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Case Report

Lipemia Retinalis Diagnosed Incidentally After Laser Photocoagulation Treatment for Retinopathy of Prematurity

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Abstract

A preterm infant who underwent bilateral laser photocoagulation for the treatment of stage 3 retinopathy of prematurity (ROP) is presented because she was incidentally diagnosed with grade 3 lipemia retinalis by dilated fundoscopy at post-laser 2 weeks. Meticulous ophthalmologic examination is imperative in premature newborns for not only ROP screening but also detecting any concomitant ocular abnormalities, which can be sight-threatening or even life-threatening.

Keywords: Hypertriglyceridemia, lipemia retinalis, newborn, retinopathy of prematurity

Introduction

Lipemia retinalis (LR) is an unusual and rare pathology characterized by whitening of the retinal vessels due to high plasma triglyceride levels and scattering of light by triglyceride-laden chylomicrons. Early findings are generally observed in the peripheral retina; however, pathognomonic symptoms tend to appear at the posterior pole when plasma triglyceride levels exceed 2,500 mg/mL.[1,2,3] The disease can be graded as early, moderate, or marked according to retinal appearance.[4] As long as LR is not associated with complications like retinal vein occlusion, it generally does not interfere with visual acuity.[4] However, various electroretinographic changes may be noted in cases with LR.[5]

Case Report

The first daughter of non-consanguineous parents was born prematurely at a gestational age of 26 weeks. Her birth weight was 680 grams and she had grade 1 intraventricular hemorrhage with patent ductus arteriosus as well as respiratory distress syndrome. She was first screened for retinopathy of prematurity (ROP) at postnatal 6 weeks and was diagnosed with stage 1 zone 2 ROP. During her follow-up examinations, the baby developed bilateral stage 3 zone 2 ROP with plus disease, and was referred to our pediatric retina unit for laser photocoagulation at 35 weeks of gestational age. Her medical history revealed mechanical ventilation for 4 weeks during her neonatal intensive care unit stay, where she received erythrocyte transfusion and her patent ductus arteriosus closed completely with ibuprofen treatment.

Prompt laser photocoagulation treatment was scheduled soon after her initial ophthalmological examination at our pediatric retina unit. At the post-laser second week visit, dilated fundoscopy showed widespread pigmented laser burns sparing the posterior pole, and significant regression of plus disease without complications in both eyes (Figure 1A, B). On the next post-laser visit at 39 weeks of gestational age, dilated fundoscopy

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revealed no progression in ROP disease; however, bilateral retinal findings were compatible with LR. Color fundus photography also showed that all retinal vessels had a milky aspect with salmon-colored retina appearance bilaterally (Figure 2A, B). There was no family history of any lipid disorders. Since the diagnosis of grade 3 LR was made, consultation was immediately requested with the division of pediatric metabolism and blood sampling was scheduled. Her serum triglyceride level was measured as higher than 10,000 mg/dL (normal range: <150 mg/dL) and her serum low- and high-density lipoprotein levels were also found to be 490 mg/dL (normal range: <130 mg/dL) and 19 mg/dL (normal range: >40 mg/dL), respectively. The blood sample was otherwise within normal range for liver, kidney, hematologic, and endocrinologic functions, thereby ruling out secondary hyperlipidemia. Her general physical examination also did not reveal any symptoms such as hepatosplenomegaly or xanthomas that could be associated with high triglyceride and/or cholesterol levels. Simultaneously, molecular genetic testing for apolipoprotein C-II deficiency and lipoprotein lipase (LPL) deficiency was performed by sequencing of *APOC2* and *LPL* genes, respectively, and revealed normal results.

The newborn was then started on a strict low-fat diet consisting of 10-15% of her daily caloric intake comprised of fat with weaning on skimmed milk. Serum triglycerides and total serum cholesterol levels gradually decreased to normal range within 6 weeks. On her last visit, dilated fundoscopy showed effective laser burns with no signs of ROP activity, as well as total regression of LR (Figure 3A, B). Her parents were strongly advised to follow her low-fat diet and continue close follow-up by a metabolic diseases specialist.

