Course of lipaemia retinalis in acquired generalised lipodystrophy

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DESCRIPTION

Acquired generalised lipodystrophy is a rare skin disorder characterised by severe fat loss in large areas of the body especially the face, arms and legs. This is associated with ectopic fat deposition, leptin deficiency and metabolic abnormalities like insulin resistance, hypertriglyceridaemia, high low density lipoproteins (LDL) and low high density lipoproteins (HDL) levels.¹ It is an extremely rare disease with over 100 cases reported so far in literature. Lipaemia retinalis refers to abnormal deposition to TG in retinal vessels and entire fundus due to underlying high serum triglycerides (TG) levels.² This condition is usually associated with high TG levels >1000 mg/dL.³ Abnormal deposition of TG produces a salmon-coloured fundus associated with creamish discolouration of retinal arteries and veins in the advanced stages. The disease usually begins in the periphery and progresses towards the centre as the TG levels increase. Lipaemia retinalis is usually asymptomatic and does not affect vision until associated with vascular occlusion.⁴ We report a course of a rare ocular manifestation of a very rarely reported systemic disease.

A 45-year-old woman diagnosed with acquired generalised lipodystrophy was referred to us for fundus evaluation. Blood triglyceride and cholesterol levels were more than 10 000 mg/dL and 1050 mg/dL, respectively, at the time of presentation. Best-corrected visual acuity in the right eye (OD) was 6/18, and 6/12 in the left eye (OS). Anterior segment evaluation revealed dense senile cataract in both the eyes (OD > OS). Fundus examination showed presence of salmon coloured fundus with whitening of retinal arteries, and veins suggestive of lipaemia retinalis in both the eyes (OU) (figure 1A shows lipaemia retinalis in the OS, arteries marked

Learning points

- ▶ Routine fundus evaluation is warranted in patients with lipodystrophy syndromes.
- ▶ The clinical ocular findings of lipaemia retinalis should quickly resolve, once the triglyceride levels return to normal.

in blue and veins marked in yellow). Fundus images of the OD were hazy due to presence of dense cataract, and are thus not shown. Loss of vision in OU was attributed to the presence of cataract in both the eyes.

Aggressive treatment with lipid-lowering drugs lead to lowering of triglyceride, and cholesterol levels to 1416 mg/dL and 318 mg/dL, respectively, at 6 months of follow-up. Fundus examination showed complete resolution of lipaemia retinalis in OU (figure 1B shows resolution in OS).

This report highlights the association of very rare ocular condition with acquired generalised lipodystrophy. Patients with lipodystrophy disorders should be referred for fundus evaluation to rule out lipaemia retinalis. Though lipaemia retinalis is visually not disabling, timely intervention to control serum TG levels can reverse the disease, and prevent development of any vascular occlusions or retinal ischaemia.

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Figure 1 (A) shows salmon-coloured fundus with whitening of retinal vessels suggestive of lipaemia retinalis. (B) shows normal fundus glow with resolution of whitening of retinal vessels.

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