PUZZLING VISUAL FIELD LOSS IN PATIENTS WITH PRIMARY EMPTY SELLA

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Abstract

Several patterns of visual field loss including bitemporal or binasal defects have been sporadically reported in empty sella syndrome (ESS). The authors present automated and Goldmann perimetry findings in three female patients (age range 54-64 years) with ESS, normal intraocular pressure, good visual acuity, and non-glaucomatous cupping of the optic disc. Irregular bilateral peripheral constriction with normal appearance of the optic disc were found in the first patient. The second patient showed a moderately pale disc with an inferonasal defect in her right eye, and a pale disc with only a central and partially temporal field remaining in her left eye. The third patient had irregular constriction of the visual field and a pale disc in her right eye, and a normal field in her left eye; both optic discs were slightly elevated and increased intracranial pressure was recorded. No noteworthy progression of perimetric defects was found after a six-year follow-up. Understanding the cause of visual field defects similar to those found in these patients may be challenging, and should prompt adequate neuro-imaging focusing on possible ESS.

Introduction

While visual field disturbances are commonly associated with secondary empty sella following surgery or radiation therapy, patients with CT or MRI evidence of primary empty sella syndrome (ESS) – resulting from deficient diaphragma sellae with extension of the subarachnoid space into the sella and subsequent flattening of the pituitary gland – may have normal visual fields or the following types of visual field defect: bitemporal or binasal hemianopia or quadrantanopia, generalized field constriction, central scotoma and enlarged blind spot[1-4]. To the best of our knowledge, long-term perimetric follow-up, automated perimetry data and asymmetric perimetric findings are not often reported in patients with primary ESS. Therefore, we found it interesting to describe the clinical features of three of our patients.

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Patients and methods

Between 1990 and 1991, MR imaging showed primary ESS in three female patients. In 1992 all these patients underwent detailed ophthalmological examination with special attention being paid to disc appearance and manual kinetic (Goldmann) and automated (Humphrey central threshold program) perimetry. Endocrinological and neurological assessment, including visual evoked potentials (VEPs) and standardized A-scan measurement of the retrobulbar optic nerve, was performed. In 1998 the patients were completely re-evaluated, at which time Goldmann and automated perimetry were performed using similar strategies and programs.

Patient No. 1

RR, a 61-year-old female suffering from arterial hypertension since the age of 40; using MRI, a diagnosis of enlarged ESS was made in 1991; the endocrinological assessment showed elevated blood prolactin. In 1992, corrected (sph +1.50) visual acuity was 20/20 in both eyes; IOP was normal bilaterally (16 mmHg), as were optic disc (C/D 0.3), retrobulbar optic nerve thickness and transient and flash VEPs. Goldmann perimetry showed moderate to severe irregular bilateral peripheral contraction of the visual field. Automated perimetry showed bilaterally good sensitivity within the central 15-20° only (Humphrey 24-1 and 24-2 program). Six years later, in 1998, no noteworthy changes were seen on ophthalmological and perimetric examination.

Patient No. 2

ZA, a 64-year-old female who recently complained of headache and slow left visual loss in whom a diagnosis of partially empty sella syndrome was made by MRI in 1990. There were no endocrine disturbances. In 1992, corrected (sph + 2) visual acuity was 20/20 in the right eye and 20/25 in the left; the patient had a moderately pale right optic disc (C/D 0.4), and a pale left optic disc (C/D 0.5); IOP (14 mmHg) and retrobulbar optic nerve thickness were normal; there was reduced amplitude of transient VEPs in the left eye only. Goldmann and automated (Humphrey 30-2 and 24-2 program) perimetry: there was a quadrantic inferonasal defect in the right eye; there was only a central and partially temporal field remaining in the left eye, with depressed sensitivity. In 1998, no progression of perimetric defects could be found on central automated testing (Fig. 1) and both Goldmann perimetry and ophthalmological examination had remained unchanged.

Patient No. 3

BT, a 54-year-old female. In 1991, the patient suffered from severe headaches with bilateral true papilledema and an MRI diagnosis of ESS was made; she had elevated blood prolactin. In 1992, corrected (sph +4.50) visual acuity was 20/25 in the right eye and 20/20 in the left; the patient had an elevated, pale (OD>OS) optic disc with blurred margins and minimal venous congestion; the diameter of the retrobulbar optic nerve was enlarged with increased subarachnoidal fluid; IOP was normal (17 mmHg); and there was reduced amplitude and increased latency of transient VEPs, especially in the right eye. The ophthalmoscopic evidence of increased intracranial pressure

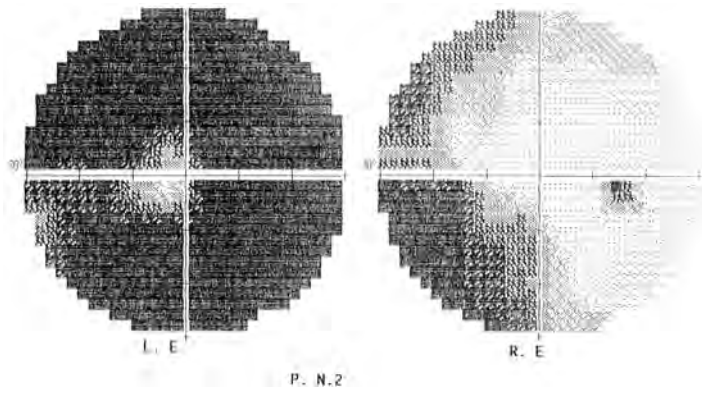

Fig. 1. Patient No. 2. *Right eye:* quadratic inferonasal defect; *left eye:* severe field contraction with depressed sensitivity (Humphrey Field Analyzer Program 24-2).

(ICP) was supported by 24-hour monitoring of the ICP (around 20 mmHg). Goldmann and automated (Humphrey 24-1 and 30-1 program) perimetry showed irregular constriction and decreased central sensitivity in the right eye, while the left eye was within normal limits. In 1998, there were no substantial changes in perimetric or ophthalmological findings, but there was a lack of echographic evidence of increased ICP.

Conclusions

Generalized visual field constriction was found in both eyes of patient No. 1 and in only one eye of patient Nos. 2 and 3. It is interesting to note that unilateral field constrictions have been found to be associated with diffuse central sensitivity depression on automated perimetry.

The co-existence of gross asymmetric field loss in patient Nos. 2 and 3, in whom the better eyes had a quadratic defect and just not an abnormality, respectively, is very rare, and probably due to asymmetric stretching and/or compromised blood supply at the chiasmal level, due to the compressive effect of the cerebrospinal fluid[3,5].

Actually in long-standing papilledema due to idiopathic intracranial hypertension, severe bilateral, usually peripheral, axonal loss has been documented[6]. Increased ICP and ESS, found in patient No. 3, are known to be associated[2], and this may occur more frequently than previously reported[7,8]. Unfortunately, ICP was not measured in patient Nos. 1 and 2, but they had normal retrobulbar optic nerves on echographic examination.

Goldmann perimetry still appears to be a good tool for the diagnosis and monitoring of patients with ESS, although automated perimetry provides much better information on the central 30° sensitivity.

The substantially stable visual field defects, lack of history of abrupt field loss, and non-glaucomatous cupping of the optic discs in our patients with ESS, militates against other pathologies such as anterior ischemic optic neuropathy and normal-tension glaucoma[9]. On the other hand, interpretation of visual field defects similar to those found

in our patients is challenging and should prompt adequate neuro-imaging, focusing on possible ESS and appropriate investigation in order to detect abnormal intracranial pressure.

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