vascular loop. Fixation was unstable and 5° inferotemporal to the fovea (Figure 2). Comparison with the fundus photograph taken 1 year earlier showed some advancement of the loop.

The left eye was healthy. There was no strabismus, and stereopsis (Lang test) was normal. Cycloplegic refraction was +2.00 diopters spherical OU. The medical history, systemic history, and examination results were unremarkable. Recent magnetic resonance tomography results did not show any intracranial anomaly of Bonnet-Dechamp-Blanc or Wyburn-Mason syndrome.

Because the location of the loop itself did not account for the total amount of visual acuity loss, we suspected relative amblyopia and recommended occlusion of the left eye for 4 hours per day.

Eight months later, visual acuity had improved to 20/30 OD and scanning laser ophthalmoscope–fundus perimetry showed a definite central shift of the retinal fixation area (Figure 2). Esotropia of +10° without correction and +3° with correction was now present.

Afterward, occlusion therapy was difficult and finally abandoned by the parents. One year later, visual acuity was 20/400 OD. A further progression of the vascular loop and a 13° temporal shift of the fixation area were present (Figure 2). Because of that and the lack of success to maintain useful vision, we stopped any treatment.

Comment. The initial drop in visual acuity may have been due to 2 factors, a slow progression of the loop approaching the fovea and relative amblyopia. The progression of the loop toward the fovea was gradual and totaled 500 μm in 33 months (180 μm/yr). Although most cases of arteriovenous malformation are thought to be stable,2 anatomical changes of arteriovenous malformations have previously been described4 but not linked to visual loss.

On the other hand, relative amblyopia is likely to have been a contributing factor. It is defined as a reversible functional deficit superimposed on a congenital organic abnormality or pathologic changes acquired in early childhood. Our hypothesis was supported by the increase in visual acuity with patching and the shift of fixation. When manifest esotropia was present, strabismic amblyopia may have contributed to the final visual loss.

Not all retinal AV malformations are stationary, and small changes can induce a visual loss that may be amplified by relative amblyopia in patients during their sensitive period of visual development. Unlike this patient, most cases do not progress. A trial of occlusion therapy may be warranted in young children if visual acuity is worse than expected from the location of a central loop.

Olive Ehrt, MD Munich, Germany

The author has no relevant financial interest in this article.

Corresponding author: Oliver Ehrt, MD, Department of Ophthalmology, Ludwig Maximilians University, Mathildestr 8, D-80336 Munich, Germany (e-mail: oliver.ehrt @med.uni-muenchen.de).

Report of a Case. A 20-year-old woman had complex retinal detachment and proliferative vitreoretinopathy in her right eye. She noted a deterioration in the vision of her right eye 1 day prior to hospital admission. Her ocular history was remarkable for a progressive impairment in the peripheral visual field during the past 18 months, but she had had no dilated eye examinations in the meantime. The function of the right eye had always been worse compared with the left eye, and it was thought to be amblyopic. On examination, visual acuity was hand movements OD and 20/20 OS. Slitlamp examination was unremarkable except for anterior vitreous opacities in the right eye. Funduscopy disclosed a subtotal retinal detachment with macular involvement in the right eye. There were subretinal fibrous strands in the inferior 2 quadrants and epiretinal membrane formation with star folds peripheral to the major vascular arcades consistent with a long-standing detachment and advanced proliferative vitreoretinopathy. While the optic disc in the left eye appeared healthy, there were 2 pits noted in the disc of the right eye. A small, rather subtle depression was located at the 8-o’clock position. A larger, well-developed, obvious declivity, the bottom of which was not visible, was at the nasal disc margin (Figure 1). No diaphanous tissue was identified overlying any of the pits.

Subsequently, pars plana vitrectomy, removal of subretinal and
Two days after surgery for retinal detachment, which included an inferior retinotomy, peripheral retinectomy, endolaser photocoagulation, and silicone oil tamponade, subretinal perfluorodecalin droplets appeared in the subretinal space at the inferior margin of the optic disc. The whitish spots represent diode laser spots.

