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CASE REPORT

Glaucomatous-Like Cupping Associated with Slow-Growing Supra-Sellar Intracranial Lesions

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ABSTRACT *Purpose*: To demonstrate that slow-growing intracranial suprasellar mass lesions can be associated with optic disc cupping, clinically indistinguishable from glaucomatous optic neuropathy. *Cases*: At a neuro-ophthalmology tertiary clinic, a retrospective chart review identified four patients of presumed normal tension glaucoma and supra-sellar tumors. In all four patients with normal intraocular pressure, the appearance of the optic disc resembled typical glaucomatous optic nerve damage with disc cupping and parapapillary atrophy. *Conclusion*: In few patients, typical glaucomatous abnormalities of the optic nerve head may occur in association with slow-growing suprasellar tumors.

KEYWORDS optic disc cupping; glaucoma; Supra-sellar tumor; intracranial mass lesion; optic nerve compression; optic disc pallor.

INTRODUCTION

Glaucomatous optic neuropathy is characterized by enlargement of the optic cup due to a loss of neuroretinal rim, deepening of the optic cup due to a backward bowing of the lamina cribrosa, parapapillary atrophy, localized and diffuse loss of the retinal nerve fiber layer, and flame-shaped optic disc hemorrhages.[1] In contrast, eyes with non-glaucomatous optic nerve damage due to various conditions such as central retinal artery occlusion and intracranial tumors do not show marked enlargement or deepening of the optic cup, nor a change in the shape of the neuroretinal rim, but rather manifest as pallor of the optic disc rim. In contrast, patients after an arteritic anterior ischemic optic neuropathy, a non-glaucomatous cause for optic nerve damage, may show an enlargement and deepening of the optic cup.[2,3] Consequently, diffuse or focal loss of neuroretinal rim and an increase in parapapillary atrophy are considered almost pathognomonic for chronic glaucoma.

There are a few reports, however, in which glaucomatous-like excavation (cupping) of the optic nerve head was reported in association with compressive lesions of the intracranial segment of the optic nerve, with preservation of the color of the remaining neuroretinal rim and typical glaucoma-like visual field

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defects. Bianchi-Marzoli et al.[4] analyzed in a masked fashion the magnified stereophotographs of the optic discs of 51 eyes of 29 patients with intracranial lesions producing compressive optic neuropathy. The median ratio of cup/disc area was 0.37 and for the control eyes: 0.1, demonstrating statistical significance. They concluded that an inter-eye asymmetry in patients with unilateral optic nerve compression is convincing evidence that the enlarged cup is an acquired feature. Ahmed et al.[5] presented a series of 62 patients with normal tension glaucoma (NTG) in whom four were found to harbor a clinically relevant intracranial compressive lesion involving the anterior visual pathway. In contrast, Greenfield et al.[6] did not identify any intracranial mass lesions in a series of 23 NTG patients who were imaged. Younger age, lower levels of visual acuity, vertically aligned visual field defects, and neuroretinal rim pallor seem to increase the likelihood of a mass lesion. Lee[7] suggested that a mismatch between the degree of optic disc cupping and the degree of visual field loss, the presence of an afferent pupillary defect and rapid progression of visual loss are some "red flags" for possible nonglaucomatous optic atrophy.

It was the purpose of the present study to present four patients manifesting bilateral, yet asymmetric, optic disc cupping (Figs. 1–4), which was perceived to be clinically indistinguishable from glaucomatous optic disc cupping typically found in patients with advanced glaucomatous optic neuropathy.

CASE REPORTS

Patient 1

A 65-year-old female presented with the clinical diagnosis of NTG diagnosed four years earlier. Diagnosis was based on typical optic disc cupping, with a cup-to-disc (C/D) diameter ratio (vertical/horizontal) of 0.6/0.6 in the right eye and 0.9+/0.9 in the left eye (Fig. 1A). While the visual field of the right eye was within normal limits, the visual field of the left eye demonstrated a typical glaucomatous visual field defect with a marked inferior nasal step occupying most of the inferonasal quadrant (Fig. 1B). Highest documented intraocular pressure measurements were 18 mmHg in the right eye and 16 mmHg in the left eye. Visual acuity (VA) was 0.7 OD and 0.5 OS. There was a relative afferent pupillary defect in the left eye. Magnetic resonance imaging demonstrated a left anterior clinoid meningioma involving the left optic canal (Fig. 5A).

