Grade III Lipemia Retinalis in Premature Infant with Hyperlipidemia: A Rare Case

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ABSTRACT

Lipemia retinalis is a very rare ocular manifestation of hypertriglyceridemia, characterized by white creamy hue discoloration of retinal vessels, due to elevated triglyceride level over 2500 mg/Dl. Grading is divided as early (I), moderate (II), or marked (III) based on clinical appearance. To report a premature infant presented with grade III lipemia retinalis and vitreous opacity. A twelve-week-old male infant admitted with white pupil in the right eye, 4 days prior to admission. History of premature birth (31 weeks) and low birth weight (1700 grams) were present. Visual acuity was uncentral, unmaintained, and unsteady in both eyes. Right eye showed leukocoria, poor pupillary dilatation and absence red reflex, while whitish optic disc, creamy appearance of all retinal vessels and salmon pink appearance of the retina in the left eye. Ultrasonography of the right eye showed vitreous opacity. Hematological examination revealed milky white serum, triglyceride 9974 mg/Dl, total cholesterol 631 mg/Dl, low-density lipoprotein (LDL) cholesterol 195 mg/Dl, high-density lipoprotein (HDL) 12 mg/dl and apoprotein B 196 mg/Dl. The patient was diagnosed with grade III lipemia retinalis and vitreous opacity. White creamy appearance of retinal vessels in infant may manifested from extremely high level of triglyceride. Therefore, comprehensive laboratory examination including lipid profile is necessary.

Keywords: Lipemia Retinalis, Premature Infant, Vitreous Opacity, Hyperlipidemia, Hypertriglyceridemia

I. INTRODUCTION

Lipemia retinalis is an unusual ocular manifestation of severe hypertriglyceridemia, characterized by white creamy hue discoloration of retinal vessels [1]. It was extremely rare in children, but may occurs in premature or full-term infants and could be diagnosed simultaneously with retinopathy of prematurity (ROP) [2,3]. Since it was first described in 1880 by Heyl, very few lipemia retinalis have been reported in infants and children, mainly grade III [4,5,6]. In Indonesia, no child with lipemia retinalis associated with hyperlipidemia have been reported. We report a premature infant who presented grade III lipemia retinalis and vitreous opacity. To the best of our knowledge, lipemia retinalis in a premature infant, accompanied by vitreous opacity in the other eye, has never been previously reported.

II. CASE PRESENTATION

A twelve-week-old male infant admitted to Pediatric Eye Center Hasanuddin University Hospital with white pupil in the right eye, four days prior to admission. He was the firstborn of non-consanguineous marriage and had normal antenatal history. His birth history was premature (31 weeks gestational age), low birth weight (1700 grams) and had history of oxygen administration for 3 full days. Nutrition was based on breastfeeding, but there was a history of premature formula milk consumption. There was no significant illness history. Neither previous ROP screening nor eye examination were performed. There was no family history of hyperlipidemia.

Ophthalmologic evaluation revealed visual acuity was uncentral, unmaintained, and unsteady in both eyes, but the right eye had slower response. Anterior segment of the right eye showed leukocoria and poor pupillary dilatation, while the left eye showed normal anterior segment (Figure 1)

Figure 1. The right eye showed white and poor pupillary dilatation. The left eye showed bright red reflex and normal dilated pupil
Funducopic examination of right eye showed no red reflex due to opacity behind the lens (Figure 2A). Left eye presented whitish optic disc, salmon pink appearance of the retina and creamy white appearance of all retinal vessels (Figure 2B).

![Figure 2](image)

**Figure 2.** The right eye showed diffuse opacity behind the lens (A). The left eye (B) showed whitish optic nerve head (yellow arrow), white creamy retinal vessels in posterior pole and peripheral area (red arrow), with salmon pink appearance of the retina (white arrow).

Ultrasonography of right eye showed vitreous opacity (Figure 3). The patient was referred to vitreoretina subdivision and planned for further posterior segment evaluation. Then, he was prepared for evaluation under anesthesia.

![Figure 3](image)

**Figure 3.** Ultrasonography demonstrated vitreous opacity (white arrow) in the right eye (A). Normal in the left eye (B).

The laboratory staff had difficulty taking the blood samples during laboratory examination process due to high viscosity of the blood. But finally succeeded after the third attempt by using a large syringe. The blood specimen showed milky white color (Figure 4), and the clinical pathologist suggested for lipid profile examination to identify the possibility of hyperlipidemia.
Hematological investigation revealed blood glucose was 95 mg/dL and extremely high lipid profiles. Triglyceride 9974 mg/dL, total cholesterol 631 mg/dL, LDL 195 mg/dL, and apoprotein B 196 mg/dL, while HDL reduced to 12 mg/dl. The interpretation was very severe hypertriglyceridemia, hypercholesterolemia, elevation of LDL and apolipoprotein B. The patient’s parents had normal lipid profiles and no funduscopic abnormality. This patient was diagnosed with grade III lipemia retinalis and vitreous opacity.

The patient was consulted to pediatrician for further hyperlipidemia evaluation and management. The result was weighed 3700 grams, microcephaly, the vital signs and other physical examination were within normal limit. The patient was planned for plasmapheresis treatment since limited administration of lipid pharmacotherapy in infant. However, the parents did not agree with the treatment.

