diffuse edema within the subcutaneous tissues of the thorax and abdomen. A biopsy specimen of his skin lesions showed nonspecific inflammatory cell infiltration of the dermis. A bone marrow biopsy specimen revealed large, atypical lymphocytes filling the lumen of the capillaries in the alveolar septae (Figure 2B and C). These findings were diagnostic of malignant intravascular lymphoma of B-cell phenotype (intravascular lymphomatosis).

A regimen of hyper-CVAD (cycle 1: cyclophosphamide, vincristine, doxorubicin, and dexamethasone) chemotherapy, intrathecal methotrexate and cytarabine, and methylprednisolone was initiated 10 days after initial examination. A week later, there was a marked improvement in visual acuity (20/20 OU) and resolution of serous detachment and choroidal thickening, shown by repeat fluorescein angiography and indocyanine green angiography (Figure 1E and F). The patient continued to do well 2 months after starting therapy.

Comment. Intravascular large B-cell lymphoma is a rare form of extranodal malignant lymphoma in which the lymphoid cells are principally found within small vessels without extensive involvement of bone marrow or lymphoid tissue. Aberrant expression or deletion of certain cell adhesion molecules critical for lymphocyte trafficking and transvascular migration (CD29 and CD54) contributes to the tumor's intravascular and disseminated pattern of distribution. The tumor expresses CD20 and other B-cell markers and does not express the T-cell marker CD3. At diagnosis, the lymphoma typically involves various organs, including the central nervous system, skin, lung, kidneys, spleen, and adrenal glands. Other types of lymphoma may separately coexist with intravascular large B-cell lymphoma in the same patient. Hematologic abnormalities, like microangiopathic and autoimmune hemolytic anemia, as in our case, are often associated with this disease process. The prognosis is generally very poor because of the usually delayed diagnosis. Intravascular large B-cell lymphoma can also involve the small vessels of the choroid. The case presented herein adds the eye as another end organ involved by this rare malignancy.

B. Harold Lee, MD
Jose S. Pulido, MD, MS
Helmut Buettner, MD
Diva Salomão, MD
Clive S. Zent, MD
Thomas P. Link, BA

Correspondence: Dr Pulido, Mayo Clinic, Department of Ophthalmology, 200 First St SW, Rochester, MN 55901-0001 (pulido.jose@mayo.edu).

Author Contributions: Dr Pulido, principal investigator, had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Financial Disclosure: None reported.

Funding/Support: This project was funded by an unrestricted grant from Research to Prevent Blindness, New York, NY.


Unilateral Eyelid Swelling and Ptosis Caused by Dural Arteriovenous Fistula in an Infant

Patients with potentially life-threatening intracranial vascular abnormalities may first come to an ophthalmologist with signs or symptoms that include visual disturbances, bruits, papilledema, or periorbital congestion commonly coexisting with other neurologic and cardiovascular problems. In this report, we discuss a girl seen first at 6 months of age with the rare finding of isolated, unilateral eyelid ptosis and fullness due to a dural arteriovenous fistula.

Report of a Case. A female child was noted at birth to have right upper eyelid swelling and ptosis. Although they were believed to be related to birth trauma, there was little change in her eyelid appearance over the next few months, prompting referral to a pediatric ophthalmologist (B.J.K.) when the girl was 6 months old. Fullness of the right upper eyelid was evident (Figure 1), but no palpable mass or orbital bruit was appreciated. The remainder of her ophthalmic examination, including eyelid function, motility, and sweep visual evoked potential testing, had normal findings for both eyes. The child had otherwise enjoyed good health and achieved appropriate developmental milestones.

Magnetic resonance imaging revealed a dural arteriovenous fistula with massive engorgement and dilatation of the dural venous sinuses, including the distal superior sagittal sinus, torcular herophili, and right transverse sinus (Figure 2A). Cerebral angiography identified multiple arterial feeding vessels, the most prominent of which were derived from the occipital arteries bilaterally (Figure 2B). Also noted were bilateral dilated superior ophthalmic veins due to shunting of cerebral venous blood to the external periorbital veins. There was no evidence of intracranial mass effect, infarct, hemorrhage, or other parenchymal abnormalities.

With the poor natural history of dural arteriovenous fistula in children, we decided to attempt endovascular embolization of the lesion to prevent long-standing cerebral venous hypertension and potentially irreversible brain injury. A preoperative cardiovascular evaluation, including echocardiogram, revealed only a slightly enlarged left atrium due to mildly increased cardiac output.

At 7 months of age, the patient underwent initial endovascular treatment using a combined transve-
nous and transarterial approach with selective embolization of feeding branches from the right occipital and middle meningeal arteries, branches of the vertebral arteries, the superior sagittal sinus, and the torcular herophili (Figure 2C). Postoperatively, the child did well, and her parents noted an increase in alertness and playful activity and increased interaction with her siblings. Her right upper eyelid ptosis and fullness were unchanged, however, and an occipital bruit (first noted during a follow-up visit) remained. When the patient was 19 months of age, we performed a second embolization procedure because of persistence of part of the malformation and complaints of intermittent headaches. At age 4 years, the child’s neurodevelopment continued to be normal, although her visual acuity as measured by the HOTV method was 20/70 OD and 20/60 OS despite an otherwise normal ocular examination. The cause of her decreased visual acuity was not clear, although a component of cortical damage from chronic cerebral venous hypertension was suspected. She continues to be monitored closely by her multidisciplinary medical team.

