Optic ataxia: clinical-radiological correlations with the EMIsan¹

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SYNOPSIS After coronary by-pass surgery, a 47 year old, right-handed man developed a Gerstmann's syndrome, a visual-spatial perceptual deficit, and a gross impairment of movement under visual guidance ('optic ataxia'). Visual fields and extraocular movements were intact; he had a left hemiparesis. The EMIsan showed three lesions: a left parietal-occipital lesion; a posterior callosal lesion, and a right frontal lesion. It is hypothesized that optic ataxia in both visual fields requires bilateral lesions which, in the present case, were strategically placed so as to effectively disconnect motor cortex from visual input.

Optic ataxia is a disorder of visually guided movement, most evident when the patient attempts to reach for objects. As originally described by Balint (1909), it is usually seen in the context of severe oculomotor impairment. A similar disorder, also associated with gross oculomotor disturbances has been described under the name of visual disorientation (Holmes, 1918; Michel *et al.*, 1965). A few cases, however, have been reported with normal extraocular movements and optic ataxia confined to a single visual field (Riddoch, 1935; Stenvers, 1961; Rondot and de Recondo, 1974). Recently, understanding of the possible mechanisms of this disturbance has been enhanced by a report of a similar phenomenon in monkeys after experimental lesions (Haaxma and Kuypers, 1974).

This paper describes a patient who showed optic ataxia in both visual fields with no abnormalities of oculomotor function, visual acuity, or visual fields. He had a left hemiparesis, a Gerstmann's syndrome, and a moderate visual-spatial deficit. Computerized axial tomography

of the head (EMIsan) of this patient proved particularly helpful in providing information concerning the extent and localization of his brain lesions. On the basis of the findings reported below, a model of the mechanisms of visual control over movement is proposed which accounts for the presence of optic ataxia without disturbance of oculomotor function in either one or both visual fields.

CASE REPORT

A 47 year old, right-handed insulation contractor underwent triple graft coronary artery surgery on 28 November 1973. A few hours after surgery he had failed to waken from anaesthesia and was found to have a left hemiplegia. The patient remained unresponsive for nine days, after which his state of consciousness gradually improved. By the nineteenth day after surgery, he was able to speak and understand, but there was considerable uncertainty regarding his vision. Some examiners reported that he could count fingers and name colours, others that he failed such tasks. At times he seemed to show a left homonymous hemianopia, while at other times he 'appeared blind'. His eyes and head tended to deviate to the right; he seemed unable to move his eyes beyond the midline except on labyrinthine stimulation. On 30 January 1974 he was discharged to a rehabilitation hospital. His eye movements and

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motor strength improved, except for a continuing paresis of the left arm. In February, the patient had a tendency to extinguish in the left visual field on double simultaneous stimulation, and one examiner reported a right inferior quadrantic visual field defect on double simultaneous stimulation. He still showed a mild inconsistent paresis of gaze to the left.

On 13 March, he was transferred to the Neuro-behavior Unit of the Cleveland Veterans Administration Hospital. At that time visual fields were examined by formal perimetry, tangent screen, and double visual stimulation by the neurology and ophthalmology services independently, and no defect could be detected. The physiological blind spot was normal in size, and he had no scotoma. Extraocular movements were now full both on command and when following objects. Optokinetic nystagmus was present at times, but most observers were unable to obtain any movement.

General physical examination was always unremarkable except for a scar over his chest. The patient was fully alert and cooperative and complained only of 'poor vision'. His spontaneous speech was intact, his naming ability nearly normal, and his comprehension and repetition of speech normal. Spelling was mildly impaired. Based on the four verbal comprehension subtests the patient's WAIS Verbal IQ was 106. His attention span seemed slightly deficient (five digits forward), and he was mildly impaired on tasks requiring more active concentration. Although memory for both recent and past events and his immediate and delayed recall of stories were normal, his performance on a paired

associate learning task was erratic, tending to be mildly to moderately impaired. The patient identified the correct principles for categorizing the stimuli on the Weigl Color-Form Test (Goldstein and Scheerer, 1941) yet failed to sort the stimuli accordingly. On the Seashore tests (Milner, 1962) discrimination of rhythmic patterns was normal but memory for tonal patterns was moderately impaired. He could sing and repeat simple tunes without difficulty.

The patient showed deficits on some visual perception tasks where no motor act was required. When presented with two simple designs in vertical array, one of which was either identical with the other or in a rotated position, he tended to describe similar designs as different. On the Hooper Visual Organization Test (Hooper, 1958), in which the subject is required to identify an object from a picture which has been cut up and randomly arranged on a card, he obtained a deficient score (13 out of 30 correct). He was able to discriminate rods 5 and 15 cm but not 5 and 10 cm long. His depth perception appeared intact. Directional orientation and right-left discrimination were poor. He had moderate difficulty in describing the floor plan of his home. On a test of recognition memory for visual designs the patient was mildly to moderately deficient. He had no difficulty in recognizing familiar or in learning unfamiliar faces.