Discussion

Careful fundoscopic screening of premature infants is mandatory to identify the signs of ROP. Moreover, various ocular

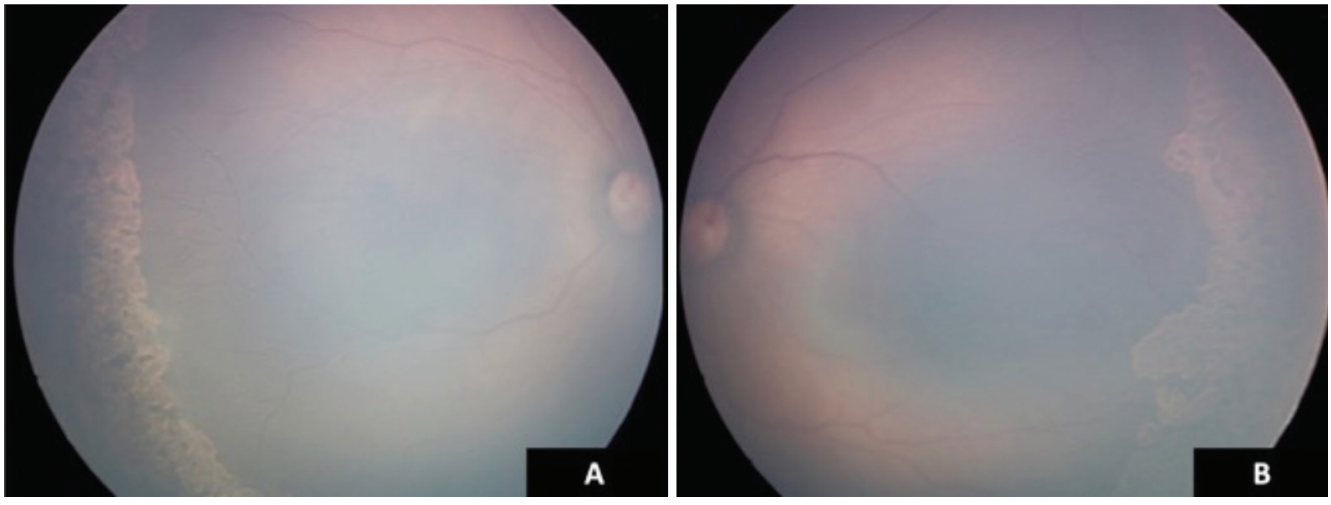

Figure 1. Color fundus photographs of the right (A) and left (B) eye after laser photocoagulation