Lipemia retinalis is an unusual funduscopic abnormality caused by an elevation of triglyceride plasma levels over 2500 mg/dL (normal <200 mg/dl) [1,4,7]. Hypertriglyceridemia occurs as result of primary disorder as genetic mutation or secondary to other diseases [1,5,4]. Secondary hyperlipidemia associated with other disease such as diabetes mellitus, biliary obstruction, nephrotic syndrome, pancreatitis, hypothyroidism, obesity, renal failure, systemic lupus erythematosus, alcoholism, medications (estrogens, beta-blockers, steroid, second generation antipsychotic, antidepressants, accutane, rosiglitazone, thiazides, bile acid sequesterants, sirolimus, antiretroviral therapy) [1,3,4,8]. In order to no history or symptoms of secondary causes were detected; thus, we suspected this case was a primary disorder.

Primary hypertriglyceridemia may lead to disruption of triglyceride synthesis or metabolism. The clinical features include systemic disorders and ocular abnormalities (palpebral xanthelasma, lipid keratopathy, lipemic aqueous, iris and retinal xanthomas, sludge retinal vein, red cell aggregation, branch vein occlusion with a marked exudative appearance, and lipemia retinalis) [9,10]. According to the Fredrickson classification, primary hypertriglyceridemia can be clinically classified into five types based on patterns of lipoprotein fractions [8-11].

Lipemia retinalis have been reported only in type I, III, and V which associated with elevation of chylomicron levels [9,10]. The three types presenting lipemic serum, very high triglyceride levels >1000 mg/dl and similar clinical symptoms. However, in type I and III the elevation of total cholesterol, triglycerides, chylomicron levels without an increase in very low density lipoproteins (VLDL) as in type V. Therefore, LDL and apo B values were usually low or normal. Based on lipid profile findings in this patient, we suspected hyperlipidemia was due to type V hypertriglyceridemia, also known as primary mixed hyperlipidemia. Type V, as a polygenic form of hypertriglyceridemia, has a complex genetic etiology, its inheritance pattern is variable, and rarely show familial occurrence. However, further genetic tests targeting potential genetic defects in triglycerides metabolism are required to establish the diagnosis [12,13].

Chylomicron is the largest lipoprotein particles, its accumulation in the retinal and choroidal circulations induces fundus discoloration due to light scatter, and resulting in pink salmon appearance in lipemia retinalis [4,5,10]. The grading is divide based on distribution of fundus discoloration, which is related to triglycerides levels (Table 1) [1,4,7]. The vessels which first affected are in peripheral zone and posterior pole later [1,10]. Based on this classification, left eye was diagnosed with grade III, which has rarely been described [4]. This grade represented his very high triglycerides level which is more than 5000 mg/dL.

Table 1. Grading of lipemia retinalis

<table>
<thead>
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<th>Grade</th>
<th>Intensity</th>
<th>Triglyceride levels</th>
<th>Fundus appearance</th>
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<tr>
<td>I</td>
<td>Early</td>
<td>&gt; 2500 mg/dL</td>
<td>White and creamy aspect of peripheral retinal vessels</td>
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Generally, lipemia retinalis is an asymptomatic disease even with extensive involvement of the vessels [3,10]. Visual acuity usually has no deterioration, but electroretinogram (ERG) amplitude may be decreased [3,4,7,14]. Lu and colleagues have described that after correcting the lipid levels, the abnormal findings were resolves within one week [14]. This condition may be temporary and improve with decreasing triglyceride levels [3]. However, the persistent lipemia retinalis can induce massive irreversible lipid exudation and lead to vision loss [10].

As the only visible human microvasculature, retinal vessels provide information about systemic vascular condition. In this case, accumulation of fatty matter that presented as creamy white retinal vessels, in accordance with this patient’s lipemic blood samples. This lipemic serum could lead to poor circulation in the whole body [15]. Fortunately, retinal circulation has autoregulation system that maintain retinal perfusion pressure relatively constant despite changes in the ocular perfusion pressure [16,17]. However, systemic circulation not autoregulate equally [17]. Therefore, this extreme hyperlipidemia condition may result in more severe damage in the other organs. This prolonged effect is not only leading to permanent visual loss but also to particular life-threatening complications, including hepatosplenomegaly, pancreatitis, coronary heart disease and other manifestations of atherosclerosis [10,18].

Vitreous opacity in the right eye was still unclear whether due to secondary of lipemia retinalis or other disease. We assumed that hyperlipidemia leads to retinal ischemic, increasing of vascular permeability, lipid particles leaked from lipemic serum eventually accumulated in vitreous cavity. Management of lipemia retinalis is immediately treat the underlying condition to obtain fundus and ERG normalization [10]. Hypertriglyceridemia treatment aims to prevent the risk of pancreatitis and/or early cardiovascular disease with lifestyle modification alone or combine with drug treatment, particularly in refractory severe hypertriglyceridemia [11,12]. Unfortunately, treatment for hyperlipidemia in this patient was not easy because of limited administration of pharmacotherapy in infant [8,12,19].

Therefore, the most possible management is plasmapheresis. Plasmapheresis has been reported as a valid and safe intervention to reduced triglycerides, chylomicron, LDL and total cholesterol levels. This treatment has been successfully treated primary hypertriglyceridemia, even in some cases that could not be confirmed by genetic studies [11,20,21]. However, delay in treatment in this patient may lead to poorer prognosis for both life and visual function, if compared to the previous study.

### III. CONCLUSION

The white creamy appearance of retinal vessels in infant may manifested from an extremely high level of triglycerides. Lipemia retinalis grading could be a reliable parameter to describe the triglycerides levels. Therefore, comprehensive laboratory examination, including lipid profile, is necessary.

### REFERENCES

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