
ABNORMAL CUPPING OF THE OPTIC DISC: CLINICAL SCREENING BEFORE PERFORMING A NEUROIMAGING EXAMINATION

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ABSTRACT

Imaging studies in typical glaucomatous cupping are usually needless. This article reviews the various clinical presentations for considering imaging in patients presenting with cupped disc.

RÉSUMÉ

La réalisation d'une imagerie du nerf optique n'est généralement pas nécessaire dans la mise au point d'un patient présentant une excavation glaucomateuse typique du nerf optique.

Le papier décrit les différentes situations cliniques où la réalisation d'une résonance magnétique nucléaire du nerf optique doit être considérée chez des patients avec une excavation papillaire.

KEY WORDS

cupping, optic nerve, magnetic resonance imaging.

MOTS-CLÉS

excavation, nerf optique, résonance magnétique nucléaire.

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received: 11.01.06

accepted: 31.03.06

INTRODUCTION

The diagnosis of typical glaucoma related optic neuropathy can normally be made on clinical grounds alone (e.g., older age, splinter optic nerve hemorrhage, positive family history of glaucoma, level of intraocular pressure (IOP), nerve fiber bundle visual field defects). In addition, glaucoma produces a specific and differentiating form of optic atrophy (i.e. cupping). Although most patients with glaucoma have an elevated IOP corrected or not with the central corneal thickness, some have a true "normal-tension" glaucoma which renders the differential diagnosis difficult with a non glaucomatous cupping. Currently the indications for performing neuroimaging in patients with cupping of the optic nerve mimicking glaucomatous optic neuropathy are not uniformly accepted. As reported by Greenfield in 1998 (2), radioimaging examinations are expensive and rarely cover a previously unknown and asymptomatic intracranial tumor or condition that could explain the morphological aspects of the optic disc and/or the defects of the visual field. Then a more selective approach based on a careful medical history and examination is very important. This article reviews the different clinical presentations of suspicious glaucomatous optic atrophy for considering neuroimaging.

WHO NEEDS NEUROIMAGING?

LOSS OF CENTRAL VISUAL ACUITY OR CENTRAL VISUAL FIELD.

Although loss of visual acuity may occur as a late finding in glaucoma, it is a "red flag" for non-glaucomatous etiologies. Likewise, a central scotoma is an unusual visual field defect for early glaucoma and typically occurs only after marked loss of the peripheral field in end-stage disease.

One condition that could produce a central scotoma and central acuity loss is autosomal dominant optic atrophy (6). These patients may present with excavation of the optic nerve that may mimic glaucomatous cupping. The earlier age of onset (often in early adulthood or childhood), preferential loss of central vision, cen-

tral scotoma with sparing of peripheral visual field, optic disc pallor, and positive family history of optic atrophy are the differentiating features.

ACUTE ONSET OR RAPIDLY PROGRESSIVE VISUAL LOSS

Primary open-angle glaucoma is a slowly progressive and chronic disorder. An acute or subacute course of visual loss is atypical for glaucoma and should prompt evaluation for other optic neuropathies. One cause for an acute onset of visual loss followed by glaucomatous-like cupping is giant cell arteritis (3,4,5). Optic disc cupping that might mimic glaucomatous cupping can occur after arteritic anterior ischemic optic neuropathy. The pallor of the remaining neuroretinal rim and lack of peripapillary atrophy may be distinguishing features in favor of giant cell arteritis (3). Likewise, acute "shock-induced" hypotensive ischemic optic neuropathy may produce a non-progressive optic disc cupping that is similar to the cupping seen with glaucoma.

MARKEDLY ASYMMETRIC OR STRICTLY UNILATERAL OPTIC NEUROPATHY

Although some secondary glaucomas (e.g., uveitic, post-traumatic, Fuchs' heterochromic iridocyclitis) are unilateral, primary open-angle glaucoma is typically bilateral. It is often asymmetric, and might even be unilateral (especially in secondary glaucoma). Strictly unilateral disease, a large relative afferent pupillary defect, or markedly asymmetric disease is a "red flag" for a non-glaucomatous etiology (that may or may not be superimposed on true glaucoma).

