FLUORESCINE ANGIOGRAPHY WITH RETCAM IN INCONTINENTIA PIGMENTI: A CASE REPORT

ANGIOGRAFÍA FLUORESCEÍNICA CON RETCAM EN INCONTINENCIA PIGMENTI: COMUNICACIÓN DE UN CASO

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ABSTRACT

Case report: The ophthalmic examination and results of fluorescein angiography using Retcam II are described in a patient with Incontinentia Pigmenti (IP).
Discussion: Angiography fluorescein is extremely valuable in detecting vascular lesions that were invisible with ordinary ophthalmoscopy. Retcam II allows documentation of these lesions which is very useful for diagnosis, treatment and follow-up of this disease (Arch Soc Esp Oftalmol 2009; 84: 529-532).

Key words: Incontinentia Pigmenti, Bloch-Sulzberger syndrome, angiography, fluorescein, diagnostic techniques.

INTRODUCTION

Incontinentia pigmenti (IP) or Bloch-Sulzberger disease is a hereditary ailment transmitted via X-linked dominant inheritance, 80% of which are NEMO gene mutations (1). It affects almost exclusively women. In the first months of life, patients develop characteristic systemic alterations in the skin, teeth, hair, bones, central nervous system and eyes. Ocular involvement occurs in 35% of cases. Although a variety of ocular alterations can emerge (strabismus, cataracts, microphthalmos, iris hypoplasia, among others), retinal lesions involve the largest threat to eyesight and may go unnoticed in a routine ocular fundus exploration. Retinopathies are expressed as a variable severity vasculopathy.

RESUMEN

Caso Clínico: El examen oftalmológico y los resultados de la angiografía fluoresceínica empleando la Retcam II, son descritos en una paciente con Incontinencia Pigmenti (IP).
Discusión: La angiografía fluoresceínica es muy valiosa detectando lesiones vasculares que eran invisibles con oftalmoscopia normal. La Retcam II permite la documentación de dichas lesiones lo cual es muy útil para el diagnóstico, tratamiento y seguimiento de esta enfermedad.

Palabras clave: Incontinencia Pigmenti, Síndrome Bloch-Sulzberger, angiografía, fluoresceína, técnicas diagnósticas.
from an avascular peripheral area (similar to that observed in retinopathy of premature babies) to a fibrous proliferation with tractional retina detachment. There can also be anomalies in the pigmentary epithelium (2) and foveal hypoplasia (3).

**CLINICAL CASE**

A 4-month old female infant referred by her dermatologist with a diagnostic of IP to discard associated ocular pathology. The skin exhibited typical lesions for this disease, the so-called Blaschko lines (fig. 1). An ocular exploration in the practice revealed good visual behavior in both eyes (BE), normal anterior segment, normal intrinsic and extrinsic ocular motility. The ocular fundus exploration (OF) with indirect ophthalmoscopy was normal in the right eye (RE), but the left eye (LE) exhibited a minute hemorrhage in the nasal part (fig. 2). Due to the difficulty involved in the exploration due to continuous movements of the patient, a new OF exploration under narcosis and fluorescein angiography was scheduled due to suspected hidden vascular injuries. Said exploration comprised a new OF exploration with indirect ophthalmoscopy and Retcam II (Massie Labs.), which allows for digital archiving of images for subsequent detailed analysis. A paramacular hypopigmented injury was observed in the RE (fig. 3) and a discrete alteration in a vessel pathway in the extreme temporal periphery. The LE exhibited a linear hemorrhage in nasal periphery without evidence of other findings. A fluorescein angiography

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**Fig. 1.**

**Fig. 2.**

**Fig. 3.**

**Fig. 4.**
was made, with IV injection of sodium fluorescein (5 ml/kg of weight) which in this case amounted to 0.35 ml. Serial photographs were taken with the Retcam II which evidenced lesions that were unobserved in the previous ophthalmoscopy.

The RE exhibited point-shaped exudations in the extreme temporal periphery (fig. 4). In the LE we observed non-perfusion of the entire nasal periphery (fig. 5). The entire affected nasal area of the LE was treated with diode laser (1,200 impacts, 300 mW, 300 ms) (fig. 6).

**DISCUSSION**

Together with neurological pathologies, retinal vascular alterations are the main cause of vision loss in these patients (4). Retinal lesions can lead to tractional retina detachment with a pessimistic prognosis. In the instant case, the alterations are bilateral and asymmetrical, as is usually the case in IP (5). The RE exhibited minor alterations in the posterior pole as well as in the periphery. However, the LE exhibited an important nasal ischemia subsidiary to prophylactic treatment.

It is very important to verify OF frequently in the first year of life of these patients and consider fluorescein angiography if the slightest alteration is evidenced, because the lesions which can lead to retinal detachment could go unnoticed in routine ophthalmoscopy. Retcam II allows for the archiving of its images for detailed analysis, which is very useful for the diagnostic, treatment and follow-up of this disease.

**REFERENCES**