University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 **2021, Vol. 7(1)**

A rare case of Lipemia Retinalis

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Abstract : 38days old male child, a case of hypertriglyceridemia combined hyperlipidemia, with normal prenatal, natal and postnatal history came for evaluation. Both eyes anterior segment was normal. Fundus examination of both eyes showed a clear media, disc appeared normal, salmon colored retinal arteries and veins without skip areas were seen. Serum triglycerides level increased three hundred times. Serum cholesterol level showed ten folds increase. Aim of treatment is to lower the lipid levels. This case is published for its rarity.

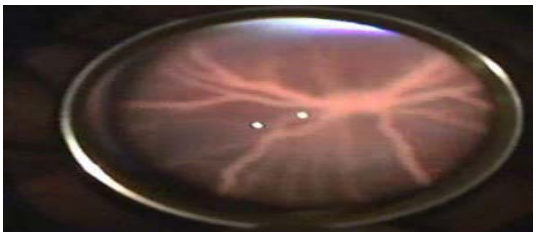
Keyword : Lipemia retinalis, hypertriglyceridemia, hypercholesterolemia, chylomicrons

Introduction

Lipaemia retinalis refers to salmon-colored retinal arteries and retinal veins due to elevated lipids, most commonly hypertriglyceridemia. It is an ophthalmic manifestation of a systemic disorder. It is a rare finding.

Case report

38 days old Mohamed Arshad was referred from Institute of child health as a case of **hypertriglyceridemia/combined hyperlipidemia** for ophthalmic evaluation. First born child, full term normal vaginal delivery, cried immediately at birth, second degree consanguineous marriage, no history of neonatal intensive care unit admission. Both eyes anterior segment was normal. Both eyes fundus examination showed clear media, disc appeared normal, **salmon colored retinal arteries and veins without skip areas were seen.**



Fundus showing salmon colored retinal arteries and veins without skip areas

Laboratory values Normal values

Serum lipase – 62 (145-216) U/L
Lactate dehydrogenase – 346 (150 - 360) U/L
Lipoprotein A – **>50** (Upto 30)mg/dL
Serum cholesterol – **2000** (>170)mg/dL
High density lipoprotein – 45 (35- 82) mg/dL
Low density lipoprotein – **310** (63 - 140) mg/dL
Serum triglycerides – **30650** (27 - 125)mg/dL
Total cholesterol/High density lipoprotein – 44.4 (<4.5)
Highly milky blood sample

Highly milky blood sample of this patient

Treatment given at Institute of child health

Diet given was skimmed milk Child was put on fenofibrates
Exchange transfusion was done to reduce triglycerides to <1000 mg/dl Child was followed up periodically

Discussion

Lipaemia retinalis has been graded O to III by Vinger and Sachs. 4 Lipemia retinalis was first described by Heyl in 1880.

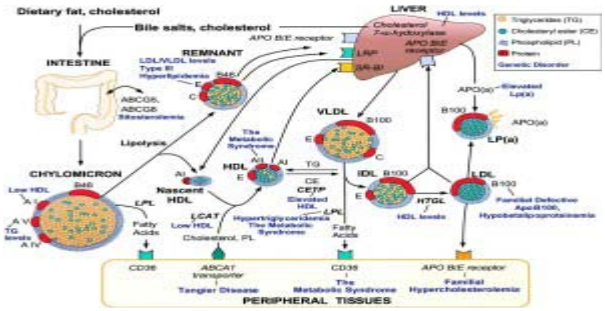
Etiology

- Type I and V hyperlipoproteinemia4
- Poorly controlled diabetes mellitus5
- Mixed retinopathy

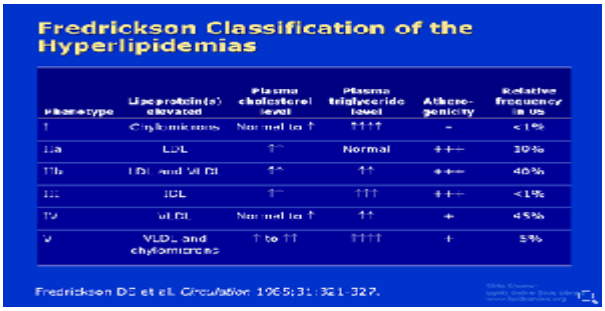
Pathophysiology

Elevated serum lipid levels may cause retinal vascular obstruction. The lipid abnormalities are typically heritable lipid metabolic abnormalities. The visualization of high levels

of **chylomicrons** in blood vessels accounts for the fundus appearance.1



cholesterol metabolism pathways



Fredrickson Classification of Hyperlipidemias

Type 1 hyperlipoproteinemia

Autosomal recessive disorder.

Incidence 1 in 1,000,000

Type I hyperlipoproteinemia exists in several forms:

Lipoprotein lipase deficiency (type Ia), due to a deficiency of lipoprotein lipase (LPL) or altered apolipoprotein C2, resulting in elevated chylomicrons.4 Familial apoprotein CII deficiency (type Ib) a condition caused by a **lack of lipoprotein lipase activator**. Chylomicronemia due to **circulating inhibitor of lipoprotein lipase (type Ic)** Type I hyperlipoproteinemia usually presents in childhood with eruptive xanthomata and abdominal colic. Complications include retinal vein occlusion, acute pancreatitis, steatosis and organomegaly, and lipaemia retinalis.

Clinical signs

Pale conjunctiva may result from lipemic conjunctival blood vessels.2 Fundus examination reveals salmon colored retinal arteries and veins without skip areas.2 Serum triglycerides usually tend to exceed 1000mg/dL when this diagnosis is made.3

Differential Diagnosis

Retinal vascular whitening can be observed in prior **retinal vascular occlusion or in retinal vasculitidies**. These are typically confined to the retinal arterial or venous systems and typically have **skip segments** where the retinal vasculature is unaffected.2 Lipaemia retinalis shows diffuse retinal arterial and venous involvement.

Prognosis and management

The visual prognosis can be good for patients who do not develop retinal vascular occlusive disease.2 Treatment is directed towards **decreasing the causative lipemic factor**.2

Reference

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