Epiretinal membranes, inferior retinotomy, peripheral retinectomy, endolaser photocoagulation, and silicone oil tamponade were performed. Perfluorodecalin was used intraoperatively, and all visible PFD was removed prior to air-fluid exchange and silicone oil filling. Postoperatively, the retina was completely attached and visual acuity improved to 20/60 OD. Two days postoperatively, several small droplets of PFD were noted in the subretinal space inferior to the optic disc margin. The number of droplets increased during the next 2 days, and some were advancing toward the macula (Figure 2). Therefore, another surgical procedure was performed 7 days after the initial surgery to remove all visible subretinal PFD. This was accomplished in the presence of the silicone oil tamponade by gently advancing the subretinal PFD droplets toward the inferior retina spatula (DORC Internation, Zuidland, the Netherlands). Once released from the subretinal space, the droplets were removed via a fluid needle and concurrent pressure increase generated by a conventional oil pump (Gueuder, Heidelberg, Germany). At the end of this procedure, no more PFD droplets were visible in the subneurosensory space. However, 1 day later, new subretinal PFD droplets appeared again at the inferior disc margin. The amount of droplets increased during the subsequent days, however, at a slower rate than before. Thirteen days later, in addition to the PFD droplets, silicone oil appeared in the same subretinal area with progressive enlargement of the retinal detachment toward the fovea.

Silicone oil was then removed, and the subretinal PFD and silicone oil was released via the inferior retinotomy (Figure 3). The vitreous cavity was now filled with air and 16% C3F8 gas. A subsequent retadhachment associated with proliferative vitreoretinopathy, including epiretinal membrane formation and traction, necessitated another surgical procedure. This time, no PFD was used intraoperatively and prior to silicone oil tamponade, diode laser endophotocoagulation was applied around the inferior of the optic disc to generate chorioretinal scars that would prevent silicone oil from passing into the subretinal space. Three months later, the retina remained attached, no new subretinal PFD or silicone oil occurred, and best-corrected visual acuity was 20/100 OD.

Comment. Congenital pits of the optic nerve head represent a rare abnormality, occurring in approximately 1 in 11,000 patients. Their pathogenesis is not completely understood but is thought to be related to anomalies of closure of the fetal fissure. Pits of the disc are usually unilateral but may occur in both eyes. Most often, they are present on the temporal edge of the disc, but they may be multiple and have been observed in other meridians. Clinically, the pits may vary considerably in size, ranging from subtle, barely visible depressions to well-developed, obvious declivities.

Serous macular detachment develops in 25% to 75% of eyes with optic nerve pits. The pathophysiologic explanation of this complication is controversial. Several theories have been brought forward including leakage from the vessels at the base of the pit, leakage from the parapapillary choroidal circulation, leakage from the cerebrospinal fluid, and leakage from the vitreous cavity.

The case reported here clearly demonstrates that there may be a communication between the vitreous cavity and the subretinal space via the optic pit and that this may be the cause for retinal detachment. The most likely explanation for the appearance of subretinal PFD and silicone oil bubbles several days postoperatively is a reservoir posterior to the optic disc plane not visible by biomicroscopy and a communication between the optic disc pit and the subretinal space. Various possibilities may be considered for such a reservoir including the subarachnoid space around the optic nerve or posterior herniation of abnormal tissue composing the true “floor” of the pit. While instant leakage of PFD under the retina was noted intraoperatively in the presence of a morning glory disc anomaly by Bartz-Schmidt and Heimann, this is, to the best of our knowledge, the first description of a delayed occurrence of PFD and silicone oil in the presence of optic pits.

It has been demonstrated in an animal study that india ink placed in the vitreous cavity flowed into the pit and subretinal space, supporting the hypothesis that the subretinal fluid originates from the vitreous cav-
ity. Bonnet et al reported eyes with optic pits and associated retinal detachment managed with intravitreal gas injection, in which gas bubbles migrated through a small break in tissue overlying the optic pits into the subretinal space postoperatively. Coll et al reported a communication between the subretinal space and the vitreous cavity in the morning glory syndrome.

