Therapies such as carbon dioxide laser, sclerosing therapy, and radiotherapy have mixed outcomes. Observation is often recommended, since the hemorrhagic cyst and accompanying proptosis will often spontaneously resolve. It has been suggested that surgery to decompress these lesions be reserved for cases of optic nerve compromise, poor cosmesis, pain, disabling diplopia, or the threat of amblyopia.

Our case presents an unusual manifestation of orbital lymphangioma in which deterioration of vision was the sole complaint and retinal folds the initial clinical finding. We believe that mononucleosis was the precipitating event for enlarge-
ment of the orbital lymphangioma in this patient.

Sherman W. Reeves, MD, MPH
Dianna L. Miele, MD, MBA
Julie A. Woodward, MD
Thomas J. Cummings, MD
Megan C. Eales, COT
Paul Kurz, MD
Michael J. Cooney, MD

Correspondence: Dr Reeves, Box 3802, Duke University Eye Center,
Durham, NC 27710 (sherman.reeves@duke.edu).

Financial Disclosure: None.


Ewetzky's (294) case was that of a boy of 3 and a half years who had signs of inflammation in 1 eye, lasting for a week, and relapsing for a short time, after a month's free interval.

After the operative removal of the worm, which was found to be the larva of a fly of undetermined species, recovery rapidly took place.