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Original Article

Chiasmal syndrome: Clinical characteristics in patients attending an ophthalmological center

Aline Astorga-Carballo; Juan Carlos Serna-Ojeda *; Mayra Fabiola Camargo-Suarez

Abstract

Purpose: To present the clinical characteristics of a group of patients with the diagnosis of chiasmal syndrome who attended a large ophthalmological institute.
Methods: Retrospective, observational clinical study with the review of medical records of patients with a diagnosis of chiasmal syndrome. The following variables were assessed: demographic characteristics, chief complaint upon presentation, best-corrected visual acuity (BCVA), presence or absence of diplopia, pupillary responses, optic nerve head morphology, etiology, and results from the ancillary tests including Ishihara test, Goldmann visual field (GVF) perimetry and neuroimaging.
Results: A total of 104 met the inclusion criteria, with a median age of 52 years (range 4–86 years). Fifty-four patients (51.9%) were referred to our institution with a diagnosis of a causative etiology for chiasmal syndrome, while in 50 (48.1%) the diagnosis was performed at our center. The most common presenting symptom was low visual acuity in 57 patients (54.8%), and the most common GVF defect was bitemporal hemianopsia in 39 patients (78 eyes, 39.8%). Pupillary abnormalities were present in 58 patients (55.7%), the optic nerve revealed pallor at any degree in 67 patients (64.4%) and the Ishihara test was affected in 65 patients (62.5%). The most common diagnosis was pituitary macroadenoma.
Conclusion: The ophthalmologist participates in the diagnosis and rehabilitation of patients with chiasmal syndrome. Low visual acuity is the most common symptom at presentation, and bitemporal hemianopia the most frequent GVF defect. Examination of the optic nerve head and pupillary responses, and ancillary tests including Ishihara test and neuroimaging are relevant for diagnosis.

Keywords: Chiasmal syndrome, Intracranial tumor, Perimetry, Ishihara test, Bitemporal hemianopsia

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Introduction

The chiasmal syndrome is a constellation of signs and symptoms that include changes in the visual field, decreased visual acuity and atrophy of the optic nerves, which are associated with lesions in the optic chiasm.[1–4] Other symptoms such as diplopia, alterations in chromatic sensitivity, changes in the appearance of the head of the optic nerve, headache and systemic manifestations secondary to variations in the pituitary hormones may also arise.[5]

The etiology varies from congenital, traumatic, iatrogenic causes to extrinsic or intrinsic lesions. The most common intrinsic lesion is the pituitary adenoma which may promote compression of the optic chiasm causing visual disturbances.[1,6–8] Therefore, a suspicion starting with the clinical

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Instituto de Oftalmologia ''Conde de Valenciana'', Mexico City, Mexico

* Corresponding author at: Instituto de Oftalmologia ''Conde de Valenciana'', Chimalpopoca 14, 06800 Mexico City, DF, Mexico.
e-mail address: juanc.sernao@gmail.com (J.C. Serna-Ojeda).

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history, a correct diagnosis, prompt management and rehabilitation is essential for these patients.

The ophthalmologist may be involved with the care of patients with chiasmal syndrome, performing the diagnosis at first, therefore making the reference for further neurological management and also in the visual rehabilitation and control. In this study, we present the clinical characteristics of a group of patients with the diagnosis of chiasmal syndrome who attended a large ophthalmological institute during a period of 6 years.

Material and methods

This is a retrospective, observational clinical study; we reviewed the medical records of all patients with a diagnosis of chiasmal syndrome at the Instituto de Oftalmologia ''Conde de Valenciana'' in Mexico City from 2009 to 2014. As our institution is an ophthalmological center, we diagnose patients and refer them for multidisciplinary management to other institutions, but also receive patients after treatment for visual rehabilitation.

Patients with suspected chiasmal syndrome underwent a complete medical history, an ophthalmologic examination including best-corrected visual acuity (BCVA), intraocular pressure by applanation tonometry, anterior segment biomicroscopy and fundoscopy. Other ancillary tests were performed including: Ishihara test for chromatic sensitivity, Goldmann visual field (GVF) perimetry with a standard III/2e stimulus but then individualized according to each patient, computed tomography (CT) scanning or magnetic resonance imaging (MRI), and laboratory tests including basal hormone levels and/or dynamic hormone measurements depending on the tumor suspected. If available, the histopathologic diagnosis was obtained from some of the referral centers. We included the patients that came back for follow-up and the diagnosis was completed.

We assessed the following variables: demographic characteristics, chief complaint upon presentation, BCVA, presence or absence of diplopia, pupillary responses, optic nerve head morphology, results from the available studies and etiology. Visual acuity was analyzed in the logMAR format but reported again in the Snellen chart format.

