Short communication

Spontaneous resolution of macular detachment associated with optic disc coloboma

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ABSTRACT

Case report: A 24-year-old male presented with visual acuity loss of the right eye. Examination of the right eye fundus showed a serous detachment of the posterior pole associated with optic disc coloboma. The patient refused surgical treatment. After six months an improvement in visual acuity was observed with the disappearance of macular subretinal fluid.

Discussion: The macular detachment is a known complication of the optic disc coloboma. Different therapeutic options have been used, although spontaneous re-attachment may also occur as shown in this case.

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Resolución espontánea de desprendimiento macular asociado a coloboma del disco óptico

RESUMEN

Caso clínico: Paciente varón de 24 años que acude por disminución de agudeza visual del ojo derecho. La exploración oftalmológica de dicho ojo mostraba un desprendimiento seroso del polo posterior asociado a coloboma del nervio óptico. Se propuso tratamiento quirúrgico que el paciente rechazó. A los 6 meses se objetivó una mejora de la agudeza visual con desaparición del líquido subretiniano a nivel macular.

Discusión: El desprendimiento del polo posterior es una complicación conocida del coloboma del nervio óptico. Se han utilizado distintas opciones terapéuticas, aunque como demuestra este caso también puede acontecer una resolución espontánea.

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Introduction

Optic nerve cup defects are the result of alterations in the development of the papilla and comprise a range of abnormalities including optic disc coloboma, congenital optic disc fossa, morning glory anomaly, peripapillary staphyloma and megalopapilla.\textsuperscript{1}

Optic nerve coloboma is characterized by increased disc size with total or partial excavation (generally inferior and temporal) which is well defined with shiny white surface and with normal retinal vessels along the edge of the defect. The inferotemporal neuroretinal ring is extremely thin or absent while the superior ring is generally well preserved. Involvement can be bilateral or unilateral and can appear sporadically or following a dominant autosomic inheritance pattern.

Patients with optic nerve coloboma may exhibit coloboma-like defects in other ocular structures such as retina and choroids, iris and ciliary body.

Case report

A male, 24 years of age, visited our center due to diminished visual acuity in the right eye (RE) with one month evolution. Personal history included borderline intellectual coefficient, liver transplant due to cryptogenetic cirrhosis and polycitema under study. Family history was not relevant and ophthalmological examination of his parents gave strictly normal results.

The corrected visual acuity (CVA) of RE was 0.3 (\(-1 \text{sph,} -1.25 \text{ at 80° cil}\) and 0.8 (\(-2.5 \text{sph,} -0.75 \text{ at 105° cil}\) in LE. The ocular fundus of RE revealed inferotemporal optic disc coloboma with serous detachment of the posterior pole and yellowish macular sediments. The LE only showed a papillary cup defect at the temporal level with alterations in the papillary vascular pattern and in the peripapillary retinal pigment epithelium (RPE), more compatible with congenital optic disc fossa than with coloboma (Fig. 1A and B). The remainder of the ophthalmological examination produced normal results without finding other ocular colobomatous defects.

Fluorescein angiography (FA) of the RE revealed late hyperfluorescence at the papillary level in the coloboma area and discrete hyperfluorescence from early times at the level of the serous detachment. The macular area also showed heterogeneous hyperfluorescence which increased with time without exhibiting contrasted diffusion at the edges and with hyperfluorescent focal points enhanced over the underlying hyperfluorescence (Fig. 2A–C). The FA of the LE only exhibited hypofluorescence in early times in relation to the optic disc fossa (Fig. 3). RE indocyanine green angiography revealed macular hypofluorescence in late times probably matching the sedimentations shown by the OF (Fig. 4).

Optic coherence tomography (OCT) (Zeiss Stratus, time domain technology) of RE revealed the typical posterior pole serous detachment image with an optically empty space between the RPE and the neurosensory retina. In addition,
hypo-reflectiveness was shown at the level of the fovea in and under the RPE (Fig. 5).

The angiographic image of the RE suggested the presence of a subretinal neovascular membrane, a highly infrequent association with optic disc coloboma even though it has been described in the literature.\(^1\) This could not be confirmed with indocyanine green or OCT. The image was more compatible with the existence of subretinal fibrosis and RPE alterations secondary to the chronic nature of the macular serous detachment.

Discussion

Posterior pole serous detachment is a complication associated to optic disc coloboma which frequently occurs in the second and third decade of life.\(^1,3\) There are several theories

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Fig. 3 – Left eye fluorescein angiograph taken during the first visit to our hospital. Time: 40 s.

Fig. 4 – Right eye indocyanine green angiograph taken during the first visit to our hospital. Time: 7 min.

Fig. 5 – Right eye macular optic coherence tomography of the right eye taken during the first visit to our hospital.

Fig. 6 – Right eye retinograph taken after spontaneous resolution of the serous macular detachment associated to optic disc coloboma.

Fig. 7 – (A and B) Right eye posterior pole optic coherence tomography taken after the spontaneous resolution of the macular serous detachment associated to optic disc coloboma.

Treatment for the RE was proposed with posterior vitrectomy associated to argon laser photocoagulation in the temporal edge of the disc with a fluid/gas exchange, but the proposal was rejected by the patient and his family.

At month 6, the patient referred improved visual acuity and exhibited the following CVA: RE: 0.4 (−1.75 sph, −1.25 at 80° cil) and LE: 0.8 (−2.5 sph, −0.75 at 105° cil). The RE ocular fundus did not exhibit the subretinal liquid at the macular level (Fig. 6), which was confirmed with OCT (Fig. 7), even though RPE alterations were observed.
about the origin of the subretinal liquid (SRL) that causes the serous detachment. One sustains that the SRL proceeds from the vitreous fluid and another that it is due to the diffusion of the retrobulbar fluid within the subretinal space. Some authors also propose a regmatogenous etiology.

Several therapeutic options have been applied for treating macular serous detachment associated to optic papilla coloboma. The best visual results seem to be obtained with a combination of argon laser photocoagulation of the peripapillary area combined with intraocular injection of gas (C3F8 or SF6) and posterior vitrectomy. As demonstrated by the instant case, the serous detachment can also resolve spontaneously. A possible explanation to said spontaneous resolution is the interruption of communication between the coloboma and the papillomacular array due to fibrosis and RPE alteration secondary to the chronic nature of the process.

It is essential to carry out a systemic examination of patients with optic disc coloboma because associated anomalies have been described in various systems including the central nervous, digestive and cardiovascular systems, genitourinary apparatus, muscle-skeletal system, skin and nasopharyngeal apparatus. In addition, the CHARGE association is relevant: coloboma optic nerve, heart disease, choanal atresia, growth retardation, genital hypoplasia and ear abnormalities.

REFERENCES