Newborn Lipemia Retinalis Resolved with Exchange Transfusion

Lipemia Retinalis do Recém-nascido Resolvida com Transfusão Permuta

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ABSTRACT

The purpose of this study was to report the case of a newborn with lipemia retinalis and the clinical evolution after treatment.

RetCam fundus photographs were taken before and after treatment with exchange transfusion and fat restriction diet.

A 1 month and 25 days old female with a history of 3 days irritability with bilateral leukocoria and a creamy pale blood sample was treated for primary hyperlipidemia. After treatment, symptoms resolved, leukocoria disappeared and fundus exam appearance became normal.

Lipemia retinalis in the newborn can fully resolve with appropriate systemic treatment.

KEYWORDS: Exchange Transfusion, Whole Blood; Hyperlipidemias/complications; Hyperlipoproteinemia Type I/diagnosis; Hyperlipoproteinemia Type I/therapy; Infant; Retinal Diseases/diagnosis; Retinal Diseases/therapy.

RESUMO

O objetivo deste estudo foi relatar um caso clínico de uma recém-nascida com lipemia retiniana e a evolução clínica após o tratamento. As fotografias de fundo de olho foram obitdas antes e após o tratamento com transfusão e dieta com restrição de gordura. A recém-nascida, 1 mês e 25 dias de idade, com história de irritabilidade de 3 dias, leucocoria bilateral e amostra de sangue com aspecto creme pálido, foi tratada para hiperlipidemia primária. Após o tratamento, os sintomas e a leucocoria desapareceram, e a aparência do exame de fundo ocular tornou-se normal. A lipemia retiniana no recém-nascido pode ser totalmente resolvida com tratamento sistêmico apropriado.

PALAVRAS-CHAVE: Doenças Retinianas/diagnóstico; Doenças Retinianas/tratamento; Hiperlipidemias/complicações; Hiperlipoproteinemia Tipo I/diagnóstico; Hiperlipoproteinemia Tipo I/tratamento; Lactente; Transfusão Total.

INTRODUCTION

Lipemia retinalis is a rare ocular manifestation of systemic hyperlipidemia, without significant visual affectation that resolves with systemic treatment and needs no ocular intervention.

CASE REPORT

Our patient is a 55 days old first-born Hispanic female of a 28 years old from a non-consanguineous couple with no history of hereditary or metabolic diseases, nor any major dysmorphology. Had normal pregnancy and birth, documenting only oligoamnios at 38th week, because of which, an induced labor was held five days later. At age 52 days, the pediatrician documented an inguinal hernia, which was reducible. Since then, the child became irritable, and presented a single day febrile peak (38°C), that resolved with acetaminophen. The mother's chief concern was that the infant was inconsolable since that exploration. The Emergency department documented bilateral leukocoria without any other findings, therefore an ophthalmological evaluation was requested. Meanwhile, a complete blood cell (CBC) was ordered, which had a pale creamy pink color. The metabolic count had triglycerides at 37719 mg/dL, total cholesterol at 1070, HDL 363, VLDL and LDL over 400 (the maximum objective measurement). Hemoglobin count was reported 4 g/dL. The rest of the CBC and metabolic count was unreliable because of the above results. At the ophthalmic exam, the patient fixed and followed with each eye. Anterior segments, intraocular pressure (IOP), pupillary reflexes and eye movements were normal. The fundus exam documented both retinas to be pale and the blood vessels were completely white and dilated. The periphery, nerve and macula had no other alterations (Fig. 1). The patient was admitted to the hospital to perform an exchange transfusion at the Intensive Care Unit, and underwent on a parental lipid-free diet until a medium chain triglyceride oil diet was established. Fig. 2 was taken 3 days after the exchange transfusion was performed and the triglycerides level was at 743 mg/dL. The patient continues control at the nutritional and metabolic center of the hospital and at our service with no ocular complications (Fig. 2).