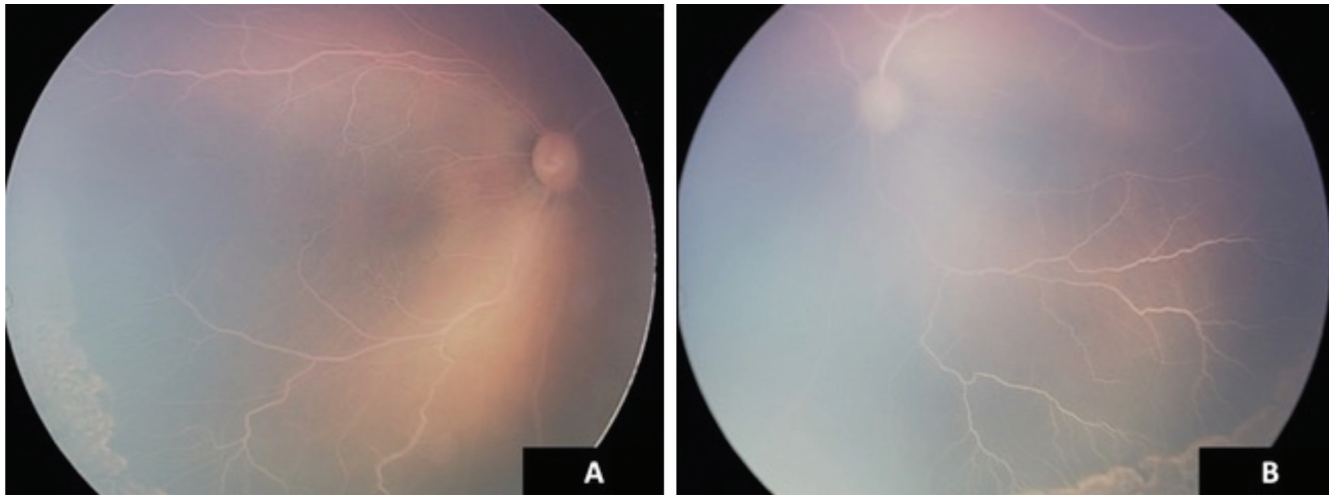

Figure 2. Color fundus photographs depicting milky colored retinal vessels with salmon-colored retina appearance in the right (A) and left (B) eye 4 weeks after laser photocoagulation

and retinal pathologies can be diagnosed incidentally during dilated routine examinations.[6] Several metabolic disorders can also be detected in newborns during a thorough ophthalmic examination, even in very early stages before any prominent symptoms and signs become evident. In 1880, Heyl first described LR, which is characterized by milky-creamy white colored retinal vessels. This typical appearance generally occurs when serum triglyceride level is higher than 2500 mg/dL. Isolated hyperlipidemia without accompanying hypertriglyceridemia does not present with LR.[1,2,3,4,5,6,7] Hypertriglyceridemia may also occur in familial disorders like apolipoprotein C-II deficiency, LPL deficiency, and endogenous circulating low-density lipoprotein inhibitor.[1] Yin et al.[8] reported a homozygous p.G215E mutation in the *LPL* gene in a 6-week-old full-term baby with hypertriglyceridemia and LR.

Babies born preterm and those with intrauterine growth restriction may be more prone to LR due to major risk factors for hypertriglyceridemia such as low LPL activity, parenteral nutrition, and consumption of medium chain triglyceride formulas, which are very common supplements for low-birth-weight infants.[6,7,8] Persistence of uncontrolled high serum levels of cholesterol and triglycerides may lead to possibly fatal morbidities including premature atherosclerosis and coronary artery disease, pancreatitis, and hepatic failure, especially in preterm and term newborns and infants.[6,7,8,9,10] Hence, early diagnosis of LR in such cases is crucial. Although it is known that LR generally does not affect visual acuity in sufferers, Dinc et al.[11] reported a pregnant Turkish women with no family history of hyperlipidemia who developed bilateral severe visual deterioration and whose visual acuity improved after delivery and increased to 20/20 with a normal fundoscopic appearance in both eyes after low-fat diet, physical exercise, and drug treatment with 3-hydroxy-3-methyl-glutaryl-coenzyme A inhibitor.