Patient 2

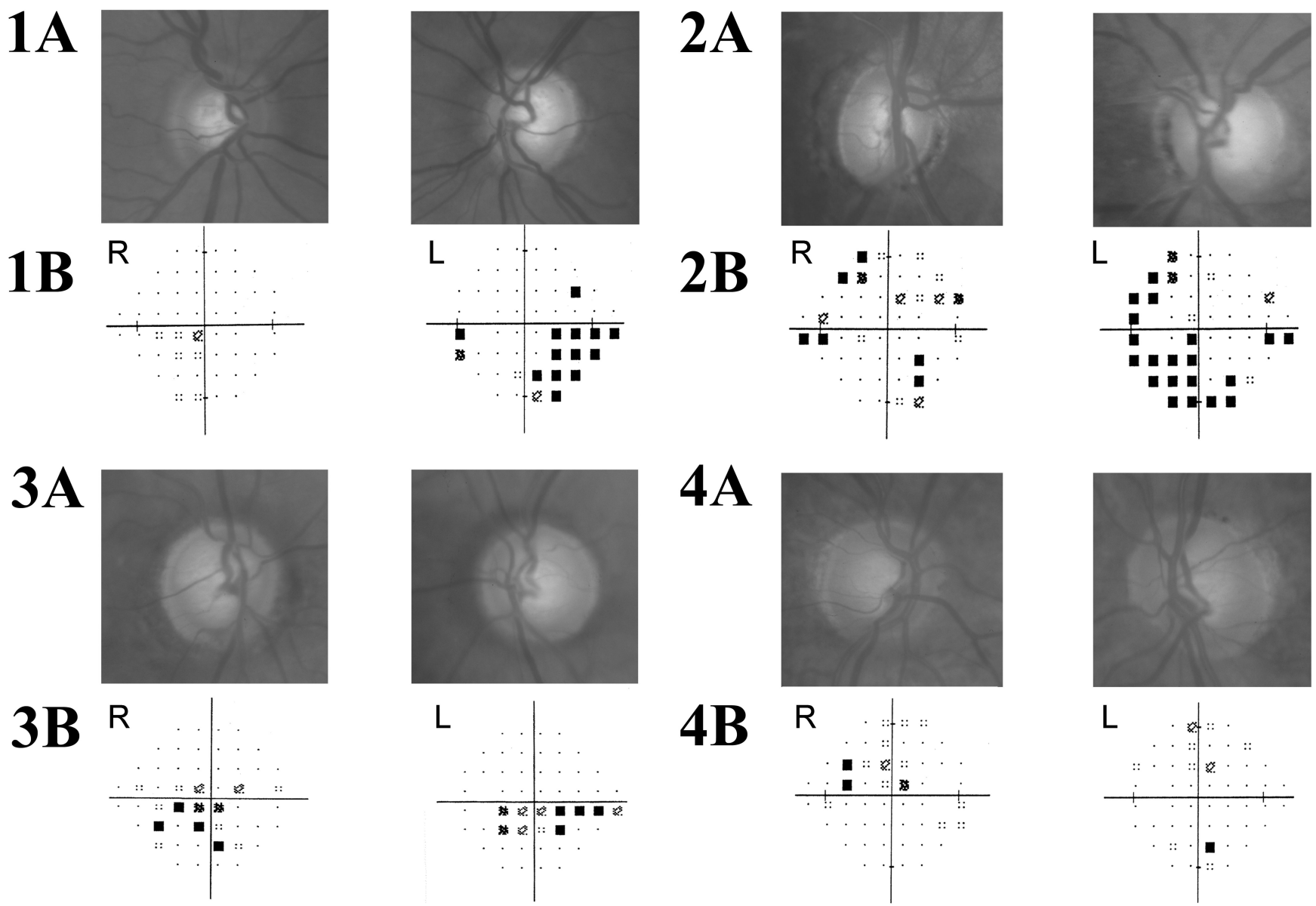
A 71-year-old female with a history of chronic schizophrenia was hospitalized due to somnolence and headaches accompanied by reduced visual acuity. Neuroimaging demonstrated a large tuberculum sella meningioma compressing both optic nerves (Fig. 5B). Preoperative visual acuity was 0.2 in both eyes with a relative afferent pupillary defect for the left eye. Highest recorded intraocular pressure measurements were 16 mmHg for both eyes. Fundus examination revealed optic disc cupping as typically found in eyes with glaucoma, more pronounced in the left eye than in the right eye. The optic discs demonstrated a C/D ratio of 0.9+/0.8 in the right eye and 0.9/0.7 in the left eye (Fig. 2A). Visual fields demonstrated non-specific scattered changes, a bilateral inferior nasal step, and a biarcuate visual field defect for the left eye mainly localized to the temporal hemifield, with an inferior vertical cutoff (Fig. 2B).

