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Optical Coherence Tomography Findings in Foveal Schisis

Juvenile retinoschisis is a congenital X-linked recessive retinal disorder. Patients may develop nystagmus, decreased central vision, or strabismus. Fundus findings vary considerably, from the absence of a foveal reflex to the presence of a large, elevated schisis cavity involving the fovea. The protean finding is a foveal schisis. Logically, the peripheral retinoschisis is found in the nerve fiber layer. However, on review of recent literature, it is not clear where the foveal split occurs. The progression of juvenile retinoschisis is associated with changes in the fovea and underlying retinal pigment epithelium. Additional information may be gleaned from fluorescein angiography, electroretinography, or genetic studies.

Optical coherence tomography (OCT) is a recent advance in retinal imaging; the techniques of this modality are described elsewhere. Few case reports describe OCT imaging of this disease. We demonstrate the use of OCT to highlight unique foveal findings in a patient with juvenile retinoschisis.

**Report of a Case.** A 26-year-old man came to our office for evaluation of decreased vision and foveal schisis. This patient was previously examined and diagnosed when he was 10 years old. The best-corrected visual acuity was 20/60 OD and 20/50 OS. Refraction was +0.50 sphere OU. Pu-

![Figure 1](http://example.com/figure1.png)

**Figure 1.** Red-free photographs demonstrate foveomacular schisis in both eyes.

![Figure 2](http://example.com/figure2.png)

**Figure 2.** Two-dimensional optical coherence tomographic (OCT) scan of the right eye, with horizontal (A) and vertical (B) sections through the macula. Note the broad central area of flat foveal tissue surrounded by multiple levels of schisis. Horizontal (C) and vertical (D) sections reflect similar pathologic features in the left eye.
pillarary reflexes were normal. The optic discs were both normal. Foveal schisis and internal limiting membrane changes were observed in each eye (Figure 1). No vitreous detachment was present. Peripherally, there were a localized choroidal retinal scar in the right eye and a relatively flat retinoschisis cavity in the left.

Electroretinography demonstrated decreased b-wave amplitude, which was consistent with the diagnosis. After adequate dilation, OCT (Optical Coherence Tomography version 3000; Zeiss Humphrey Instruments, Dublin, Calif) was performed. Six-millimeter radial sections of each macula were completed. The OCT figures demonstrated schisis of at least 2 retinal layers adjacent to the fovea. The center of the fovea, however, was not elevated (Figure 2).

Comment. X-linked juvenile retinoschisis is a retinal dystrophy that may have a variety of clinical findings. Fundus findings often mimic cystoid macular edema; however, there is no leakage on fluorescein angiography. The pathologic feature involves a split in the nerve fiber layer and may be related to Muller cell dysfunction. Funduscopy demonstrates foveal schisis in virtually all patients and peripheral retinoschisis in half of the cases. Clinically, pigmentary changes can develop in the fovea with loss of foveal schisis across time. Moreover, the plications described with X-linked juvenile retinoschisis reflect a true split in the retina, as supported by the OCT findings. The OCT indicates that the split involves multiple retinal layers in the same cross section, including the nerve fiber layer and/or deeper layers. The broad area of flat central tissue on OCT suggests that foveal schisis may collapse with subsequent alterations of the retinal pigment epithelium and overlying retina. Menchini et al have shown similar OCT results in myopic females with unilateral macular retinoschisis.

In conclusion, OCT may offer new insight into the pathologic features of this condition. In this case, it was used to reveal unique foveal pathologic features of a patient with a clinical diagnosis of X-linked juvenile retinoschisis.

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Bilateral Serous Retinal Detachment Due to Protein-Losing Enteropathy

Protein-losing enteropathy (PLE) refers to a group of disorders causing hypoproteinemernia and edema in the absence of proteinuria or defects in protein synthesis. It is characterized by increased protein loss in the gastrointestinal tract and is commonly suggested by the presence of peripheral edema and low serum albumin and globulin levels, in the absence of renal and hepatic disease. We describe an unusual patient with a corticosteroid-responsive PLE who developed bilateral serous retinal detachments (RDs) coincident with a flare of her enteropathy. With appropriate treatment of the enteropathy, there was resolution of the serous detachments.

Report of a Case. A 47-year-old woman was diagnosed as having idiopathic PLE 8 years before referral to the Retina Service of the Department of Ophthalmology and Visual Sciences, University of Wisconsin–Madison.

In 1995, she was seen at the Mayo Clinic, Rochester, Minn, complaining of swelling over the face and lower extremities. The findings from the physical examination revealed anasarca. She reported a history of multiple episodes of edematous facies and lower extremity edema responsive to intermittent short courses of oral corticosteroid therapy.

Laboratory analysis revealed severe hypoalbuminemia, with serum albumin levels measuring 1.4 g/dL (normal range, 3.5-5.5 g/dL). Renal and liver function test results were unremarkable. The antimicrobial antibody was mildly elevated, although a complete rheumatologic evaluation ruled out rheumatologic disease, including systemic lupus erythematosus.

Protein-losing enteropathy was suspected. Twenty-four-hour stool analysis revealed a high level of fecal ß2–antitrypsin concentration at 642 mg/dL (118.13 μmol/L) (normal, <54 mg/dL [<9.94 μmol/L]), and 24-hour serum analysis for ß2–antitrypsin clearance was 964 mL/24 h (normal, <27 mL/24 h). (Intestinal clearance of ß2–antitrypsin has been shown to be a reliable diagnostic test for PLE.) Esophagogastroduodenoscopy and gastric biopsy revealed a congestive gastropathy. A diagnosis of PLE was made.

Given the patient’s history of responsiveness to corticosteroid administration, she was successfully treated with intravenous corticosteroids (methylprednisolone), intravenous furosemide, and albumin, with resolution of the anasarca. The PLE was subsequently well controlled with 10 mg of methylprednisolone (Medrol) every other day and 50 mg of mercaptopurine (Purinethol) daily for the subsequent 8 years.

The patient developed a systemic viral illness 4 weeks before being seen at the Department of Ophthalmology and Visual Sciences, University of Wisconsin–Madison clinic. As her upper respiratory symptoms began to resolve, she sought care from a primary care physician because of complaints of increased abdominal girth, profound anorexia, diarrhea, and shortness of breath.

The medical workup revealed abnormal liver function test results, including a low serum albumin...