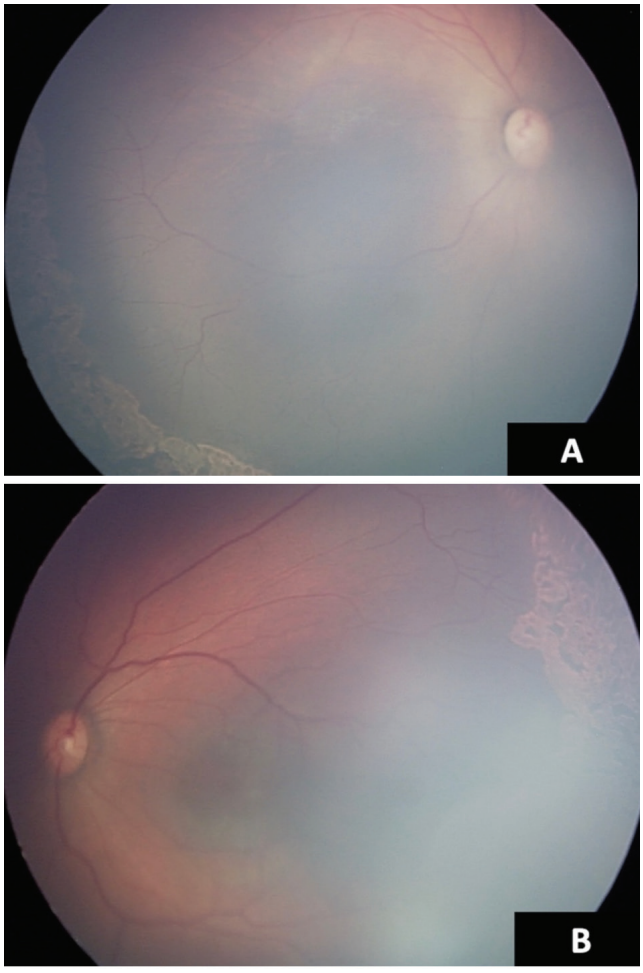

Figure 3. Totally normal color fundus photographs of the right (A) and left (B) eye after low-fat diet

In the present case, retinal findings totally regressed in accordance with the rapid decrease in serum triglyceride and cholesterol levels after the commencement of a strict low-fat diet. Systemic assessment and pertinent laboratory tests should be carried out in patients with LR in order to determine the underlying cause, which may seriously affect the end organs due to high serum lipid levels. Therefore, early diagnosis can even be life-saving. Moreover, examination of the parents and siblings, genetic counseling, and dietary recommendations are among the essential steps to be taken.

Ethics

Informed Consent: Since presented case is a minor, detailed ınformed consent was obtaıned from her parents before laser photocoagulation for the treatment of retinopathy of prematurity.

Peer-review: Externally and internally peer reviewed.

Authorship Contributions

Surgical and Medical Practices: T.Ö., P.T.K., H.O., Concept: T.Ö., A.O.S., Design: T.Ö., E.K.Y., T.K.K., Data Collection or Processing: T.Ö., E.K.Y., P.T.K., Analysis or Interpretation: T.Ö., A.O.S., Literature Search: T.Ö., E.K.Y., Writing: T.Ö., E.K.Y.

Conflict of Interest: No conflict of interest was declared by the authors.

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References

1. Zahavi A, Snir M, Kella YR. Lipemia retinalis: Case report and review of the literature, JAAPOS. 2013;17:110-111.
2. Shinkre ND, Usgaonkar UPS. Lipemia retinalis in a 27 day old neonate: A case report. Indian J Ophthalmol. 2019;67:954-957.
3. Vinger PF, Sachs BA. Ocular manifestations of hyperlipoproteinemia. Am J Ophthalmol. 1970;70:563-573.
4. Nagra PK, Ho AC, Dugan JD Jr. Lipemia retinalis associated with branch retinal vein occlusion. Am J Ophthalmol. 2003;135:539-542.
5. Lu CK, Chen SJ, Niu DM, Tsai CC, Lee FL, Hsu WM. Electrophysiological changes in lipemia retinalis. Am J Ophthalmol. 2005;139:1142-1145.
6. Jain A, Mochi TB, Braganza SD, Agrawal S, Shetty BK, Pachiyappan B. Lipemia retinalis in an infant treated for retinopathy of prematurity. J AAPOS. 2017;21:254-257.
7. Cermakova I, Pattenazzo A, Perrone S, Burlina A, Zannin ME. Grade III lipaemia retinalis in a newborn. Acta Ophthalmol. 2010;88:141-142.

8. Yin HY, Warman R, Suh EH, Cheng AM. Exceptionally elevated triglyceride in severe lipemia retinalis. Int Med Case Rep J. 2016;9:333-336.
9. Horton M, Thompson K. Lipemia retinalis preceding acute pancreatitis. Optometry. 2011;82:475-480.
10. Cortner JA, Coates PM, Liacouras CA, Jarvik GP. Familial combined hyperlipidemia in children: clinical expression, metabolic defects, and management. J Pediatr. 1993;123:177-184.
11. Dinc D, Dincer N, Eltutar K. Lipemia retinalis: Case report. Turkiye Klinikleri J Med Sci. 2012;32:1433-1436.