Case report

Reversible Holmes’ syndrome complicating cardio-pulmonary resuscitation

K J Fullerton, K Balnave

Accepted 24th July 1985.

The two cardinal features of the syndrome described by Holmes in 1918\(^1\)\(^,\)\(^2\) are a disorder of ocular movement and a disorder of spatial orientation. The former is manifested by severe problems of ocular fixation and accommodation, such that the patient finds great difficulty in fixing his or her gaze on an object and, having done so, has even greater difficulty in moving the point of fixation to look at something else. This means, for instance, that he cannot follow a moving object with his eyes. The second feature means that the patient has difficulty in orientating objects in three-dimensional space, so that he cannot pour a glass of water without spilling it, and, as in this case, has difficulty in lifting food with a fork or spoon. It can also result in disturbance of topographical memory so that he cannot find his way around, even in familiar surroundings. Other features found in some, but not all of Holmes’s original six cases include: hemiparesis, hemisensory loss, speech disorders, visual field defects, visual neglect and apraxia.

CASE HISTORY

A 57-year-old right-handed housewife was admitted to the coronary care unit following an acute inferior myocardial infarction. Three hours after admission she had the first of a series of four episodes of ventricular fibrillation. During the last of these, she required a period of cardio-pulmonary resuscitation lasting 45 minutes and artificial ventilation for a further 48 hours. It was another four days before a normal level of consciousness was restored. She then complained of being unable to eat because she ‘kept missing her food with her fork’ and of being unable to read.

On examination of the central nervous system there was no weakness, sensory loss, hemianopia, or unilateral neglect. Comprehension of speech and cognition were unimpaired, but she had a nominal dysphasia. When she wore her normal glasses, visual acuity was bilaterally 6/6 though she said that it took several seconds for objects to come into focus. She was able to move her eyes in any direction on command, though if the eyes were fixed on a particular object and a...
torch was flashed in her peripheral visual field, the normal saccadic searching eye movements in the direction of the new stimulus were absent. On being asked to read, she could normally make out a single word but scanning movements were absent.

There was great difficulty in orientating objects in three-dimensional space, so that she missed when asked to grasp an object held out by the examiner, and could not pour a glass of water, or pick up a piece of food with her fork. There was a disorder of topographical memory so that she frequently had to stop and ask for directions on her way to the toilet, which she could clearly see and recognise from her bed. In addition, she demonstrated all the signs of a Gerstmann syndrome (right/left disorientation, finger agnosia, acalculia, agraphia). She had good insight into all her disabilities.

At the time of discharge, four weeks after the cardiac arrests, her dysphasia had resolved, but her other signs were unchanged so that she had difficulty finding her way around her own home. Unfortunately, she refused to attend follow-up or for CT scan, and was not seen for a further two months. She then consented to attend an outpatient clinic, but not to undergo further investigations. Neurological examination at this time was entirely normal and her disorder of topographical memory had resolved to the extent that she could go out to the local shops. She has remained well at subsequent review.

**DISCUSSION**

Holmes’ syndrome is best considered as a development of that described by Balint in 1909, though an earlier description exists. Balint did not note the specific problem of ocular movement which is a major feature of Holmes’ syndrome but referred to ‘optic ataxia’ in his patients. It is probable, however, that both Holmes and Balint were describing the same clinical picture, and the terms are therefore interchangeable, though some authors have argued otherwise.

Reports have been published of sporadic single cases and short series of patients with the disorder, including a case initially presented in this journal. Although few cases have come to post mortem, or had the benefit of newer radiological techniques, it is agreed that the causative lesion is bilateral posterior parietal lobe damage, as in Holmes’ six victims of penetrating head injury.