Comment. Dural arteriovenous fistula in the pediatric population is rare, often multifocal, and generally more complex than adult dural arteriovenous fistula.1,2 In adults, an insult from trauma, intracranial surgery, thrombophlebitis, or dural venous thrombosis is believed to initiate an inflammatory cascade with ensuing neovascularization and angiogenesis, resulting in shunts at the arteriolar level.3 The exact causative triggers in children are unknown, although both embryologic and intrauterine-acquired etiologies have been proposed.1,2 Signs and symptoms in infants typically include cardiac failure, hydrocephalus, hemorrhage, and seizures, although some children may only have bruits, proptosis, or papilledema.1,2 The increased risk of heart failure necessitates a comprehensive pediatric cardiology evaluation. In our patient, normal internal jugular venous drainage was compromised by shunted arterial flow, leading to asymmetric anomalous drainage of venous blood through the cavernous and periorbital venous system, resulting in unilateral upper eyelid swelling and mechanical ptosis.

The major goal of therapy is to disconnect the fistula from its arterial supply and ensure that alternative venous outlets remain patent and functional.2 Infants with multifocal lesions tend to have more serious neurological sequelae than adults, including neurocognitive delay, hemorrhage, or death.
although the advent of newer endovascular treatments has improved overall outcomes.\textsuperscript{1,3} In complex lesions with numerous arterial feeding arteries, multiple therapeutic embolizations may be required to definitively cure these life-threatening fistulae. Our case illustrates that children with potentially life-threatening intracranial vascular abnormalities may first visit the ophthalmologist with seemingly benign symptoms, making prompt diagnosis and treatment essential.

Gregory J. Griepentrog, MD
Beverly Aagaard-Kienitz, MD
Burton J. Kushner, MD
Bermans J. Iskandar, MD
David M. Gamm, MD, PhD

Correspondence: Dr Gamm, T607 Waisman Center, 1500 Highland Ave, Madison, WI 53705 (dgamm@wisc.edu).
Financial Disclosure: None reported.


Vicarious Menstruation in Primary Localized Conjunctival Amyloidosis

Recurrent subconjunctival hemorrhage has been found to occur in various circumstances, including in primary conjunctival amyloidosis.\textsuperscript{1} It usually occurs spontaneously and warrants ocular and systemic examination. However, if it occurs precisely every month in association with menstruation, then it may be termed vicarious menstruation.

Report of a Case. A 30-year-old woman had an 8-year history of unilateral recurrent subconjunctival hemorrhaging that commenced each month on the first day of her menstrual cycle and cleared after 7 to 10 days. The monthly timing of the bleeding was so precise that she found it a reliable indicator for day 1 of each menstrual cycle. The initial onset of symptoms appeared to have occurred shortly after the birth of her first child. Examination revealed a fleshy, thickened, discrete conjunctival mass in the inferior fornix (Figure 1). This appearance raised the suspicion of conjunctival lymphoma or amyloid; hence, a biopsy and debulking of the lesion were performed.

Histological studies of the biopsy specimen revealed findings consistent with conjunctival amyloidosis (Figure 2). Light microscopy did not reveal any ectopic endometrial tissue. There was no significant inflammatory response, nor was there any vascular or lymphatic channel abnormality.

Systemic examination and laboratory investigations (including serum amyloid measurement) for evidence of systemic amyloid disease had results that were unremarkable, suggesting that the amyloid deposition was most likely primary and localized in nature. Use of an oral contraceptive pill to regulate hormone levels had no effect on the timing, frequency, or duration of the subconjunctival hemorrhage. Debulking of the lesion resulted in cessation of the hemorrhaging. However, identical cyclical symptoms recurred 2 years later when the mass regained its size, requiring further debulking.

Comment. The close association between the size and existence of the amyloid mass with the incidence of the subconjunctival hemorrhages, in addition to the laterality of the signs, suggests that amyloid deposition was responsible for the bleeding. The recurrent hemorrhages may well have been due to amyloid in the blood vessel walls in a similar fashion to purpura found in primary systemic amyloidosis. However, the timing suggests that the microangiopathy was responsive to hormonal fluctuations of the menstrual cycle. Although serum amyloid P component levels are known to fluctuate with hormonal changes,\textsuperscript{2} this was not found to be the case in our patient. On the other hand, the hemorrhages could have been due to physiological vascular changes that occur in menstruation confounded by the localized amyloid; hence, debulking of the lesion was merely reducing the vascularity of the conjunctival mass. However, histological evidence showed that the lesion was not a highly vascular structure, implying that the vessels were merely more prone to bleeding.

Subconjunctival hemorrhages have been found to occur in pri-