Using descriptive statistics, categorical variables were evaluated using percentages and numerical variables were assessed using measures of central tendency for non-parametric distribution.

The study adhered to the tenets of the Declaration of Helsinki and was approved by the Institutional Ethics Committee.

Results

One hundred and eighty-two patients with diagnosis of chiasmal syndrome were identified. A total of 104 met the inclusion criteria, from which 62 (59.6%) were female and 42 (40.4%) male. The median age of the patients was 52 years (range 4–86 years). Fifty-four (51.9%) patients were referred to our institution with a diagnosis of a causative etiology of chiasmal syndrome, while in 50 (48.1%) the diagnosis was performed at our center.

The BCVA at presentation was a median of 20/60 (range 20/20 to no light perception) in the eye with poorer vision, with 41 (39.4%) patients with a BCVA worse than 20/400.

The most common presenting symptoms were low visual acuity in 57 (54.8%) patients, changes in the peripheral visual field in 20 (19.2%), systemic symptoms secondary to hormonal imbalance in 18 (17.3%), headache in 10 (9.6%), diplopia in 1 patient (0.9%) and pupillary changes in 1 patient (0.9%). Nine (8.6%) patients were initially misdiagnosed and treated for glaucoma for a median time of 4 years (range 1–7 years), for changes in the appearance of the optic nerve head, until the visual field defect was recognized as suggestive of a chiasmal syndrome.

Regarding the GVF perimetry. each eye of the patients was evaluated separately (Figs. 1 and 2). Six patients didn't have a visual field registered, then 196 eyes of 98 patients were analyzed. The most frequent visual field defect was bitemporal hemianopsia in 39 patients (78 eyes, 39.8%), followed by temporal hemianopsia in 18 (9.2%) eyes, quadrantanopsia in 17 (8.6%) eyes, increased blind spot in 16 (8.2%) eyes, altitudinal defect in 9 (4.6%) eyes, nasal scotoma in 7 (3.5%) eyes, homonymous hemianopsia in 3 patients (6 eyes, 3.1%), cecocentral scotoma in 5 (2.5%) eyes, and constricted peripheral field in 3 (1.5%) eyes. Eight (4.1%) patients presented amaurosis in one eye and 29 (14.8%) eyes didn't present any GVF defect.

In 58 (55.7%) patients, pupillary abnormalities were reported, from which 54 (51.9%) patients had a relative afferent pupillary defect, and 4 (3.8%) had a fixed dilated pupil. The optic nerve revealed pallor at any degree (from 1+ to 4 +) in 67 (64.4%) patients. The Ishihara test was affected in 65 (62.5%) patients.

The neuroimaging studies were available at our institution for 34 (32.7%) patients with the confirmation of the presence of an intracranial tumor (Fig. 3), being the most common a pituitary macroadenoma in 17 (50%) patients, followed by an unspecified size adenoma in 6 (17.6%), a pituitary microadenoma in 5 (14.7%), an arachnoidocele in 4 (11.7%), craniopharyngioma in 1 patient (3%) and a pilocytic astrocytoma in 1 patient (3%). The rest of the patients had an imaging study in the referral neurologic center confirming an intracranial lesion, but it was not possible to obtain the specific information.

Discussion

In this study, we extensively analyzed the clinical characteristics of a group of patients with the diagnosis of chiasmal syndrome. We found relevant data, most of that remained in accordance with other studies, but with the particularity that those were patients who attended a large ophthalmological institute.

The optic chiasm is a flattened ring composed of a tuft of fibers located at the junction of the anterior wall of the third ventricle with its floor.[9] It contains approximately 2.4 million afferent nerve fibers that anteriorly reach both eyes and then continue along the optic tract. It measures approximately 8 mm in anterior to posterior axis, 15 mm wide and 4 mm high.[9,10] Most chiasmal syndromes can be classified as intrinsic (thinning of the chiasm) or extrinsic (adjacent structures causing compression).[6]