He was nearly normal in abstracting verbal information from visual stimuli. Naming of objects shown to him was carried out with only a few hesitations. Colour naming was more erratic; he occasionally misnamed colours, calling them by names of

FIG. 1 Samples of the patient's attempt to (a) copy a square and (b) write his name. Note the inability to draw a horizontal line.

closely related hues. When presented with an array of objects or pictures, he could not count them or describe them consistently. On the other hand, when given a single, complex scene he was able to describe the picture relatively well. Analogously, the patient could correctly read single letters or numbers, while his performance with larger sets of written information was quite erratic. He was usually unable to read correctly numbers with three or four digits, often adding a digit at the end of the string rather than simply ignoring a number. His performance on reading single words was erratic, and on a few occasions he correctly read an entire sentence that was presented in large letters on two lines. He could neither read nor set a clock. Mental arithmetic was performed poorly, whether problems were presented orally or visually. Finger localization was severely impaired.

In striking contrast with the mildly to moderately impaired and erratic performance on tests of spatial organization, visual comparison, and verbal description of visual material, he was consistently very severely impaired on any task that required coordination of movement with visual perception. When attempting to reach out with his right hand or either foot to touch an object presented to him, the patient tended to reach to the left and groped until he came into contact with it. Yet his eyes appeared to be directed toward the object. When asked to point to an object, he pointed very vaguely in its direction, declaring himself satisfied with his performance when questioned. He showed similar groping movements when asked to touch an extended part of his own body—for example, to grasp his left thumb with his right hand—although he was able to touch his body accurately when he was, for instance, saluting or drinking, movements presumably less dependent on visual control. Once he had grasped an object, he handled it appropriately and could name it when blindfolded (even in his paretic left hand when he was assisted in palpating it). He could demonstrate the use of an object in its absence and could carry out complex movements on command. He was unable to draw a horizontal line and consistently drew vertical lines rotated nearly 45 degrees, usually to the left. Drawing, writing, or copying even the simplest form or letter was grossly impaired (Fig. 1). He would not attempt cursive script, and his block letters tended to resemble a series of diagonal lines running across the diagonal of the paper. Constructing a block design or assembling objects was impossible, although he often recognized that his performance was erroneous. He could frequently identify the object that he could not assemble, and, as noted above, though his total score on the Hooper test was deficient, he succeeded in identifying a

number of objects from examining a picture of a random arrangement of cut-up parts. While blindfolded, he could not place even one of 10 blocks in a form board. It is possible that even without visual input such tasks may require the coordination of movements with internal representation of visual information. Using vision, he also failed completely but tended to grope in the vicinity of the correct hole.

The rest of his neurological examination showed a mild left spastic hemiparesis affecting especially the distal segments of the left upper limb. Strength of flexor muscles was nearly normal in his left lower limb. There was no facial, tongue, or palatal asymmetry; cerebellar testing was normal. He was able to stand but could walk only with assistance because of poor visual-motor coordination. Sensory examination was normal.

It was difficult to determine the patient's visual acuity, because he was often unable to identify accurately a small number or letter pointed out to him but at other times he read fairly small print and identified small objects at a distance, suggesting functionally normal visual acuity in both eyes. Fundi were normal. Pupils were equal and reactive to light and accommodation.

FIG. 2 EMIsan performed at the level of the lateral ventricles. Three areas of low density are seen: in the left parieto-occipital junction (short solid arrow); in the right frontal lobe (open arrow); and in the posterior corpus callosum (long solid arrow). (EMIsan performed at the Cleveland Clinic).

Routine haematological and blood chemistry examinations were normal. Skull radiographs were normal. An EEG performed on 7 January 1974 showed slowing (delta activity) overlying the right frontal lobe. Brain scintiscan performed on 20 March was negative in the anteroposterior, postero-anterior, and both lateral views. The anterior vertex view, however, showed an area of increased uptake of isotope in the right frontal area. An EMIsan (Fig. 2) at the level of the lateral ventricles showed one area of low density of approximately 4 cm in diameter lateral to the right frontal horn; a second, somewhat less than 3 cm in size, in the left parietal convexity lateral to the occipital horn; and a third in the posterior corpus callosum just to the left of the midline. These lesions were considered to be infarcts. The ventricular system showed no displacement.

