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Cognitive Visual Disturbances in Chiasmal Disorders: A Review on Exceptional Visual Disabilities as Based on 30 Years' Clinical Experience

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Abstract

Purpose: To direct attention to a generally ignored cognitive element in chiasmal symptomatology, with the aim of reducing the diagnostic delay in such cases, where complaints are typically vague. A minority of chiasmal patients thus present with cognitive ignorance or mis-interpretation in visual space, probably caused by conductive loss of information as usually required for higher brain centres to induce relevant eye movements.

Methodology: Personal experience from more than 30 years' neuro-ophthalmological service is overviewed, primarily based on observation of abnormal behaviour during routine clinical testing for visual acuity (eventually summing up to 64 patients). Neuro-psychological testing and extended imaging (including functional positron emission tomography imaging as a pilot study) were carried out in part of the series.

Major findings: A main feature has been lateralizing behaviour during unilateral visual acuity testing, showing a lateral ignorance on the board. Many also presented with inability to read print and a few had bizarre mis-interpretations in visual space.

Conclusions: The cognitive aspects have gained little attention in the literature. As a guide to early diagnostic attention, however, acknowledging aberrant behaviour in the eye clinic may be a marker and thus an important step for achieving a better prognosis after surgery for a chiasma-related condition.

Keywords Chiasmal disorder; Cognitive defect; Neglect-like behaviour; Neuropsychological evaluation; Functional PET scan

Introduction

A combined delay by the patient and the doctor is a common feature of chiasmal disorders. For the patient, the main reason seems to be unawareness of visual loss as being associated primarily with slow growth of the basic lesion (typically pituitary adenoma, craniopharyngioma, or skull base meningioma). Delay can also arise due to the insidious and asymmetric nature of the initial visual field drop-outs, the defects of one eye being compensated for by the other when binocular. Complaints, when acknowledged, are usually vague.

When seen in the clinic, however, a minority of patients may present more specific features that include cognitive elements, the most frequent being a lateralizing ignorance under customary testing for single eye visual acuity [1,2]. Usually this manifests as varying degrees of nasal preference on the board, or a temporally located ignorance (NP/TI). Some patients experience an inability to read print, despite apparently normal or fair visual acuity data. A few also describe mis-interpretation of moving events in visual space. The following is suggested as a kind of collective definition: a cognitive ignorance or mis-interpretation in visual space, probably related to conductive loss of information to supratentorial connections and not readily compensated for by relevant eye movements.

Reviewing more than 30 years of personal neuro-ophthalmic experience, the aim of this article is to direct attention to such occasional cognitive signs, a topic that has gained little attention in the literature, including textbooks [3]. For the eye specialist, it is the incentive to order a visual field examination and to ask for further evaluation as indicated.

Clinical Findings

Single eye visual testing showing NP/TI

A typical response may be recognition merely of the nasal column of symbols when testing single eye visual acuity, whether on a Snellen type board or using projected optotypes. The lateralizing behaviour is abolished when binocular, although the NP/TI pattern recurs on monocular re-testing; despite fresh conception of a full board, the usual saccades for searching the line cannot be mobilized.

Over 3 decades, varying degrees of this aberrant visual behaviour have been acknowledged in a total of 64 adult patients, in one eye or in both, and often with a side difference when of binocular occurrence [2]. It may be associated with visual acuity loss, but has also occurred in patients with a decimal best corrected visual acuity of 1.0 in both eyes. One such patient with full monocular acuities presented 4-5° paracentral heteronymous scotomas as the only visual field manifestation; both eyes were strictly nasal on the chart, and reading books had become impossible. In cases with markedly reduced visual acuity (e.g. 0.16–0.05), single eye testing may even add the most central nasal 10–20° to the hemianopic temporal visual field loss . Paradoxically, fixation usually appears stable in such cases despite the

apparent inclusion of the central target for fixation in the functional scotoma. When occasionally combined with temporal hemianopia to midline in the fellow eye, the 2 recordings suggest a central vertical bar as not recognized. At binocular testing, however, no drop-outs are marked correspondingly. Due to a less strained convergence, tangent screen testing at a distance of 1 m (e.g. white 10/1000) may show better compliance than the close working conditions demanded by static computerized or Goldmann kinetic perimetry. A full central field is given, including the monocularly suggested missing vertical bar.

Visual lacunae, disorientation in space

- Four examples can be given.

A couple walking on the street towards a 58-year-old female patient with visual incapacities after previous chiasmal surgery was perceived as having a total of 3 legs. Further, the patient could not decide whether the shared leg was wearing trousers or a skirt.

