OPTIC NERVE DRUSEN AND DEEP VISUAL FIELDS DEFECTS

DRUSAS DEL NERVIO ÓPTICO Y DEFECTOS CAMPIMÉTRICOS SEVEROS

CALVO-GONZÁLEZ C1, SANTOS-BUESO E2, DÍAZ-VALLE D2, RECHE-FRUTOS J1, MORICHE-CARRETERO M3, BENÍTEZ-DEL-CASTILLO JM2, GARCÍA-SÁNCHEZ J2

ABSTRACT

Purpose: Optic nerve drusen needs to be included in the differential diagnosis of pseudopapilledema. As the identification of this entity by funduscopy may be difficult, ultrasonography has thus become the gold standard for its diagnosis. Severe optic nerve drusen has been correlated with a reduction of the nerve fiber layer measured by optic coherence tomography and with the presence of serious visual field defects. To demonstrate the relationship between extensive optic nerve drusen and visual field defects.

Method: A prospective observational study of the visual fields of a series of 5 patients with severe optic nerve drusen diagnosed by ultrasonography.

Results: Visual field defects of widely differing severity, from inferior nasal quadrant to severe hemivisual field defects, were described in each patient studied.

Conclusions: Visual field defects of diverse severity are common in patients with deep optic nerve drusen. For that reason ultrasonography and/or opti-

RESUMEN

Objetivo: Las drusas del nervio óptico deben ser incluidas en el diagnóstico diferencial de pseudopapiledema, ya que pueden resultar dificilmente identificables mediante funduscopy. Por ello se hace necesaria la utilización de la ecografía, como gold standard en su diagnóstico. Se ha correlacionado la severidad de las drusas papilares con la reducción del espesor de la capa de fibras nerviosas medidas por tomografía óptica de coherencia y con la existencia de defectos importantes en el campo visual. Demostrar la existencia de una alteración campimétrica asociada a la presencia de drusas del nervio óptico extensas.

Métodos: Estudio descriptivo prospectivo de campimetrías realizadas por una serie de 5 pacientes afectos de drusas del nervio óptico severas diagnosticadas mediante ultrasonografía.

Resultados: Se han observado defectos campimétricos destacables de diversa severidad en todos los pacientes estudiados, que oscilan entre escalón nasal y defectos de hemicampos severos.
INTRODUCTION

Optic nerve drusen (OND) constitute hyaline globule-like formation with a diameter of 5-1,000 microns, commonly situated in front of the lamina cribrosa. OND contain amino acids, ribonucleic acids and small amounts of iron and calcium, which makes them partially calcified structures. Even though their pathogenesis remains unknown, the most accepted theories suggest an alteration in the axoplasmic flow of ganglion cells. In addition, the small scleral channels seem to be more frequently associated to the presence of OND. Clinically, a prevalence of 3.4 for each 1,000 adults has been described. However, histological studies on cadavers point to higher prevalence rates. ONDs are usually bilateral and asymmetric and seem to favor the female gender. Frequently ONDs are associated to ocular vascular abnormalities such as presence of ciliary-retinal arteries, marked tortuosity, abnormal bifurcations, retinal-choroidal collaterals, hemorrhage, peripapillary neovascularization and occlusive vascular diseases, among others (1). ONDs can cause abnormal elevation of the papilla, making it necessary to establish differential diagnosis with papilledema.

Clinically, ONDs can reduce central visual acuity (infrequent) and produce campimetric defects, some of which simulate a glaucomatous pattern. This finding, together with a difficult interpretation of the optic disc due to the presence of drusen, increases the complexity of diagnosis and follow-up of patients with possible glaucomatous damage. Likewise, the severity of clinically visible OND has been correlated with reductions in the thickness of the nervous fiber layer measured by Optic Coherence Tomography (OCT) and with visual field defects (2). The most reliable OND diagnostic is provided by mode B echography, although they can also be detected by means of fluorescein angiography, CT scan, electrophysiological methods and funduscopy for visible drusen.

This article presents 5 case reports which evidence the existence of visual field defects which are permanent, deep and with glaucomatous pattern in patients with severe ONDs, as well as the usefulness of echography and high definition photography in differential diagnosis of raised papilla.

SUBJECTS, MATERIAL AND METHODS

A prospective study was made on five patients (10 eyes) diagnosed with OND. The inclusion criteria were: 1) OND diagnostic obtained by funduscopy and/or mode B echography, 2) severe visible drusen, and 3) high reliability obtained in perimetric tests. The exclusion criteria were: 1) ocular hypertension or previously diagnosed glaucoma, 2) evidence of other eye diseases, and 3) presence of macular pathology secondary to alterations related to drusen or other visual deficit causes.