The occurrence of ‘minor forms’ of the syndrome have led to the suggestion in this case, as in the case of Gerstmann’s syndrome, that the clinical findings represent a constellation of separate neurophysiological deficits, related only because it is possible for them to be caused by a single vascular or traumatic event. Certainly, the disorder of ocular fixation is similar to that found in the affected visual field of a stroke patient with marked unilateral visuo-spatial neglect. In two of Holmes’s patients and one of Hécaen’s, the ocular disorder was more marked in one visual field. A lateralised disorder of topographical memory also occurs in visuo-spatial neglect, and a non-lateralised form in bilateral stroke. It is interesting to speculate, therefore, whether some of the features of Holmes’ syndrome could be explained on the basis of bilateral visuo-spatial neglect. Despite the possibility that Holmes’ syndrome is not a distinct entity, it is important, as in the case of Gerstmann’s syndrome, that the various features should be recognised in the clinical setting. Because they are unusual, it is possible that they could go unrecognised, or be misinterpreted as evidence of mental confusion.
The clinical findings in this patient accord well with Holmes's original cases, and all the features of the syndrome were present. There are two notable points to be drawn from this case report. First, Holmes' syndrome developed following cardiac arrest. There have been previous reports of Holmes' syndrome complicating occlusion of both internal carotid arteries and cardiac surgery, the posterior parietal 'watershed' areas being particularly susceptible to anoxia. Holmes' syndrome is a potentially disabling complication of cardiac arrest which may not be recognised because of its unusual clinical features, and may be commoner than previously supposed. Second, the syndrome resolved completely over a three-month period. Previous studies have tended to suggest that, once it has developed, Holmes' (or Balint's) syndrome is a permanently disabling condition.

As in other forms of cerebrovascular damage, the exact mechanisms of recovery are unclear. It may be postulated that recovery of local oedema and the revitalisation of a 'penumbral' area of cerebral tissue which had not been irretrievably damaged were involved. In the past, this disorder was often diagnosed some time after the cerebral damage had occurred, when the period of maximum spontaneous recovery had already passed, and it is feasible that the condition remained undiagnosed in those in whom recovery had occurred.

REFERENCES

1. Holmes G. Disturbance of visual orientation. Br J Ophthalmol 1918; 2: 449-68, 506-18.
2. Holmes G. Disturbance of visual space perception. Br Med J 1919; 2: 230-1.
3. Balint R. Seelenlähmung des Schauens, optische Ataxie, räumliche Störung der Aufmerksamkeit. Monatschr Psych Neurol 1909; 25: 51-71.
4. Badal J. Contribution à l'étude des cécités psychiques. Alexie, agraphie, hémianopsie inférieure, trouble du sens de l'espace. Arch Ophthalmol 1888; 140: 91-117.
5. Benton AL, Meyers R. An early description of the Gerstmann syndrome. Neurology 1956; 6: 838-42.
6. Michel F, Jeannerod M, Devic M. Trouble de l'orientation visuelle dans les trois dimensions de l'espace. Cortex 1965; 1: 441-56.
7. Allison RS. The parietal lobes: clinical and pathological aspects of their dysfunction. Ulster Med J 1970; 39: 1-19.
8. Allison RS, Hurwitz LJ, Graham White J, Wilmott TJ. A follow-up study of a patient with Balint's syndrome. Neuropsychologia 1969; 7: 319-33.
9. Hécaen H, Ajuriaguerra J. Balint's syndrome (psychic paralysis of visual fixation) and its minor forms. Brain 1954; 77: 373-400.
10. Kase CS, Troncoso JF, Court JE, Tapia JF, Mohr JP. Global spatial disorientation. J Neurol Sci 1977; 34: 267-78.
11. De Renzi E. Disorders of space exploration and cognition. Chichester: Wiley, 1982: 57-137.
12. Kinsbourne M, Warrington EK. A disorder of simultaneous form perception. Brain 1962; 85: 461-86.
13. Critchley M. The parietal lobes. London: Arnold, 1953: 203-25.
14. Brain RW. Visual disorientation with special reference to lesion of the right cerebral hemisphere. Brain 1941; 64: 244-72.
15. Roth M. Disorders of the body image caused by lesions of the right parietal lobe. Brain 1949; 72: 89-111.
16. Dunn TD. Double hemiplegia with double hemianopia and loss of a geographical centre. Trans College Phys Philadelphia 1895; 17: 45-55.
17. Meyer O. Ein- und doppelseitige homonyme Hemanopsie mit Orientierungsstörungen. Monatschr Psychiatr Neurol 1900; 8: 440-56.
18. Russell RW, Bharucha N. The recognition and prevention of border zone cerebral ischaemia during cardiac surgery. Q J Med 1978; 187: 303-323.

© The Ulster Medical Society, 1985.