

## Case Report

### Lipemia Retinalis in an infant with Acute Pancreatitis

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#### Abstract:

A rare case hyperlipoproteinemia in a 22-day-old infant who presented not only high blood levels of cholesterol and triglycerides but also an ocular manifestation described as lipemia retinalis. The fundus abnormality cleared as the lipid profile improved. Lipemia retinalis is an important and reliable parameter of high levels of chylomicrons and triglycerides and should be considered as a significant clue while diagnosing.

**Key words:** Lipemia retinalis, Pancreatitis, Hyperlipoproteinemia, Hyperlipidemia

#### Introduction:

Heyl was the first to describe lipemia retinalis in 1880(1). This rare condition can appear with some types of hyperlipidemia (2). It consists of a creamy white aspect of the retinal vessels on fundus examination (3).

The variation of the aspect in the fundus has been graded: grade I(early) - white and creamy aspect of the peripheral retina vessels; grade II(moderate) - the creamy color of the vessels extends towards the optic disc ;grade III (marked) - the retina appears with a salmon color and all arteries and veins present a milky aspect (2,4). The aspect of grade III has rarely been described (2).

Hyperlipoproteinemia is classified into five types, all with high plasma levels of lipoproteins and triglycerides. Type I: hyperchylomicronemia and hypertriglyceridemia and confirmed by lipoprotein lipase deficiency; Type II: major elevation of the LDL (low density lipoprotein), VLDL (very low density lipoprotein), cholesterol and triglyceride; Type III: high levels of LDL, VLDL, cholesterol and triglyceride; Type IV: elevation of VLDL and triglyceride; Type V: elevated levels of VLDL, chylomicrons, cholesterol and triglycerides (3,5).

The ocular features are: xanthelasma, iris and retinal xanthomas, lipid keratopathy and lipemia retinalis (5).

We decided to describe this case because especially lipemia retinalis grade III is itself rare and rare especially again in a child with only 22 days of life and such an early presentation is not described in literature.

#### Case Report:

22-day old Full term low birth baby admitted in Neonatal intensive care unit with 1950 gms with history of necrotizing

enterocolitis, neonatal jaundice, abdominal distension was referred for eye examination.

On ophthalmologic evaluation, both pupils were noted to be equal in size and shape, corneas were transparent, there was normal ocular tension and indirect ophthalmoscopy revealed grade III lipemia retinalis; whitish optic disc, creamy white appearance of all retinal blood vessels, indistinguishable veins and arteries, except by their large caliber and ivory hue of the fundus (Figure 1a,b,c)

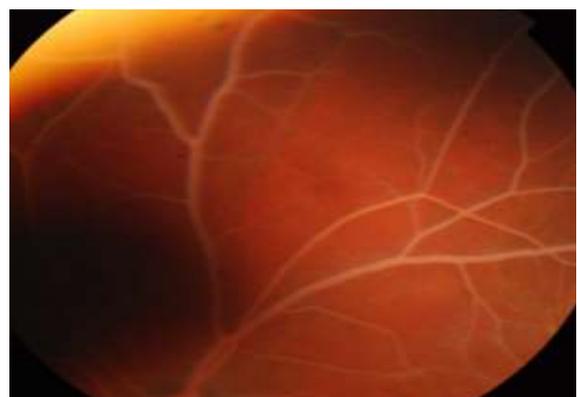
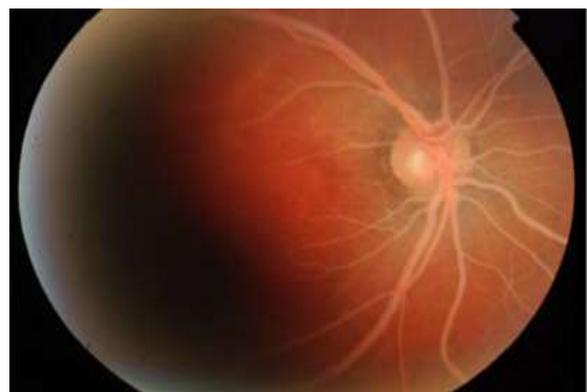




Figure 1a,b,c - showing creamy white appearance of all retinal blood vessels

Based on her ocular findings, her blood investigations were ordered; the blood and radiological tests were diagnostic of acute pancreatitis. Blood was pink in color and serum was latescent (figure2), with gross elevation of triglycerides (41.20 mmol/L, reference interval 0.56 to 2.28 mmol/L) and total cholesterol (56.11 mmol/L, reference interval 2.20 to 4.21 mmol/L). A principal diagnosis of type 1 primary hyperlipidemia (Fredrickson classification) was made.

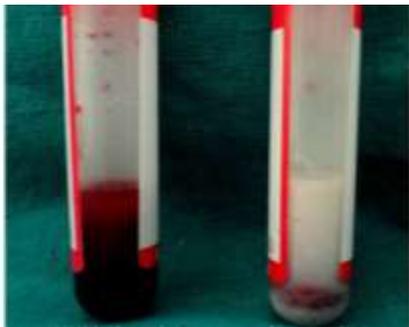


Figure 2 showing pink colour blood

Anti-infective, dietary and supportive care was administered. After a week, the lipid profile was improved: total serum cholesterol level 7.96 mmol/L; triglyceride level 12.87mmol/L and the fundi appeared normal. To our knowledge, this is the youngest patient reported with Type 1 hyperlipidemia associated with lipemia retinalis

The child survived till 4 months of age and then died due to pancreatitis.

#### Discussion:

Lipemia retinalis is an asymptomatic condition that can appear when high levels of triglycerides are present in the circulatory system (5). The retina's white milky aspect occurs when the plasma levels are near to 2000 to 2500 mg/dL or more (1-2).

It is believed (5) that the changes in the fundus are inconsequence of the elevated levels of circulating chylomicrons at the retinal vessels, but some authors also (6) defend that only the high levels are not sufficient to cause the lipemic creamy serum. The same idea is defended by other authors (7) who justified that they have some cases with elevated levels of chylomicrons and triglycerides that did not present lipemia retinalis suggesting that changes in hematocrit and in the translucence of the retinal and choroid vessels

should be considered.

After intestinal absorption the triglycerides enter the circulatory system and the chylomicrons are the way triglycerides are transported. The chylomicrons are the largest lipoprotein macromolecules, 100 to 1000 nm; VLDL are usually smaller, but some can reach 100 nm. This smaller lipoprotein is important for transportation of fat in the metabolism but does not contribute to the change of serum color (6). The blood with high levels of chylomicrons can cause the lipemia retinalis fundus at the retina and choroid vessels(2). Some authors(5) believe that the fact responsible for this effect is the dispersion of the light caused by the chylomicrons.

The deficiency of lipoprotein lipase is rare in hyperlipoproteinemia, and is characterized by the high levels of chylomicrons in serum. It is an autosomal recessive disease with an incidence of less than 1:1.000.000. Lipoprotein lipase is present in the vascular endothelium and breaks the chylomicrons into glycerol and free fatty acids; the deficiency of lipoprotein lipase causes the accumulation of chylomicrons (3).

The predetermined treatments for these cases are restriction of fat in the diet (less than 10 g/day) and elevated intake of protein and carbohydrate with anti infective therapy. Medium chain triglycerides can help because their hydrolysis is not dependent on lipoprotein lipase. The ingestion of fat soluble vitamin supplements is also necessary (3). In 1996, an article presented seven cases of hyperlipidemia but all of them were in adults (5).

#### Conclusion:

Lipemia retinalis is an important and reliable parameter of high levels of chylomicrons and triglycerides and should be considered a significant clue while diagnosing hyperlipoproteinemia

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