A 59-year-old bank manager chose to retire from service when reading text became impossible, despite a normal visual field of the fellow eye and best corrected visual acuities of 0.7 and 1.0 when first examined. As a golf player he had a visual slip of about 1 m on the putting green; for a short distance the golf ball simply got out of sight. Both complaints completely recovered after medication for his prolactinoma [2].

A 50-year-old office clerk, also with bitemporal hemianopia, was able to cover many of his former duties after relevant surgery, but could not recognize his usual station where to get off the train on return from work.

A 35-year-old man who had undergone repeated surgery for an extensive craniopharyngioma often complained of visual unawareness despite familiar surroundings.

Neuropsychological testing (n=18)

Slow and incomplete recognition of printed symbols scattered over a chart (Gothenburg test) was a feature of many eyes and indicated that the usual search by saccades were not performed in an orderly fashion [4]. Skew line division and difficulty in reporting a reading bar with 10 letters closely situated were parallel findings. A few patients could not draw a simple figure after they were shown a model (e.g. a simple house). Revision of reading glasses had no effect, but some were able to change strategy and gained moderately when re-tested.

Overall, the mis-interpretations seemed to be cognitive and not just due to reduced visual acuity or field loss. With regard to the location of the principal lesions and visual pathway conductivity, there was a slight preponderance of cases showing what might be perceived as lost lateral geniculate body stratification of information as forwarded to the right hemisphere. However, there was no support from brain imaging for supratentorial lesions, which are typically associated with visual neglect in patients with a stroke.

Findings by neuro-imaging

Computed tomography and magnetic resonance imaging are effective tools in the pre-operative evaluation of chiasmal lesions, and for follow-up of occasional residual lesions after extirpation. Usually, imaging after surgery indicates morphological changes related only to the operative field. As already mentioned, we found no hemisphere lesions in our patient series, which from text book knowledge would have been the most obvious finding in relation to neglect-like functional drop-outs perceived here as cognitive. On this background, functional positron emission tomography imaging of the brain was performed in 4 adult patients, after informed consent, however without definitive findings [5]. Only 1 of the 4 showed reduced metabolic activity, manifesting in the left hemisphere primary visual cortex and higher order visual areas. Thus, general suggestions about the mechanisms underlying the marked cognitive defects in those selected for the pilot study remained elusive, and the project was discontinued.

Discussion

Because cognitive aspects are mainly ignored in the literature, the aim of this survey was to direct attention to the minority of chiasmal patients who may present with a more specific indication of the diagnosis, as here described. In this respect, they are different from the majority of patients with chiasmal disorders, who usually have only vague complaints, if any. With early diagnosis, the prognosis after surgery clearly will be better, and many can live normal lives. Our more advanced cases typically had extensive peri-chiasmal pathology when operated upon, and a review of the clinical course often indicated that a visual field examination could have been decisive for a better outcome if carried out at an earlier contact with the medical service.

From the visual data, we can suggest no obvious mechanism underlying the neglect-like cognitive events, of which the not infrequent NP/TI behaviour at single eye visual acuity testing has remained the most obvious feature. Nor has brain imaging yielded explanations [6-8]. In theory, disturbed binocularity (e.g. hemifield slide) may be elicited when binocular [9]. Using both eyes, however, was a stabilizing factor in many cases. Orthoptic evaluation generally indicated orthophoria, and voluntary eye movements appeared intact. Some patients presented moderately reduced Titmus stereopsis, more than expected from best corrected visual acuities. Functional bitemporal post-fixational scotomas have also been discussed [3], but such visual drop-outs would presuppose fixation not on the object in space, but on a point between the object (in the above example, the golf ball that briefly disappeared close to the hole) and the eye, a behaviour that seems very unlikely.

With regard to frequency, I previously suggested that cognitive elements were part of the signs and symptoms in a few percent of patients with chiasmal lesions [2]. Low expectancy may thus contribute to the fact that they escape attention. Accordingly, experienced colleagues have often had a hesitant attitude when the item was discussed, compounded by the virtual absence of the topic in most textbooks. An important precondition for discovering the aberrant behaviour seems to be the neuro-ophthalmologist's own observation of slow or odd performance and lateralizing behaviour when testing for visual acuity. At least, this has been my primary access to the topic, but as an indication it may be lost if this part of the evaluation is performed by assistants in the clinic.

In summary, the earlier we catch lesions to the chiasm, the better. The cognitive element may therefore be a valuable early indication for the ophthalmologist when exceptional patients present with what seems to be just minor visual problems.

Conflicts of Interest

No conflicts of interest

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