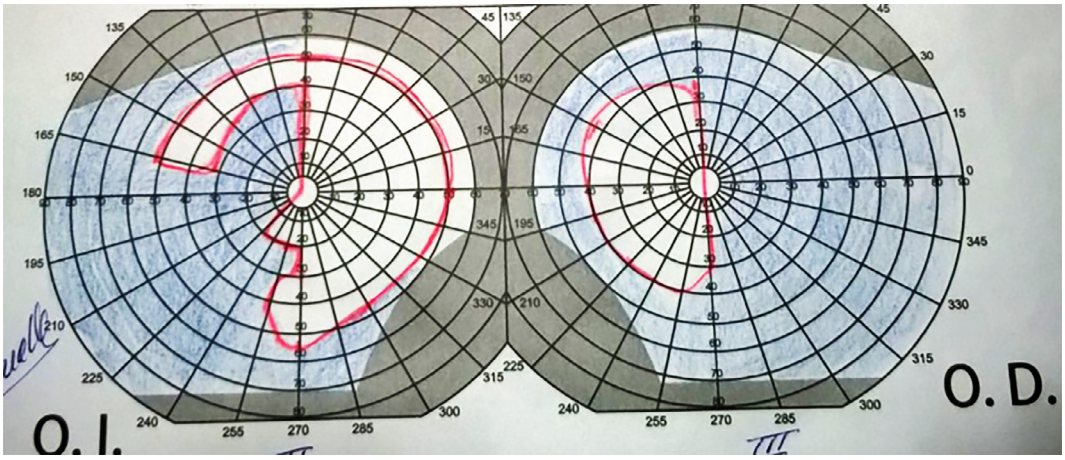

Fig. 1. Goldmann visual field perimetry showing a bitemporal hemianopsia corresponding to microadenoma.

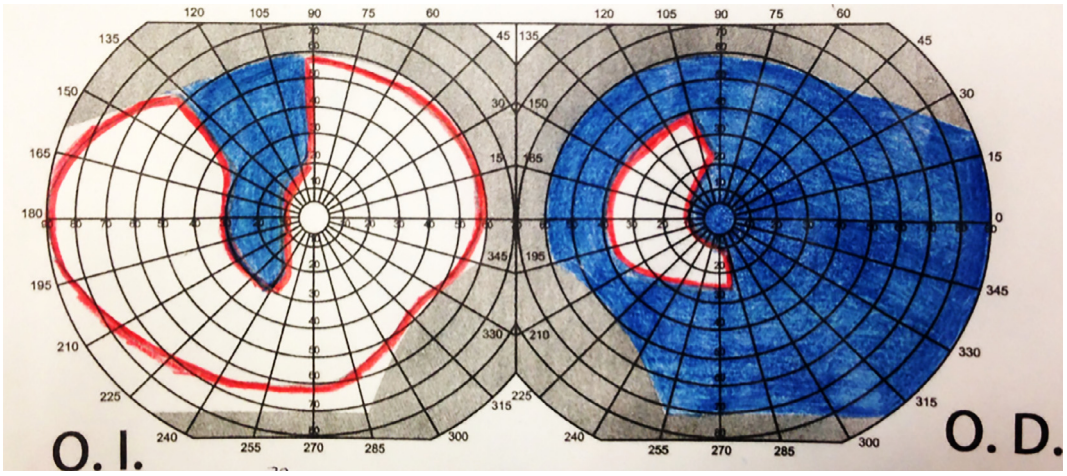

Fig. 2. Goldmann visual field perimetry showing right eye with a nasal island of vision secondary to temporal visual field loss with central involvement and left eye with increased central scotoma with tendency to form temporary hemianopia corresponding to macroadenoma.

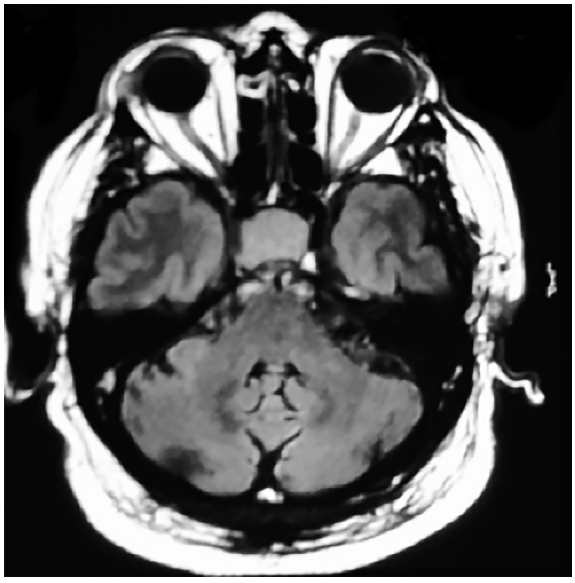

Fig. 3. Magnetic resonance imaging showing the sellar region with a tumor conditioning compression and displacement of neighboring structures corresponding to pituitary macroadenoma.