Patient 3

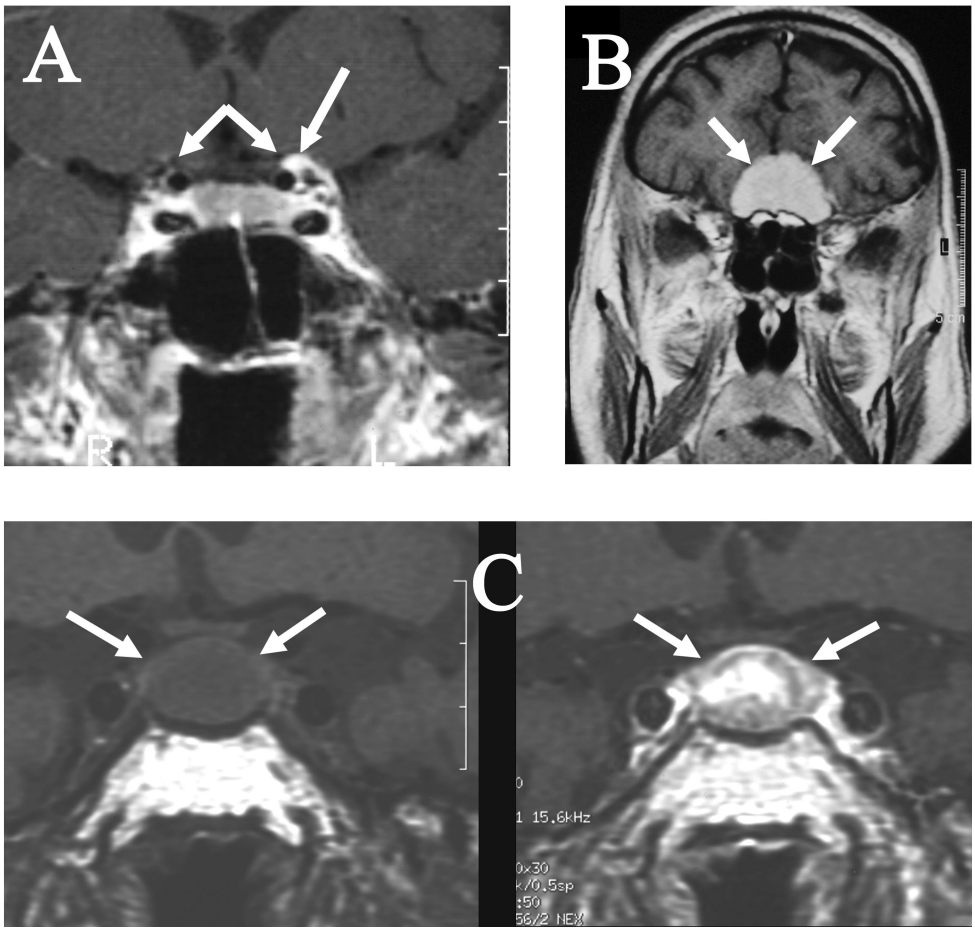
A 74-year-old male was diagnosed several years ago with NTG. Diagnosis was based on typical optic disc cupping (C/D ratio of 0.9/0.8 in the right eye and 0.8/0.7 in the left eye (Fig. 3A)) and associated visual field defects, which included an inferior arcuate-like defect approaching the horizontal raphe in the right eye and an inferior paracentral scotoma in the left eye (Fig. 3B). Additionally, the right eye showed an inferior optic disc hemorrhage at the 6:30 clock-hour location. Highest documented intraocular pressure readings in either eye never exceeded 20 mmHg. Visual acuity was 0.8 in both eyes. Following an episode of syncope and headaches, neurological evaluation revealed a 3.75 × 2.0 × 2.5 cm olfactory groove meningioma (Fig. 5B) that was consequently resected.