Ecographically (ultrasound mode-B), ONDs appear as hyper-reflecting rounded structures with acoustic shadow in mid-range gains. High definition photographs were taken by non-midriatic retinograph (CR-DGi by Canon) and mode B echographies (Toshiba Power Vision 6000 Echograph). In all cases, computerized campimeties were taken utilizing the 24.2 program of the Humphrey FA II Perimeter (Carl Zeiss, Inc, San Leandro, CA, EEUU) until reliable campimeties were obtained with a proportion of false negatives and positives below 20%. Data for maximum corrected visual acuity (MAVC) in each eye, mean defect (in dB) and campimetric involvement pattern.
RESULTS

Ten eyes of 5 patients were studied, of which 4 are men. The mean age is of 57.6 (range, 42-78). The mean corrected visual acuity is of 0.78, with a notable MAVC ≥ 0.5 (logMAR) in all patients. All cases (10 eyes) presented severe OND, confirmed by the detection of a hypertense image in the NO head with mode B echography (figs. 1 and 2).

The high reliability perimetrías obtained with Humphrey 24.2 were analyzed, identifying an involvement of the visual field (VF) in 100% of cases. The mean defect ranges between –2.39 and –22.7 (dB), with a pondered mean value of –9.8. The found campimetric alterations are the following, listed in decreasing order of frequency: 40% of inferior arched scotoma (of which half presented nasal scotoma), 40% involvement of lower hemi-field (half with slight involvement of superior hemi-field), 10% of superior arch-shaped and nasal scotoma and 10% with isolated nasal scotoma (figs. 1 and 2).

The results obtained are shown in table I.

DISCUSSION

Superficial OND can be easily identified with usual diagnostic techniques. However, deep and buried ONDs can be more difficult to identify. This situation is described more frequently in children, although there is strong evidence suggesting that with growth there is a slow and progressive increase in the size and visibility of ONDs (1). The difficult identification of buried drusen makes it crucial to differentiate them from genuine papilledema, for which it is necessary to utilize supplementary diagnostic methods to confirm the presence of this type of drusen.

There are different tests to determine the presence of drusen in the optic nerve head (ON).

The most simple and economic technique is funduscopy, although it entails an important limitation in the identification of deep or buried deposits. CT scan is useful due to its ability to show the calcium component of drusen as identifiable shiny dots. Its high cost as well as the inability to make sufficiently fine sections of the ON head region, make

Fig. 1: B-mode echography and campimetry of the RE of patient #3 with deep drusen buried in the optic nerve head.

Fig. 2: Funduscopía, B-mode echography and serial campimetrías in OND.
Similarly, fluorescein angiography is useful. Self-fluorescence of OND prior to the injection of contrast is characteristic, as well as the presence of hyper-fluorescence in the late passage thereof (1). However, the test of choice for diagnosing OND is B-mode echography because it is a method with high sensitivity and greater diagnostic yield than pre-injection self-fluorescein detection or orbital CT scan (3).

From the clinical viewpoint, visual acuity reduction due to the presence of drusen is not frequent. On the contrary, visual field defects are described as being commonly associated to OND. Several studies have demonstrated greater frequency and severity of perimetric alterations in patients with visible OND as well as older age (1-5), with campimetric defects being less frequent in patients with buried OND (6). Even though the most dramatic VF losses in patients with drusen are usually related to associated vascular complications, drusen can also be responsible for the defect. In these cases, pupillary alterations of the afferent relative type are usually found.

The most commonly found alterations, described as of slow progression, are arch-shaped defects, mainly inferior, general constriction and growth of the blind spot. Visual field loss can correspond directly or not to the location of OND. Physiopathogenic mechanisms are varied: compromise of the axon transport in one eye with small scleral canal with gradual secondary wearing out of the ON fibers, direct compression of pre-laminar fibers by drusen and ischemia of the ON head (1).

In our series we have observed campimetric defects of variable depth showing patterns similar to those described in the literature. On the other hand, the most acute defects were identified in patients with bigger drusen as shown by the echo-graph images.

By means of OCT and laser scan polarimetry (GDx) a reduction of the nervous fiber layer has been described in OND patients, greater loss in relation to a higher number and greater visibility of the drusen. OCT seems to be a more sensitive and earlier indicator to detect the loss of nervous fibers as compared with perimetry and fiber layer photography (2,7). Authors recommend B-mode echography to confirm or discard the existence of OND as well as to perform campimetric and/or OCT follow-up to monitor the involvement of the nervous fiber layer in these patients.

REFERENCES