Alien Hand Sign and Other Cognitive Deficits following Ruptured Aneurysm of the Anterior Communicating Artery

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We describe a right-handed patient who suffered a ruptured aneurysm of the anterior communicating artery (ACoA) which was clipped successfully. Computerized tomography indicated a low density area in the genu of the corpus callosum and the infero-lateral aspect of the left frontal lobe. On recovery the patient's most notable deficit was the "alien hand sign" whereby the left hand would frequently interfere with the actions of the right hand. Problems in response initiation were also evident. There was significant memory loss and performance was impaired on some tests of frontal lobe function. Discussion centres on the functional locus of the alien hand sign but other aspects of the patient's deficits are also considered.

Introduction

The anterior communicating artery (ACoA) is particularly prone to saccular aneurysms with between 30% and 40% of all cerebral vascular aneurysms being located there (Gade, 1982; Steinman and Bigler, 1986). The ACoA has a variable distribution which includes, minimally, the basal forebrain, anterior cingulate, anterior hypothalamus, septal nuclei, columns of the fornix, anterior commissure, and the corpus callosum (Crowell and Morawetz, 1977; Dunker and Harris, 1976). In addition to infarctions within its own distribution, ruptured ACoA aneurysms are frequently associated with vasospasm, particularly within the anterior supply. For this reason ACoA aneurysm patients often have frontal lobe pathology (e.g. Alexander and Freedman, 1984; Vilkki, 1985; Parkin et al., 1988; Parkin and Leng, in press).

The most prominent and widely investigated deficit following ruptured ACoA aneurysm is loss of memory. This deficit is of particular interest because it appears to vary qualitatively from "classic" instances of the amnesic syndrome such as case HM (Corkin, 1984) and alcoholic Korsakoff Syndrome (Butters, 1984). Several studies, for example, report that recognition memory is largely unaffected in ACoA patients even though recall may be quite impaired (Parkin et al., 1988; Volpe and Hirst, 1983) and Delbecq-Derousne et al. (1990) report the opposite in their patient RW. ACoA patients also seem more prone than most memory-disordered patients to manifest "fantastic" confabulatory tendencies (e.g. Stuss et al., 1978; Kapur and Coughlan, 1980; Parkin et al., 1988).
In this paper we describe a woman who suffered a ruptured ACoA aneurysm which resulted in rather different neuropsychological sequelae from those typically reported. Although memory impairment was a significant feature of her disorder, the feature of greatest note was an “alien hand sign” (Brion and Jedynak, 1972) along with clear indications of callosal disconnection.

Case Report

MP is a 46-year-old right-handed woman who woke up feeling faint and dizzy on the morning of admission. She had a sudden onset of occipital headache, blurred vision, photophobia and tingling in both arms, vomited several times but there was no loss of consciousness. On neurological examination she was conscious and orientated with normal intellect and speech function. There was neck stiffness but fundoscopy, visual acuity, and visual fields were normal. Pupils were 3 mm reacting equally to light and accommodation and external-ocular movements were full. Remaining cranial nerves were normal and examination of the upper and lower extremities revealed normal power, tone, sensation and reflexes bilaterally. Plantars were equivocal.

A CT Scan demonstrated diffuse subarachnoid haemorrhage with blood in the third and fourth ventricles and a significant blood collection in the anterior interhemispheric fissure suggesting a ruptured anterior communicating artery aneurysm. The patient remained neurologically stable and a cerebral angiogram performed on 2.4.90 showed an ACoA aneurysm originating on the left side. On 3.4.90 craniotomy and clipping of the ACoA was carried out. The aneurysm was located at the junction of the left anterior cerebral artery and the left A1 segment of the anterior communicating artery. The patient tolerated the procedure very well and had no additional neurological deficit post-operatively. She remained slightly drowsy but otherwise intact. On 6.4.90 she developed an episode of left-sided weakness associated with numbness in the left hand and slight disorientation which resolved completely after intravenous administration of Gelofusine.