DISCUSSION

After coronary surgery this 47 year old man developed a Gerstmann's syndrome and a moderate deficit in the perception of visual-spatial information. His most striking and unusual symptom, however, was a gross impairment of movement under visual guidance (optic ataxia). In analysing the neuroanatomical correlates of these deficits we will rely on the results on the EMIsan. Having presumably suffered an anoxic episode, the patient may have been left with lesions smaller than the limit of resolution of the EMIsan (approximately 1 cm). Since he recovered to the extent that he showed no motor impairment on his right side, no aphasia, and no somatosensory deficits or visual field cuts, there was no indication of other lesions. Therefore, it seems reasonable to account for the patient's neuropsychological deficits on the basis of the lesions indicated on the EMIsan.

The extraordinarily severe impairment of visually guided movement seen in the present patient would appear to be quite rare. He was not apraxic in the classic sense of Liepmann: he could perform movements well, both on command and in imitation, and could handle objects correctly upon grasping them. His difficulty in reaching for objects seems far more striking than the impairment seen in severe constructional apraxia; patients with such disorders have not been described as groping for objects as did the present patient. His behaviour, though more erratic, resembles that of subjects whose vision is distorted by a prism. His con-

dition is similar to the optic ataxia or visual dis-orientation described by other authors (Balint, 1909; Holmes, 1918; Michel *et al.*, 1965); although the present patient showed neither abnormalities of extraocular movements nor visual inattention, prominent factors in Balint's syndrome and in Holmes' 'disturbance of visual orientation'.

Instances of impairment of visually guided movement within a *single* visual field contralateral to the lesion have been reported in four cases of left (Riddoch, 1935; Stenvers, 1961) and four cases of right parietal-occipital lesions (Stenvers, 1961; Rondot and de Recondo, 1974). All eight patients had difficulty in pointing and judging distance within the impaired field and, more importantly, at least four of them had no disturbance of eye movements, hemianopia, or visual inattention. Although Rondot and de Recondo (1974) emphasized the fact that their patients, while unable to reach for objects accurately, could grasp their own body parts readily, the present patient was defective on both tasks, emphasizing the generality of visual control over movements. An experimental analogue has been offered by Haaxma and Kuypers (1974) who, by placing unilateral lesions in the white matter of the caudal parietal area of split brain monkeys, produced impairment in the visual guidance of contralateral hand and finger movements.

The following analysis of the mechanisms of visual control over movement is tentatively offered as an explanation of the severe deficit experienced by the present patient. Each motor centre receives information concerning the visual fields through two sources: (1) ipsilaterally via pathways from the visual association area, and (2) transcallosally from the contralateral frontal association area. These pathways carry information for both visual fields which has been transferred across the corpus callosum between the posterior association areas. For a lesion to impair visually guided movement in one field alone in the absence of a visual field defect, it would have to be placed in the contralateral posterior association area or in the pathway leading to it from the primary visual centre in that hemisphere.

Impairment of visually guided movement in both visual fields would require at least two

lesions, one in each hemisphere (or a unilateral lesion combined with a complete commissurotomy). In the present patient, visual information reaching the right occipital cortex was presumably intact, yet it could not be utilized by the ipsilateral motor centre due to the large right frontal lesion. Neither could it be transferred to the opposite hemisphere: the right frontal lesion presumably blocked transmission across the anterior corpus callosum, and the small posterior midline lesion disrupted posterior cross-callosal transmission. Visual pathways from the left occipital cortex may have been disrupted by either or both of the posterior lesions. The left parietal lesion may have blocked visual information before its transmission to the left motor association area; or that part of the posterior callosal lesion which extends into the posterior left hemisphere may have intersected visual pathways extending to left motor association areas, as well as blocking cross-callosal transmission to the right visual association area.

The lesion in his left parietal area most certainly accounted for his Gerstmann's syndrome (finger agnosia, right-left disorientation, dyscalculia, and agraphia). The location of the lesion responsible for his visual-spatial deficit is less evident: it is not clear on the basis of our understanding of the cerebral localization of visual perception whether or not the patient's visual-spatial deficit was due to his right frontal, his left parietal, or to an undetected right posterior lesion. It should be noted that a right posterior lesion could be placed so as to effectively disrupt transmission of visual input of the left visual field to both the left posterior association area and the ipsilateral frontal association area. Thus, although one might argue for the existence of an undetected right posterior lesion in view of the patient's visual-spatial deficit, the model described above would apply equally well.

Though apparently rare, the syndrome of optic ataxia would undoubtedly be observed more frequently, both unilaterally and bilaterally, if it were not for the masking effect of visual field defects, hemi-inattention, and hemiparesis. Furthermore, it seems possible that some patients with subtle 'subclinical' manifestations of a disorder like that described above might be identified by careful testing of visual localization by the technique of Ratcliff and Davies-Jones (1972).

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