Lipemia Retinalis due to Secondary Hyperlipidemia in Type 1 Diabetes Mellitus

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Abstract
Lipemia retinalis is a rare and asymptomatic condition which occurs when high levels of triglycerides and chylomicrons are present in blood. We report a rare case of secondary hyperlipoproteinemia in a 27 year old type 1 diabetes patient who presented with diabetic ketoacidosis and this peculiar ocular manifestation. The fundoscopic abnormality and creamy white serum cleared as the level of chylomicrons in the plasma dropped with intensive insulin therapy.

Introduction
Lipemia retinalis is an unusual retinal manifestation of diabetes mellitus, which occurs in the setting of markedly elevated levels of serum triglycerides; in which retinal vessels become cream colored due to the scattering of light by the triglyceride-laden chylomicrons. It is thought to be directly related to the serum triglyceride level.

Case Report
We report the case of a 27 year old male patient admitted in our hospital with 2 days history of giddiness, vomiting, abdominal pain and difficulty in breathing. He was a recently diagnosed diabetic (Type 1) on ayurvedic treatment. Physical examination showed him to be drowsy, dyspneic, dehydrated and with other features suggestive of diabetic ketoacidosis (DKA). He had no cutaneous or tendinous xanthomas or xanthelasmas. He had no visual disturbances.

On Day 1, fundus was examined for diabetic retinopathy in the ward and it revealed that vessels in the posterior pole and peripheral area of each eye had a creamy appearance. Both arterioles and veins were flat and ribbon like and their colour was salmon pink around the disc and creamy milk white in the periphery. Optic disc and macula were normal and there was no evidence of diabetic retinopathy. Ophthalmologist opinion was sought for fundus examination and they confirmed the same finding.

The picture was suggestive of lipemia retinalis (Figure 1).

On day 1, blood samples showed milky white serum at the top of blood column. Laboratory study showed a fasting blood glucose level of 568 mg/dl, urine sugar 3+ and urine acetone was positive. His lipid profile showed total serum cholesterol level of 940 mg/dl, serum triglyceride level of 1400 mg/dl, serum HDL of 230 mg/dl and serum LDL of 580 mg per/dl. Other investigations like complete hemogram, renal function test, liver function test, thyroid function test, Serum amylase and Lipase were within normal limits. As there was no evidence of xanthomas, xanthelasmas or hepatosplenomegaly and no family history of hyperlipidemia, primary hyperlipoproteinemic disorders were ruled out. A diagnosis of diabetic ketoacidosis and secondary hyperlipidemia was made. Intensive insulin therapy and supportive care were provided. Atorvastatin 40 mg and Fenofibrate 400 mg once daily were started.

Four days after treatment, laboratory tests showed a rapid decrease in the lipid profile with a total serum cholesterol 300 mg /dl; triglyceride 340 mg/dl; HDL – 70 mg/dl; LDL -230 mg/dl (Table 1) and fasting blood glucose 102 mg/dl.

Examination of the fundus on Day 5, showed a normal appearing retinal vessels except those at the periphery which had a creamy white colour (Figure 2). Fundus fluorescein angiography (Figure 3) was normal indicating normal flow in the retinal vessels.

This is a rare case of lipemia retinalis complicating a newly diagnosed diabetic presenting with DKA with associated secondary Hyperlipidemia. The normalization of serum lipids and reversion of retinal vessel alterations
Lipemia retinalis first described by Heyl in 1880,2 is a rare condition occurring in the setting of markedly elevated triglycerides level, but not of high cholesterol.3 It can occur in all age groups and in both primary and secondary hyperlipidemia. Among primary hyperlipoproteinemias, Lipemia retinalis occurs in type 1 (familial chyomicronemia) and type 5 (familial hypertriglyceridermia) disease.4 The secondary causes are untreated diabetes mellitus, starvation ketoacidosis, chronic alcoholism, acute pancreatitis, nephrotic syndrome, leukemia, hypopituitarism, hypothyroidism, obesity and drugs like high dose thiazide, beta blockers, OCP, tamoxifen, glucocorticoids, isotretinoin, antiretroviral therapy and atypical antipsychotics.

Type I hyperlipoproteinemia is the best-characterized genetic cause of hypertriglyceridermia and is caused by a deficiency or defect in either the enzyme or its cofactor, apo C-II. Lipoprotein lipase which is found in the endothelial cells of capillaries, hydrolyzes triglycerides in chylomicrons and VLDL transforming them into their respective remnants. Lipoprotein lipase requires insulin for its full activity.

Hypertriglyceridermia is the hallmark of diabetic dyslipidemia. In both insulin-deficient (type 1) and insulin-resistant (type 2) uncontrolled and untreated diabetes the activity of lipoprotein lipase enzyme is decreased and triglyceride rich lipoproteins (chylomicrons and VLDL) cannot be metabolized appropriately to their remnants, which imparts a creamy white colour to the blood and the retinal vessels like in our patient.

With the initiation of insulin therapy in our patient, production of TG decreased and further conversion of chylomicrons and VLDL to its remnants gets restored and leads to disappearance of creamy white vessels. Lipemia retinalis is thought to be directly linked with the serum triglyceride level; typically, the retinal findings do not occur until the triglyceride reaches 2500 mg /dl.5 In our patient even though the triglyceride was 1400 mg/dl, characteristic features of lipemia retinalis was present.

In our patient, initial lipid profile value was elevated including triglycerides and HDL. Though the HDL was raised, due to high level of Total cholesterol, the HDL cholesterol was expected to be elevated in a ratio of 4:1 (Total cholesterol/HDL cholesterol).

The repeat lipid profile on 4th day showed total cholesterol- 300 mg/dl; HDL - 73 mg/dl; LDL - 200 mg/dl and the ratio of total cholesterol/HDL cholesterol (4:1) still maintained.

So far was fewer than 100 cases of lipemia retinalis have been reported. Out of them, 75% were due to untreated, only the rest 25% were due to familial hyperlipoproteinemias. Out of diabetics, 75% were males and their average age of incidence was 25.6

In conclusion lipemia Retinalis is a very rare condition in diabetic patients and is reversible with early insulin therapy. This case report highlights the importance of suspecting fundus change (lipemia retinalis) in diabetic patients with DKA, though there will not be any visual symptoms. So fundus examination becomes mandatory in all diabetics.

### References