There are a variety of clinical manifestations in the chiasmal syndrome, with low vision as the initial presentation of many of the optic chiasm injuries.[1] It usually presents as a temporal visual field defect by the central compression of the optic chiasm.[2,3] In our series of patients, the decreased vision as the most common manifestation may be related to the advanced disease at presentation, because even when the median BCVA was 20/60, we also mention that almost 40% of patients had worse than 20/400 in the eye with poorer vision. Another frequent complaint is headache which is usually referred as retro orbital, which has been attributed to some causes including mechanical compression of the mass, inflammation of the structures adjacent to the tumor, and sometimes hormonal dysregulation. In some cases, patients may consult for diplopia due to the injury of the oculomotor nerves.[5]

Ogra and coworkers reported in 2014 a group of patients with pituitary adenomas, with visual loss as the most common reason for presentation (39%), followed by endocrine abnormalities (21%) and headache (15%).[8] Another study in patients with a variety of intracranial tumors in Bangladesh, reported the following ophthalmic manifestations: visual blur (91.1%), visual field defect (71.4%), optic disc changes

(50.0%), pupillary light reaction defect (48.2%) and color vision defect (46.4%).[1] Masaya-anon in a study from a neurological institute in Thailand in patients with intracranial tumors, reported as common neuro-ophthalmological findings, blurred vision (88.6%), visual field defects (80.5%), abnormal optic discs (69.7%) and relatively afferent pupillary defect (43.6%).[11] In contrast to our results whose main finding was low visual acuity in 54.8% of the patients, 64.4% with changes in the optic nerve, relative afferent pupillary defect in 51.9% and an altered Ishihara test in 62.5%.

In some cases of compressive neuropathy with chiasmal tumors, patients have passed for an equivocal diagnosis of open-angle glaucoma. The compressive optic neuropathy is associated with pallor of the optic nerve and visual field defects that obey the vertical meridian.[12,13] Compressive optic neuropathy is associated with significantly thinner nasal and temporal sectors compared with open-angle glaucoma discs.[14] In this study, we reported 9 patients who were initially treated by open-angle glaucoma until the visual field defect was recognized as suggestive of chiasmal syndrome.

The unique configuration of the optic chiasm, causes that a lesion typically produces changes in visual function, particularly visual field defects that are in some cases diagnostic[15]; the pattern of visual field loss reflects the location. Typically, the pituitary adenoma compresses the chiasm from its caudal aspect, causing a bitemporal hemianopsia that starts affecting the lower fibers and then the upper fibers. Because of that, the bitemporal hemianopsia may be reported as frequent as 41%, which can be in higher proportion in the groups including only pituitary tumors[8], compared with other groups reporting an incomplete involvement of the temporal hemifields in both eyes as the most frequent distribution of scotoma pattern related to a variety of chiasmal lesions in only 22% of the cases.[7] Our study found as the most frequent visual field defect the bitemporal hemianopsia in 39.8% of the patients with GVF perimetry, being consistent with the pituitary adenoma as the most common etiology. Other causes like craniopharyngiomas, produces a bitemporal hemianopia that initiate in the lower portion of the visual field.[16]

Several studies have evaluated the etiology of the patients with chiasmal syndrome. The retrospective series by Wadud et al. in 2014 with histopathology confirmation in all the cases, reported that the most common type of intracranial tumor was pituitary adenoma (58.0%), followed by craniopharyngioma (20.5%), and posterior fossa tumor (12.5%).[1] Schiefer and coworkers reported in the same way that the majority (65%) of chiasmal lesions were caused by pituitary adenomas, followed by craniopharyngiomas (12%), astrocytomas (9%), and meningiomas (8%).[7] The previous studies coincide with the findings of our study with the pituitary adenoma as the most common etiology in chiasmal syndrome, but in contrast with the report of a neurological institute of patients with neuro-ophthalmic manifestations with the most common intracranial tumor being meningioma (45%), and the pituitary adenoma just behind in 32.9% of cases.[11]

Mejico et al. conclude that an early age of onset, unilateral optic pallor, afferent pupillary defect, absolute or complete visual field defect, visual field defect more severe inferior than superior or a combination of these findings are suggestive of a different etiology of pituitary adenoma.[15]

Ogra reported that the majority of patients with pituitary adenoma have a good visual acuity despite having visual field defects, making the assessment of the visual field essential to rule out chiasmal compression.[8]

Once the diagnosis is made, the challenge is to determine what type of injury is responsible and the type of treatment that should be provided, which often requires complementary imaging studies.[5] The progression of the injury can cause compression of adjacent structures, including the optic nerves and cavernous sinus, resulting in further vision loss, oculomotor nerve deficits and hypopituitarism.[6] Perimetry is the most effective follow-up study for the detection of visual impairment.[17]

The main limitations of our study are the retrospective design and the lack of histopathology in most of the cases as most of the patients were treated in another institution.

This study adds to the body of literature of patients with chiasmal syndrome, highlighting the importance of the ophthalmologist in diagnosis and rehabilitation. Low visual acuity is the most common symptom at presentation, and bitemporal hemianopia the most frequent GVF defect. Examination of the optic nerve head and pupillary responses, and ancillary tests including Ishihara test and neuroimaging are relevant for diagnosis.

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Conflict of interest

There is no conflict of interest to disclose.

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