Article • Case Report: Lipemia Retinalis in a 15-Year-Old

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ABSTRACT

Background: High cholesterol impacts blood vessels in every part of the body. Lipemia retinalis is a rare retinal presentation of severe hypertriglyceridemia (usually greater than 2000 mg/dl), resulting in a creamy-white discoloration of retinal blood vessels. The following case presents a pediatric case of lipemia retinalis.

Case Report: A 15-year-old Hispanic female presented for an annual diabetic eye exam. She reported that her vision had been blurry OU since she lost her glasses. Her medical history included type 2 DM, diagnosed 1 year prior. Her last blood glucose was 208 mg/dL, and she did not know her HbA1c. Her medications included metformin and insulin. DFE revealed creamy-white colored arteries and veins in both eyes and a salmon-colored fundus, but no evidence of diabetic retinopathy. OCT showed increased signal in the blood vessel lumens. Her lab work showed total cholesterol of 1328 mg/dL and triglycerides of 7337 mg/dL. The PCP started the patient on a low-fat diet, antitryglyceride, and anticholesteral medication.

Conclusion: Visual sequelae associated with lipemia retinalis is rare, as most patients are visually asymptomatic. There is no treatment for the lipemia retinalis itself, other than managing the underlying systemic metabolic pathologies. Coordination with the primary care physician is required.

Keywords: cholesterol, lipemia retinalis, pediatrics, retina

Introduction

Lipemia retinalis is a very rare retinal presentation of severe hypertriglyceridemia (usually greater than 2000 mg/dl) resulting in a creamy-white discoloration of retinal blood vessels (normal triglyceride levels are 50-149 mg/dL). It was first described in 1880 by Heyl. It can affect people of any age; pediatric and adult cases have been reported. Severe hypertriglyceridemia is defined as triglyceride levels greater than 500 mg/dl, occurring in approximately <0.2% cases of children and adolescents in the United States.

Lipemia retinalis has been described as having three distinct stages. Early stage is when peripheral vessels are affected but central blood vessels are normal in coloration. Moderate stage occurs when the creamy-colored blood vessels extend towards the optic disc. The severe/marked stage is when all retinal blood vessels are involved. A case of severe/marked lipemia retinalis follows.
Dilated fundus exam revealed creamy-white colored arteries and veins in both eyes, and a salmon-colored fundus, but no evidence of diabetic retinopathy (Figures 1 & 2). Optical coherence tomography showed increased signal in the blood vessel lumens (Figure 3). Based upon her retinal blood vessel appearances, she was diagnosed with lipemia retinalis. The patient denied any evidence of dermatological eruptive xanthomas occasionally associated with lipemia retinalis. She was referred urgently to her pediatrician for same-day complete blood count, comprehensive metabolic panel, and a lipid panel. Her lab work showed random blood glucose of 398 mg/dL (reference range: 100-139 mg/dL), total cholesterol of 1328 mg/dL (reference range: <200 mg/dL), and total triglycerides of 7337 mg/dL (reference range: <150 mg/dL) (Table 1). The PCP recommended that the patient start on a low-fat diet, antitryglyceride, and anticholesterol medication, but the patient never returned for follow-up with the PCP, nor for repeat eye examination.

**Discussion**

The pathological appearance of lipemia retinalis stems from the visualization of triglyceride-filled chylomicrons circulating in the retinal vessels that result in light scattering and production of a turbid, salmon-pink to creamy-white color based upon the plasma triglyceride level. Notably, hypercholesterolemia alone does not produce this
Pancreatitis and familial hyperlipoproteinemias (types 1, 4, 5) appear to be the most common associations with lipemia retinalis.\(^5,6,9,16\) To the authors’ knowledge, the largest triglyceride level associated with lipemia retinalis was reported to be 24,500 mg/dL in a 12-week-old infant.\(^5\)

Visual sequelae associated with lipemia retinalis is rare, as most patients are visually asymptomatic upon presentation, but retinal occlusive disease has been reported previously.\(^2,4,9,12,15\) There is no specific treatment for the lipemia retinalis itself, other than managing the underlying systemic metabolic pathologies.\(^5,6,9,15\) Currently existing technologies can be used efficiently to diagnose, document, and follow patients with lipemia retinalis. These technologies include: fundus photography, widefield scanning laser ophthalmoscopy, electroretinography, optical coherence tomography, and optical coherence tomography angiography.\(^1,2,4,6-11,14,16\)

Urgent investigation of serum lipid levels should be sought, along with possible causes of elevated triglyceride levels, such as pancreatitis or a familial lipid disorder.\(^1,6,10\) There is an increased risk of cerebrovascular disease, coronary artery disease, myocardial infarction, atherosclerosis, and pancreatitis with lipemia retinalis, so urgent management with a primary healthcare provider is imperative.\(^6,7,9,10,15\)

Treatment options include medication, diet, and lifestyle modifications.\(^6,9,10\) Standard pharmacotherapy typically includes one or more of the following medications: fibrates, nicotinic acid, polyunsaturated fatty acids, volanesorsen, and statins.\(^6,9,10,11\) The standard treatment goal is to maintain the fasting triglyceride levels <500 mg/dL to help prevent the onset of pancreatitis.\(^6,11\) The retinal blood vessel appearance returns to normal (i.e., reddish hue) with appropriate treatment of the underlying triglyceridemia.\(^4,6,8,10,12,15\) Normalization of the lipemia retinalis appearance occurs quickly in reverse order of appearance onset and reportedly occurs within 1-3 weeks in most cases after initiation of appropriate treatment.\(^4-6,8,10,12,15,17,18\)

### Conclusion

This case highlights the importance of recognizing the retinal signs of lipemia retinalis and referring urgently to the patient’s primary care physician for timely lab work and initiation of systemic medications to help minimize associated ocular and systemic risks.

### References


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