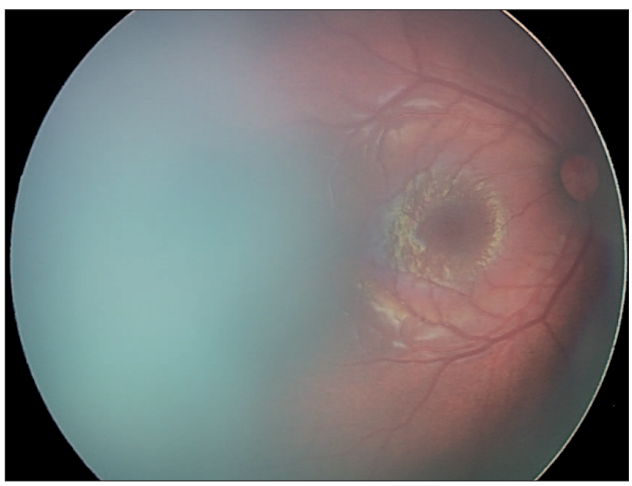

Figure 1. Patients right eye showing a Grade III lipemia retinalis.

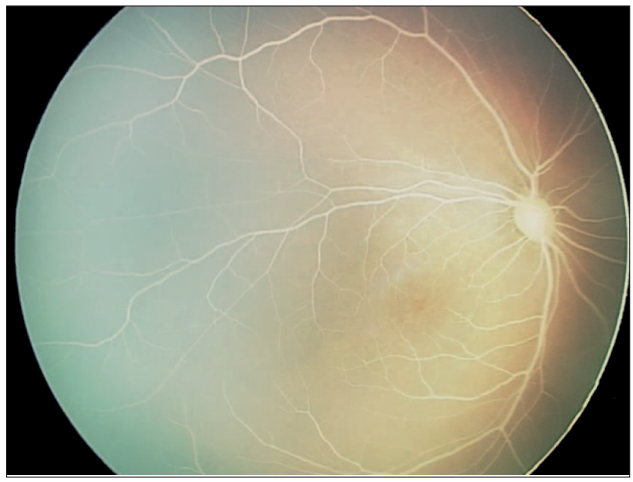

Figure 2. Patients right eye three days after exchange transfusion. The grayish area temporal to the macula is an artefact of the Ret Cam® used for that day´s exploration, fundus exam was completely normal.

DISCUSSION

Lipemia retinalis is a rare condition associated with elevated plasma levels of triglycerides described since 1880. It can be found in people whose triglycerides levels overcome 1000 mg/dL. Cermàkovà *et al*[1] and Rayner *et al*[2] explain that concerning primary hyperlipidemias, it is seen in Type I, III, IV and V (types in which hyperchylomicronemia is seen); type II has high LDL levels, and no case of lipemia retinalis has been described in this hyperlipidemia. Clinical findings grading are: Grade I shows white and creamy aspect of peripheral retinal vessels; Grade II has creamy-colored vessels extending towards the optic disc; Grade III shows salmon colored retina with all vessels having milky aspect, as shown by Nampoothiri *et al*[3]. This condition produces no visual effect. It has only been described to produce electroretinogram changes, such as decreased a- and b- waves responses in both cones and rods. Correction of the lipid levels reverses these changes within a week. Fu *et al*[4] explains that TG levels over 2000 mg/dL predispose to pancreatitis, and also produce falsely low hemoglobin, sodium and bilirubin levels. Treatment is centered to prevent pancreatitis, not the lipemia retinalis. The mainstay of management is fat restriction, for example using medium chain triglyceride oil based diets and a low-fat maintenance diet. Kavey *et al*[5] suggests exchange transfusion (or apheresis) is helpful to treat acute phase and to prevent pancreatitis and congestive heart failure. This case uniqueness is due to the fact that it is the first time lipemia retinalis has been seen with leukocoria as the presenting sign, and this is the highest recorded level of triglycerides in an infant with lipemia retinalis.

CONTRIBUTORSHIP STATEMENT / DECLARAÇÃO DE CONTRIBUIÇÃO:

GRV: Draft of the manuscript, literature research,

LAC: Contributed to the draft of the manuscript, critical review, approval of the final version.

RESPONSABILIDADES ÉTICAS

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ETHICAL DISCLOSURES

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