HEMIANOPIC VISUAL FIELD LOSS

The nerve fibers originating in the retina follow a specific topographic arrangement, and glaucoma produces nerve fiber bundle defects that respect the horizontal meridian and insert into the blind spot. The typical glaucomatous field defects include generalized depression, concentric constriction, focal depression/constriction, barring of the blind spot, Bjerrum scotoma, paracentral scotoma, arcuate scotoma, nasal step, temporal wedge, superior or inferior

altitudinal loss, and central and temporal islands. The amount and location of the visual field loss should closely parallel the degree and location of the optic disc cupping.

Lesions that affect the optic chiasm or retrochiasmatal pathways produce hemianopic visual field loss that is atypical in glaucoma. Any degree of hemianopic field loss (i.e., bitemporal hemianopsia or homonymous hemianopsia) which respect the vertical meridian is a specific sign for a compressive lesion and tumors (e.g., pituitary adenoma, meningioma) (1,7). Significant mismatch between the degree of cupping and the severity of visual field loss arise suspicion for non-glaucomatous optic atrophy.

NEURORETINAL RIM PALLOR

The key differentiating feature of non-glaucomatous cupping is pallor of the remaining neuroretinal rim. Glaucomatous cupping (i.e., focal notching, sloping to the temporal rim, barring of the circumlinear vessels) produces loss of axons and secondary pallor within the cup. Optic atrophy beyond this cupped area (i.e., pallor of the remaining neuroretinal rim) is suggestive of non-glaucomatous optic atrophy (4).

CONCLUSION

In summary, neuroimaging is considered in patients with glaucomatous cupping who have atypical features including loss of central acuity, rapid or acute loss of vision, central or hemianopic field loss, mismatch between the degree of cupping and the degree of visual loss, an unexplained unilateral optic neuropathy, or pallor of the remaining neuroretinal rim. Neuroimaging in patients with typical glaucomatous cupping and typical glaucomatous visual field loss is not generally performed even if the intraocular pressure is normal.

This paper has been presented during the meeting of the Belgian Glaucoma Society on 23.11.2005

REFERENCES

- (1) CESTARI D.M., RIZZO J.F. III – The Neuroophthalmic Manifestations and Treatment Options of Unruptured Intracranial Aneurysms. *International Ophthalmology Clinics*. 2004; 44:169-187.
- (2) GREENFIELD D.S., SIATKOWSKI M., GLASER J.S., SCHATZ N.J., PARRISH R.K. – The cupped disc. Who needs neuroimaging? *Ophthalmology* 1998;105:1866-1874.
- (3) HAYREH S.S. – Optic disc morphology after arteritic anterior ischemic optic neuropathy. *Ophthalmology*. 2001; 108; 1586-1594.
- (4) TROBE J.D., GLASER J.S., CASSADY J.C., HERSCHLER J., ANDERSON D.R. – Nonglaucomatous excavation of the optic disc. *Arch Ophthalmol*. 1980; 98:1046-1050.
- (5) TROBE J.D., GLASER J.S., CASSADY J.C. – Optic atrophy. Differential diagnosis by fundus observation alone. *Arch Ophthalmol*. 1980; 98:1040-1045.
- (6) VOTRUBA M., THISELTON D., BHATTACHARYA S.S. – Optic disc morphology of patients with OPA1 autosomal dominant optic atrophy. *Br J Ophthalmol* 2003;87:48-53.
- (7) WATTS P., NEWSOM R., MCALLISTER J. – Sphenoidal ridge meningioma masquerade: glaucoma with a sphenoidal ridge meningioma. *Eye*. 1996;10:629-634

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