NONGLAUCOMATOUS OPTIC DISK ATROPHY AND EXCAVATION IN THE ELDERLY

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SUMMARY

The causes of nonglaucomatous optic disk atrophy and excavation are enumerated in people 65 years or older: congenital anomalies, myopia, ischemic disorders, transsynaptic degeneration, traumatic, compressive, hereditary, toxic and infectious optic neuropathy.

RÉSUMÉ

Les causes de l'excavation non-glaucomateuse de la papille pâle dès l'âge de 65 ans sont rapportées: anomalies congénitales, myopie, maladies ischémiques, dégéneration transsynaptique, névrite optique par accident, maladie héréditaire, produit toxique ou maladie infectieuse.

SAMENVATTING

Een overzicht van de niet-glaucomateuze oorzaken van een bleke geëxcaveerde papil bij 65-jarigen en ouderen: aangeboren afwijkingen, bijziendheid, ischemische aandoeningen, transsynaptische degeneratie, compressieve aandoeningen, erfelijke ziekte, traumatisch en infectieus oogzenuwlijden.

KEY-WORDS

Nonglaucomatous optic disk atrophy and excavation

MOTS-CLÉS

L'excavation non-glaucomateuse de la papille pâle

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INTRODUCTION

One of the presentations of a pale optic nerve head is an optic disk (O.D.) with an excavation. An excavated O.D. is one of the characteristics of glaucoma, a common cause of blindness (24). Pallor and excavation of the O.D. have many causes and the appearance of the O.D. alone can't differentiate the origin of the optic neuropathy (1, 12, 26, 33, 34). This paper deals with the causes of nonglaucomatous O.D. atrophy and excavation in people aged 65 years or older.

GLAUCOMATOUS O.D. EXCAVATION

Glaucomatous O.D. excavation is characterized by loss of the neuroretinal rim area (focal or diffuse obliteration), an increase in the absolute size of the optic cup and in the cup/disk ratio, enlarged chorioretinal atrophy, and asymmetry of these features between the two eyes (16). The most important characteristic of glaucomatous cupping is the vertical elongation of the cup and a corresponding nerve fiber bundle visual field defect. The O.D. damaged by glaucoma typically has cupping which is larger than the pallor (the normal eye has cupping equal to the area of pallor) (26). O.D. excavation with pallor of the remaining neuroretinal rim suggests a cause other than glaucoma.

CAUSES OF NONGLAUCOMATOUS O.D. EXCAVATION

Congenital O.D. anomalies

Congenital anomalies of the O.D. may mimic glaucomatous disk changes. These anomalies are: congenital deep cup, O.D. with anomalous embryonic hyaloid glial-vascular remnants, coloboma of the optic nerve, tilted disk and optic pit. Ophthalmologists should know that central nervous system anomalies are common in patients with malformed O.D. and that patients with these anomalies often exhibit some abnormality in the visual fields. These abnormalities may mimic glaucomatous field defects or mimic field defects due to a compressive lesion. The tilted disk is the most common lesion, which can hide a glaucomatous optic neuropathy. Unlike acquired macrodisk due to high myopia and glaucomatous optic neuropathy, congenital O.D. anomalies do not cause progressive field defects (2).

Pallor of the O.D. associated with high myopia

In eyes with high myopia the O.D. will stretch leading to an acquired macrodisk: the O.D. is larger, more oval and tilted and surrounded by varying degrees of parapapillary atrophy (16). The diagnosis of glaucomatous O.D. excavation by fundoscopy is difficult because the large, abnormally shaped O.D. often has a shallow, concentric cupping. Furthermore:

- The normal red color of the O.D. is not always seen in highly myopic eyes.
- The myopic changes in the posterior pole of the fundus and the large parapapillary area of atrophy are responsible for field loss that is similar to field loss due to glaucoma.
- Normal tension glaucoma is not unfrequent in highly myopic eyes.
- O.D. hemorrhages, frequently seen in normal tension glaucoma, are less often seen in cases of high myopia with normal tension glaucoma.
- Nerve fibre layer defects (an early sign of glaucoma) are more difficult to observe in cases of high myopia.

Ischemic disorders

Anterior ischemic optic neuropathy (AION)

AION is the most common cause of acute optic neuropathy in patients over 65 years. It can be nonarteritic (nonarteritic AION, often associated with arteriosclerosis or diabetes mellitus) or arteritic (arteritic AION associated with giant cell arteritis) (15, 20). At the early stage the characteristics are a pale, swollen nerve head with hemorrhages, an altitudinal field defect and a history of abrupt visual loss (3). The end stage in the arteritic form is neuroretinal rim loss, pallor and excavation. However, in contrast to glaucoma, the end stage in the arteritic form is not associated with an enlarge-

ment of parapapillary atrophy. The end stage in the nonarteritic form shows segmental or diffuse pallor without excavation (9, 23).

Posterior ION (PION)

PION is established as a definite clinical identity and is caused by ischemia due to occlusion of one or more nutrient arteries, which supply the posterior part of the optic nerve in the orbit. Diagnosis of PION is made when neither disk edema nor hemorrhages are present in the acute stage and atrophic disk changes follow about 5-6 weeks later (14). There is always a history of systemic disease or severe hypotension (severe blood loss, hypotension during anesthesia). Development of cupping and pallor in PION have been reported (29).

Central retinal artery occlusion

Excavation of the O.D. after central retinal artery occlusion has rarely been reported and then only long after the event (1).

