ENFERMEDAD DE COATS RECIDIVANTE DIAGNOSTICADA EN SUJETO ADULTO

ADULT-ONSET OF RECURRENT COATS DISEASE

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ABSTRACT

Case report: A male diagnosed with Coats disease at the age of 30 years, had a relapse of this condition 14 years later. His first symptom of blurred vision occurred in both episodes and similar findings of unilateral retinal exudation and lipid deposition at boundary were seen. Angiography showed the characteristic early hyperfluorescence of the telangiectasias and late leakage of dye.

Discussion: Coats disease is relatively uncommon and, as the degree of involvement varies from case to case, a randomized, prospective study of treatment would probably not be feasible. Despite the characteristic features of this disease, the differential diagnoses must be considered in both children and adults (Arch Soc Esp Oftalmol 2007; 82: 555-558).

Key words: Coats disease, retinal telangiectasias, retinal exudation, late recurrence, decreased visual acuity.

RESUMEN

Caso clínico: Paciente varón que con 30 años tuvo el primer episodio de enfermedad de Coats y recurrencia 14 años más tarde. Clínicamente ambos episodios tienen como síntoma de inicio visión borrosa y exploración típica de exudación masiva retiniana con corona de exudados duros. La angiografía presenta hallazgos característicos de telangiectasia retiniana con dilatación anormal de vasos con hiperfluorescencia precoz y fuga tardía.

Discusión: Coats es una entidad clínica poco frecuente y con grado de severidad variable, que hacen difíciles los estudios randomizados prospectivos. Pese a los hallazgos característicos, hay que descartar otros diagnósticos tanto en el niño como en el adulto causantes de esa misma clínica.

Palabras clave: Enfermedad de Coats, telangiectasias retinianas, exudación retiniana, recidiva tarda, disminución de agudeza visual.
INTRODUCTION

An atypical case of relapsing Coats disease in adults is introduced herein. First described by George Coats in 1908, this is a unilateral idiopathic vasculopathy characterized by the presence of telangiectasias, massive exudation and exudative retinal detachment (RD). It appears more frequently in males and is typically unilateral. Its hereditary nature has not been established, nor an association with systemic diseases. It is usually diagnosed during the first decade of life (1,2). Usually, it is more aggressive when the onset takes place at an early age (3).

CASE REPORT

A 44-year-old male arrives in the emergency room reporting blurry vision in his right eye (RE). The patient was examined in our unit back in 1990, 14 years earlier, due to a first episode of blurry vision. Visual acuity (VA) was 9/10 in the RE and 20/20 in the LE. The exploration yielded no findings, except in the eye fundus (EF), which revealed exudation, microhemorrhages, discreet exudative RD in the peripapillary and inferior temporal region. He was diagnosed with Coats disease, in spite of the fact that it usually does not affect adult subjects, based on the appearance of the eye fundus, the angiography (FAG) and the absence of a personal history of unilateral exudative retinopathy. A general examination (blood tests, serology and chest X-rays) revealed no alterations. Argon laser photocoagulation was applied in three sessions. Exudates fully reabsorbed a year and a half later, preserving VA of 20/20, and leaving only the laser scar behind (fig. 1).

Fourteen years later, during the new episode of blurry vision, VA was 9/10 in his RE. The anterior pole was normal and IOP was 16 mmHg in BE. The RE’s EF showed a wide nasal exudative RD surrounded by hard exudates, microhemorrhages and macular star (fig. 2). The RE’s FAG revealed the characteristic premature hyperfluorescence of telangiectasias and microaneurisms (fig. 3), with late leakage (fig. 4). The patient was treated with laser photocoagulation in three sessions during a two-year period, gradually improving (fig. 5) until full resolution was achieved (fig. 6). The LE exploration was normal during both episodes. Currently, the patient is asymptomatic, with VA of 20/20.

DISCUSSION

Atypical manifestations of Coats disease may be appreciated in children with low VA, strabismus or leukocoria. In a series covering 150 cases, Shields observed that the mean age at diagnosis was 5 years; although ranging from 1 month to 63 years of age, adults are rarely diagnosed with this disease from the beginning (2). In both cases, it mainly involves vascularization. In adults, alterations take place in the equatorial and peripheral regions, hemorrhages, macroaneurism-related hemorrhages are more frequent and progression is slower than in children (3). Coats disease may influence the proli-
feration and migration of endothelial cells due to mutations in the genes responsible for the norrin protein. Different defects are likely to result in different phenotypes for the disease, with different degrees of severity and age at the onset (3,4).

In the present case, there was no previous history of RD, diabetes, exposure to radiation, episodes of intraocular inflammation, tapetoretinal degeneration or vascular occlusion. These diseases may have an exudative retinal response, similar to the clinical appearance of Coats disease, known as Coats-like syndrome (3). Gass classifies those subjects suffering from high blood pressure, polycythemia vera, coronariopathy and diabetes under a different category, even though he considers telangiectasias to be idiopathic (4).

Shields classified the disease according to the type and location of injuries (2). If only retinal telangiectasias are observed, it is a stage 1. If there is also exudation, it is stage 2 (2A, extrafoveal or 2B, foveal). Stage 3 is defined by the presence of exudative RD and stage 4 is linked to full RD and secondary glaucoma.

It is difficult to perform prospective and randomized studies on this disease and the different treatments available, since it is not frequent and its severity varies widely. Treatment should aim at obliterating telangiectasias in order to stop exudation and re-attach the RD if necessary. In the early stages, this may be achieved by means of laser photocoagulation.

Fig. 3: RE FAG: Hyperfluoresence of abnormal retinal vessels.

Fig. 4: RE FAG: At a late stage, intraretinal and subretinal fluid leakage and staining.

Fig. 5: RE EF: Gradual reabsorption of exudates after laser photocoagulation.

Fig. 6: RE EF: Re-attached retina, without exudate. Laser marks and retinal atrophy in the nasal sector can be observed.
gulation or cryotherapy. The most advanced cases also require RD surgery. Laser photocoagulation may be taken into consideration at the beginning if telangiectasias are located in the periphery and there are large exudation areas (1). If untreated, the disease’s natural progression may lead to complications such as full bullous RD, neovascular glaucoma and phthisis bulbi (5). If the exudation area is limited to a single quadrant or is located in the nasal sector, visual prognosis is more favorable.

In order to assess the treatment’s efficacy, it is advised to wait for at least 3 months, since exudation reabsorbs very slowly, which was the case of the present patient. According to clinical findings, treatment may be repeated (1). Exudation and/or subretinal fluid may lead to an irreversible fibrosis process that determines visual prognosis in cases involving the macula. Relapsing telangiectasias and exudation have been described in 7 percent of cases ten years after administering a satisfactory treatment (1,5), which may be multiple and applied during intervals of different duration, whereas follow-up should be implemented for life (in children, approximately every 6 months (5), due to the lack of complaints and the likelihood of developing amblyopia). At the clinical level, there is neither leukocoria nor strabismus in the case of adults and it tends to be very asymptomatic, nor does it lead to major complications, such as neovascular glaucoma (3).

REFERENCES