MANAGEMENT OF BILATERAL CENTRAL VEIN OCCLUSION IN A PATIENT WITH ARTHROGRYPOSIS CONGENITA AND AUTOIMMUNE NEPHROTIC SYNDROME: CASE REPORT

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Purpose

To describe a case of a 30-year-old female with arthrogryposis congenita and autoimmune nephrotic syndrome who presented bilateral central retinal vein occlusion complicated with vitreous hemorrhage treated with pars plana vitrectomy (PPV).

Methods

Case report.

Results

A 30 years old, white, female patient was seen at ER complaining of decreased visual acuity for 5 days in both eyes.

PMx was positive for arthrogryposis congenita and autoimmune nephrotic syndrome. Best correct visual acuity (bcva) was 20/400 in right eye (od) and 20/200 in the left eye (os). Fundus exame showed in both eyes: central retinal vein occlussion, hard exsudates and serous macular edema with retinal detachment (figure 1 and 2).

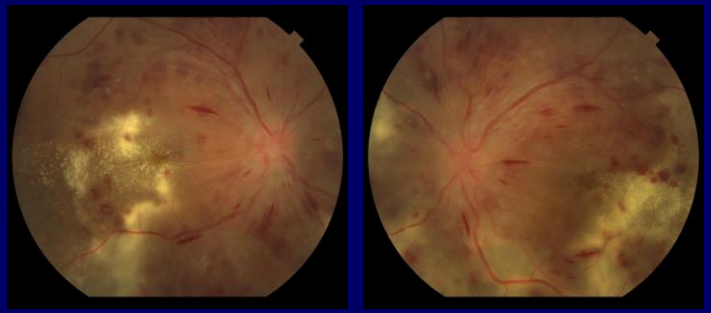

Figure 1: Central retinal vein occlusion in ou.

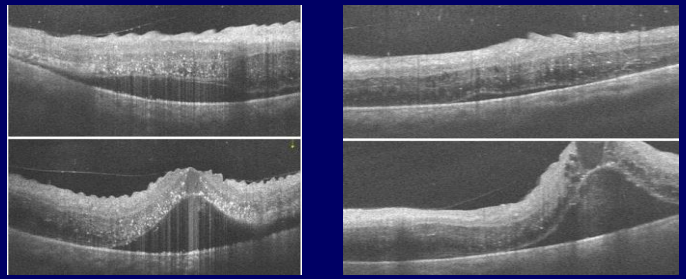

Figure 2 : Macula edema at OCT in ou.

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Patient was initially treated by a nephrologist. Despite normalizing albumin level, the patient persisted with the same fundoscopic and OCT findings and it treated with intravitreous ranibizumab in both eyes.

Five months later, BCVA of OD improved to 20/40, but OS was 20/400 due to persisting non-clearing vitreous hemorrhage.

Patient was submitted to a PPV in OS. At the last of follow-up, BCVA was 20/30 od and 20/25 os.

Fundus examination showed pigmentary clumps in the macula (figure 3). OCT showed foci of disruption of the outer retina and ellipsoid zone with no macular edema (figure 4). Microperimetry showed reduced foveal sensitivity correlated the pigmentary clumps in both eyes.

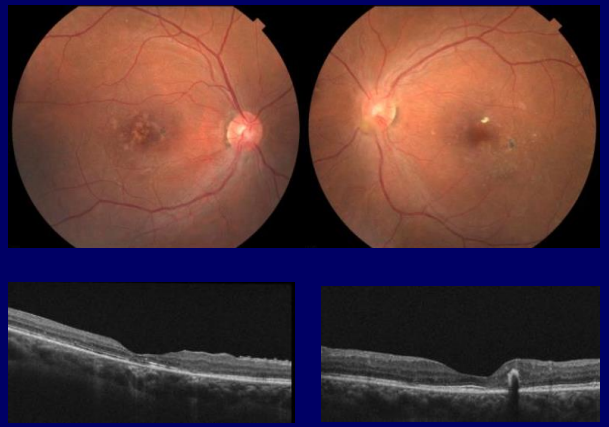

Figure 3 and 4: Fundus examination and OCT in ou.

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Discussion

Arthrogryposis congenita, is a complex, etiologically diverse disease characterized by multiple congenital joint contractures.

This report shows case of bilateral central retinal vein occlusion with macular edema and vitreous hemorrhage with variable course, different from the standard RVO cases. Arthrogryposis and autoimmune nephrotic syndrome were complicating factors in the management of this challenging condition.

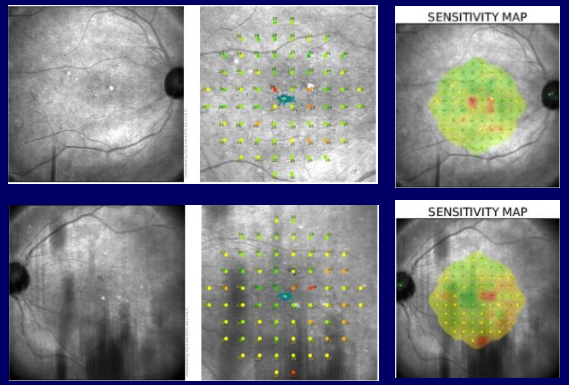

Figure 5: Microperimetry