Senile degenerative sclerosis of the vessels in the optic nerve head

Cases of cavernous degeneration of the O.D. due to senile degenerative sclerosis of the vessels in the nerve head causing chronic ischemia of the retrolaminar segment of the optic nerve head have been reported (7, 11, 30).

Transsynaptic retrograde degeneration

- The large optic excavation in children with periventricular leucomalacia is probably caused by retrograde transsynaptic degeneration across the lateral geniculate nucleus (6).
- Characteristic in neuritis due to multiple sclerosis is atrophy of the temporal side of the disk as a result of involvement of the papillomacular bundle. Band O.D. atrophy and O.D pallor and excavation have also been reported in diseases of the visual pathway due to multiple sclerosis.
- Transsynaptic degeneration as a mechanism of O.D. pallor and excavation has been described in lesions of the occipital lobe (4, 25).
- Patients with Alzheimer disease have an increased cup-to-disk ratio and cup volume and decreased disk rim area. There is antegrade degeneration secondary to retinal ganglion cell degeneration and retrograde degeneration originating at the optic tract or cortical level (35).

Traumatic optic neuropathy

In traumatic optic neuropathy there is visual loss caused by deformational forces that affect the optic nerve and can be categorized in direct and indirect injuries (31). Direct injuries result from an open wound with injury to the optic nerve. Indirect injuries can be caused by forces transmitted through the bones of the skull or by stretching of the optic nerve. Radiation injury is also a cause of traumatic optic neuropathy (22). Pallor and excavation of the O.D. due to traumatic optic neuropathy have been reported (19).

Compressive optic neuropathy

Compressive lesions affecting the anterior visual pathway may produce O.D. pallor and excavation due to axonal degeneration. Most of these lesions are benign and amenable to therapy (18). Some of the compressive lesions affecting the anterior visual pathway over the age of 65 are pituitary macro-adenoma, suprasellar meningioma, craniopharyngioma, giant carotid-ophthalmic artery aneurism, calcification, dilatation and ectasia of the internal carotid artery, optic neuropathy of Graves' disease (18, 28). There are no characteristic O.D. findings in compressive lesions affecting the anterior visual pathway (1). Therefore neuroimaging studies are often necessary to evaluate patients with a pale and excavated O.D. (13). Some differences are worth mentioning:

- Age: compressive lesions affecting the anterior visual pathway are more common before the age of 50 years; glaucoma is more common after the age of 50.
- Onset: patients with compressive lesions affecting the anterior visual pathway do not have a history of abrupt visual loss; patients with ION do and may have systemic diseases (arteritis, diabetes mellitus).

– Family history: patients with compressive lesions affecting the anterior visual pathway do not have a family history; patients with glaucoma sometimes do.
– Central visual acuity: patients with compressive lesions affecting the anterior visual pathway have often a visual acuity less than 5/10, whereas glaucoma patients may keep a central visual acuity of 10/10 even in a more advanced stage of their disease (12).
– Visual field defects: classic glaucomatous visual field defects are nerve fibre bundle visual defects that correspond to the vertical cupping of the O.D.. The visual field defects in compressive lesions affecting the anterior visual pathway produce bitemporal defects or vertical aligned defects that border the vertical meridian. However virtually any field defect may occur with any optic neuropathy (10).
– Pupillary abnormalities: afferent pupillary abnormalities are also seen in a blind eye due to glaucoma and therefore are not helpful in differentiating glaucoma from compressive lesions affecting the anterior visual pathway.

Leber's hereditary optic neuropathy

The onset of visual loss typically occurs between the ages of 15 and 35 years in most pedigrees. Exceptionally Leber's hereditary optic neuropathy can also become manifest after the age of 65. Visual acuity rapidly deteriorates to 20/200 and visual field defects typically are central or cecocentral absolute scotomas surrounded by a narrow rim of relative scotoma. In the acute stage the fundus shows a telangiectatic microangiopathy around the O.D., swelling of the nerve fibre layer around the O.D. and absence of leakage from the O.D. on fluorescein angiography. Eight weeks later the O.D. becomes pale. In some cases the O.D. shows pallor and excavation. Most patients suffer from definitive vision loss; although some may recover partially their vision. The fellow eye is affected within weeks or months (5, 8, 17, 18, 32).

Methanol - induced O.D. excavation

Consumption of methanol causes toxic effects on the optic nerve (papilledema within the first two days) and extensive O.D. excavation probably due to retrograde ganglion cell loss, secondary to acute demyelinisation (8, 27). Visual impairment may range from mild blurring of vision to complete blindness. Visual field defects nearly always include the cecocentral area.

Parasyphilitic optic neuropathy

In cases of tabes dorsalis an inflammatory process in the posterior part of the optic nerve will cause optic nerve atrophy. The O.D. will become uniformly pale but the rest of the fundus will otherwise be normal. The borders of the O.D. will be sharply outlined. The lamina cribrosa usually can be plainly seen at the bottom of the physiologic cup. As there is recession of the gray-white O.D., it is necessary to differentiate such O.D. from glaucomatous excavation (21, 36).

CONCLUSION

There are many causes of nonglaucomatous O.D. excavation in people over 65 years of age. Most important to differentiate these causes are: the best corrected visual acuity, the shape of the visual field defect, the onset (abrupt or slow), the family history, the presence of systemic diseases, the result of the neuroimaging studies. An accurate and early diagnosis can often avoid visual impairment.

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