Nerve Head Anomaly Associated with Pituitary Tumor

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A 54-year-old Caucasian female was seen in the School of Optometry, University of Waterloo clinic for field testing to rule out possible glaucoma. She had had a pituitary tumor (histologically confirmed as a chromophobe adenoma) removed 10 years previously. Field testing and fundus inspection ruled out glaucoma: there were no nasal steps and no scotomas in the arcuate areas.

Field testing (Figs. 1 and 2) did indicate some midline-type superior bitemporal defects. Moreover, the blindspots on field testing were displaced downward and out (although the patient has only a modest myopia (around 1.5 D). Ocular fundus examination revealed a fairly striking situs inversus OS1 (Fig. 3).

Comment:

The visual field findings in this case serve to remind the clinician that targets such as the Goldmann I-2 target (or finer, dimmer targets) should be used when seeking midline-type defects, as the stronger target (Goldmann I-4) failed to show a midline cut in the isopter. The Goldmann I-2 target is reasonably equivalent to a 1/1000 W target at the tangent screen, provided the screen illuminance is on the order of 108 to 160 L/sq m (10 to 15 foot-candles).

Age-matched normal perimeter data are shown with short broken lines in Figs. 1 and 2. The inner pattern of short broken lines is a normal I-2 isopter.

Fig. 1 Results of visual field testing for right eye. Three test stimuli were used: Goldmann I-2 (equivalent to a 1/1000 W target at the tangent screen when illuminance = 10 to 15 foot-candles), Goldmann I-1 (0.5 log units dimmer than I-2) and Goldmann I-4 (1 log unit brighter than I-2). A complete isopter (indicated by arrow) was plotted using the I-2 stimulus. The other two stimuli were used to assess the superior temporal portions of the field (incomplete isopters marked by arrows). For comparison, short dotted lines indicate normal age-matched normal results for the I-2 stimulus (complete normal isopter shown), and for the I-4 stimulus (partial isopter shown). The I-2 isopter shows a definite break at the midline: there is a superior temporal loss here. The dimmer I-1 target revealed not only a superior temporal loss, but an overall depression of the superior field. The brighter I-4 target showed no field abnormality, while the outer, incomplete pattern of short broken lines is from a normal I-4 isopter. Note that the patient is showing a larger-than-normal isopter in three quadrants out of four; this makes the superior temporal loss even more apparent. Note further that the patient’s I-4 data match the normal data fairly closely in the superior temporal quadrants, with no disturbance at the midline.

Fig. 2 Similar findings for the left visual field. Here, the I-1 target shows a loss with less of an overall superior character and more of a superior temporal nature.

The I-4 blindspots are shown as a pattern of unconnected dots: they are displaced in an outward and inferior direction. Such a finding is not uncommon in relatively high myopes (say greater than 5D), but the patient has only a modest degree of myopia (1.5 D). This prompts a closer examination of the fundus photos. Ordinarily, a horizontal line passing through the center of the fovea will cut across the nerve head about 1/2 of the way up from the inferior edge. This does not occur in these photos. This suggests a rather inferior positioning of the fovea relative to the nerve head. The right nerve head appears to be smaller than the left.

Taylor2 reported on an association between tumors affecting the anterior visual pathway and malformations of the optic nerve head. The tumors found in his seven cases included glioma, astrocytoma (one pilocytic and one fibrillary), a suprasellar mass (which was not biopsied), and craniopharyngioma. The
associated malformations of the nerve head included conus (frequently inferior), dysversion of the disk, situs inversus of the central retinal vessels, and hypoplasia of the nerve head (both overall and segmental).

Keane reported a further two cases in which tumors were found in the chiasmal region (one was a chromophobe adenoma, the other a pinealoma): in the first case, one disk appeared somewhat hypoplastic, while the other showed an inferior nasal conus; in the second, the left eye showed a strong superior nasal oblique direction of the optic nerve, with an associated inferior temporal conus. He felt that the nerve head findings were unrelated to the tumors.

This case would appear to be another instance of an association between a malformation of the optic nerve (situs inversus) and a tumor affecting the anterior visual pathway, in this case a pituitary tumor (chromophobe adenoma). This patient did not become aware of the tumor until her menstrual cycle stopped unexpectedly. She had had several successful pregnancies prior to this. In retrospect, it may be said that the abnormal nerve head was an early (i.e. congenital) warning which foreshadowed the development of the tumor.

The notion that the forces leading to development of a tumor in adult life are active during the earliest embryonic stages of development may seem surprising; however, this concept becomes more reasonable, as Taylor points out, when one considers that many of the tumors mentioned above are regarded as developmental in origin: they are due to the presence of pockets of embryonic tissue (embryonic rests) which undergo abnormal growth later in life.

Many people show situs inversus and other interesting nerve head anomalies, while only a few people develop tumors of the sort mentioned here. Nonetheless, this case should prompt at least a visual field assessment for patients with congenital nerve head anomalies.

References:


Fig. 3 (upper): right nerve head. Deep Elschnig II type with well-defined, unbroken neuroretinal rim. It was apparently this deep cupping which prompted referral by her GP for a glaucoma assessment.

Fig. 3 (lower): left nerve head. Central retinal vessels (as seen at bottom of cupping) appear to originate at the temporal side of the nerve head. This is typical of situs inversus. A deep Elschnig II type, also with well-defined (and unbroken) neuroretinal rim.