Our case suggests that PFD should be avoided in retinal detachment surgery in eyes with optic pits. Furthermore, intraocular air/gas tamponade may be preferable to silicone oil tamponade if feasible to prevent the complications previously mentioned. The lower interfacial tension of silicone oil was not adequate to prevent bubbles of silicone oil from passing through the small retinal opening within the optic disc. Endolaser photocoagulation for formation of a barrier to fluid movement was effective here to prevent silicone oil from passing into the subretinal space. Such laser treatment appears prudent in similar cases when air/gas tamponade is insufficient to secure retinal attachment and when silicone oil must be used. The volume of both subretinal PFD and silicone oil increased during a short period of time. It is currently unknown which unidirectional forces are operative to efficiently move fluid from the vitreous cavity through the optic pit under the neurosensory retina. A pressure gradient must be assumed whereby gravity forces are unlikely because PFD and silicone oil owing to their specific weights would be expected to behave differently. It may be speculated that the pressure gradient is the difference between the intraocular pressure at the moment of migration and the pressure within the reservoir (ie, either the extraocular tissue pressure or the pressure within the subarachnoid space).

Stefan Dithmar, MD
Florian Schuett, MD
Hans E. Voelcker, MD
Frank G. Holz, MD
Heidelberg, Germany

Corresponding author: Stefan Dithmar, MD, Department of Ophthalmology, University of Heidelberg, Im Neuenheimer Feld 400, 69120 Heidelberg, Germany (e-mail: stefan_dithmar@med.uni-heidelberg.de).


Idiopathic Juxtafoveal Telangiectasis in Association With Celiac Sprue

Idiopathic juxtafoveal telangiectasis (IJFT) is a condition of uncertain etiology that is characterized by retinal telangiectasias, superficial retinal crystalline deposits, right-angle venules, and intraretinal pigment plaques. It is capable of causing visual loss in otherwise healthy patients, and treatment remains controversial. It was first described by Reese in 1956 and has subsequently been divided into 3 groups by Gass and Blodi.

We describe a patient with IJFT and celiac sprue. To the best of our knowledge, this is the first reported case showing a potential association between these 2 conditions.

Report of a Case. A 53-year-old woman with biopsy-proved celiac sprue reported a 10-year history of blurred central vision in both eyes. Her medical history was otherwise unremarkable. She denied a history of diabetes and had a normal blood glucose level, but she declined a formal glucose tolerance test. Her medical regimen included a multivitamin, hormone replacement therapy (estrogen-medroxyprogesterone), and a gluten-free diet. Her family history included no relatives with similar visual loss.

Best-corrected visual acuity was 20/60 OD and 20/40 OS. Clinical examination showed clear media and symmetric optic discs. Macular edema and a perifoveal gray macular sheen were noted with apparent foveal thinning; no lipid or hemorrhage was present. A right-angle venule was noted in the temporal right macula (Figure 1).

Figure 1

Fluorescein angiography showed intraretinal telangiectasia with leakage for 340° in the right eye and nearly 360° surrounding the fovea in the left eye (Figure 2). Optical coherence tomography showed foveal thinning in both eyes with mild macular thickening (Figure 3).

Comment. Celiac sprue is characterized by malabsorption due to injury to the intestinal mucosa after ingestion of wheat gluten or related proteins. There is both clinical and histologic improvement on a strict gluten-free diet, and relapse occurs when dietary gluten is reintroduced. Until recently, celiac sprue was thought to be an uncommon condition in the United States, with an estimated prevalence of 1 in 3000. However, greater awareness of its manifestations and the advent of more accurate serologic tests have led to the realization that celiac sprue is relatively common, affecting approximately 1 of every 120 to 300 persons in North America.

Celiac sprue is caused by an inappropriate T cell–mediated immune response against ingested gluten, causing damage to the intestinal mucosa, and the classic histologic finding of absent villi and hyperplastic crypts. There is a strong genetic component, with more than 95% of patients expressing the HLA-DQ2 heterodimer.

Classically, celiac sprue is seen in infants with impaired growth and abdominal distention. The