Patient 4

An 87-year-old male underwent neuroimaging as part of a work-up for transient episodes of blurred vision. A large, 2.25 × 1.25 × 1.25 cm, hypophyseal adenoma, with suprasellar extension was found (Fig. 5C). On neuro-ophthalmological consultation, visual acuity was 0.9 OD and 0.8 OS. On three consecutive examinations, intraocular pressure did not exceed 16 mmHg in either eye. There was no relative afferent pupillary defect. The visual field of the right eye suggested an



FIGURES 1–4 A. Disc photographs of both eyes of each patient. B. Visual field pattern deviation plots of both eyes of each patient (standard automated perimetry, full-threshold algorithm).



FIGURES 5 **A. MRI scan of patient 1. T1-weighted images (coronal section with gadolinium) revealing a small meningioma (1 × 1 × 1.5 cm) extending off the left anterior clinoid, encroaching on the lateral aspect of the left optic nerve before it joins the optic chiasma. Single arrow: tumor; double arrow pointing at both optic nerves. B. MRI scan of patient 3. demonstrates a coronal section (with gadolinium), revealing a large olfactory groove meningioma (3.75 × 2.0 × 2.5 cm). C. MRI scan of patient 4. demonstrates a coronal T1-weighted images (7 left: without gadolinium; 7 right: with gadolinium), revealing a sellar mass (2.25 × 1.25 × 1.25 cm), enhancing with gadolinium), and touching the inferior left paracentral aspect of the optic chiasma.**

early superior arcuate defect (Fig. 4B). The optic discs showed a glaucomatous appearance (C/D ratio of 0.8/0.8 in the right eye and 0.8/0.6 in the left eye) with an almost total loss of the infero-temporal neuroretinal rim in the right eye (Fig. 4A).

DISCUSSION

NTG is a diagnosis per exclusion. While typical glaucomatous cupping combined with corresponding visual field defects and normal intraocular pressure leaves little doubt regarding the diagnosis, it may be advised to also consider the possibility of an intracranial compressive lesion as a rare cause. A thorough clinical examination may point to neurological deficits in the majority of patients, hence it is debatable whether neuroimaging is warranted for most patients with NTG, or alternatively, should be reserved only for those patients presenting with neurological signs or an atypical presentation regarding age, progression or asymmetry as has been shown and discussed by Greenfield,[6] Ahmed,[5] Girkin,[8] and others. The four patients presented in this study illustrate the value of neuro-ophthalmic assessment in patients of NTG presenting atypical features, e.g., progressive visual loss, presence of relative afferent pupillary defect, vertically aligned visual fields, and neuroretinal rim pallor.

Two of these patients were diagnosed and treated for NTG based on optic disc appearance and visual field damage for some years prior to the diagnosis of an intracranial compressive lesion. Interestingly, the patients showed not only disc cupping but additionally an optic disc hemorrhage and abnormally large parapapillary atrophy (Figs. 1–4), as has also been described for NTG.

In conclusion, the clinical features of the patients demonstrated in this report suggest that space

occupying lesions in the parasellar and suprasellar region and in the region of the optic canal may be associated with contour abnormalities of the optic disc ophthalmoscopically indistinguishable from glaucomatous optic neuropathy. From a pathophysiologic point of view, one may raise the question of whether a blockage of the circulation of the cerebrospinal fluid may play a role.[9]

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