Over the next few days, her neurological condition was fluctuating and on 9.4.90 she was noticed to respond variably to commands with no focal deficit but she was totally aphasic. By 12.4.90 she was alert, speaking in sentences with no focal deficit but there was clinical evidence of behaviour associated with frontal dysfunction. Repeat CT Scan on 12.4.90 showed some low density involving the genu of the corpus callosum extending out into the adjacent white matter of the frontal lobe and a little low density change in the infero-lateral margin of the left frontal lobe. There was no evidence of hydrocephalus. It was felt that the changes were almost certainly due to vasospasm. She was gently mobilized and was able to walk with slightly unsteady gait, requiring some assistance. She was also noted to have some left sensory and visual inattention and she continued to be disoriented and apathetic.
Neuropsychology

General observations

On admission to rehabilitation MP exhibited a range of cognitive impairments the most noticeable of which was the “alien hand” sign (Brion and Jedynak, 1972—see footnote). Her left hand frequently refused to cooperate with the right and in many instances acted in a directly conflicting manner. As her right hand opened a door or drawer the left hand would move to close it. In art therapy her right hand would place a picture onto a collage and the left hand would immediately remove it. Dressing was made difficult because the left hand would persistently undo buttons just done up by the right hand. On another occasion a test involving palpation by the right hand was prevented by the left hand grabbing the right hand at the wrist. Packing a suitcase was very difficult because the left hand would continually remove items packed by the right hand. Sometimes the left hand would act highly disruptively. In one example MP had successfully made a cup of tea when, suddenly, the left hand picked up the teapot and sprayed the other patients. On another occasion she was making an omelette when the left hand threw in a unpeeled onion, an egg still in its shell, and a salt cellar. Her husband also reported the two hands behaving in emotionally inconsistent ways with the right acting affectionately and the left pushing him away. Similarly when MP was carrying out a memory test on which she was not doing very well her outward expression was calm and co-operative. However, at the end of each trial the left hand was picking up each test card and slamming it down on the table. Subjectively MP described her alien hand experiences as like having “two naughty children in my head” who are always arguing. In the early stages it was necessary to physically restrain MP’s left arm in order for her to carry out various routine motor tasks because, on more than one occasion, she was injured by the activities of the alien hand.

A second feature of MP’s disability was a problem initiating and carrying out certain movements. When attempting to leave the lift or shower she was frequently immobilized and required help to pull her out. She also reported days when she was unable to turn right and was forced to make all her journeys continually turning left. She frequently reported urges to jump down flights of stairs or from upper storey windows. In contrast she had no difficulty carrying out simple movements, and swimming, walking, and other well learned movements were unaffected.

Footnote
Bogen (1985 uses the term “alien hand” to refer to a subset of behaviours of the category “inter-manual conflict”. Bogen describes the alien hand as one in which the offending hand “behaves in a way which the patient finds ‘foreign’ ‘alien.’ or at least unco-operative”. This definition appears to encompass most of the aberrant behaviour shown by MP’s left hand and use of the term is also consistent with other accounts of alien hand in the literature. Furthermore one should note that in his account of intermanual conflict as a separate category of deficit, Bogen describes behaviours that could also be considered as alien hand behaviour. Thus, for simplicity the term “alien hand” is preferred in our account of MP’s motor disturbance.
Frontal impairments were also apparent, particularly in tasks that involved sequenced actions—although here the confounding with alien hand symptomology makes interpretation difficult. There were not, however, any signs of confabulation and affect appeared reasonably normal although her husband did note some personality change. Loss of memory was readily apparent in that she was confused about the date and time, would frequently misplace personal items, and lose her way around the rehabilitation centre. However, her memory difficulties did not amount to an amnesic syndrome. She could, for example, remember test stimuli used in test sessions separated by 2 weeks (see below) and rarely showed the repetitive conversation typical of amnesic patients. Her spontaneous recall of past events tended to be rather poor, but, with suitable cueing, MP was often able to remember quite a lot.

**Psychometric Evaluation**

MP’s performance on various psychometric measures is summarized in Table 1. On WAIS-R MP shows evidence of intellectual impairment with a discrepancy of 31 points between her FSIQ and estimated pre-morbid IQ. She is notably poorer on the performance tests. Interpretation of this is difficult because of the confounding with alien hand behaviour. However,

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<th>WAIS-R:</th>
<th>Picture Completion</th>
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<td>Information</td>
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<td>Digit Span</td>
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<td>Vocabulary</td>
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<td>Arithmetic</td>
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<tr>
<td>Comprehension</td>
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<td>Similarities</td>
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<td>Verbal IQ</td>
<td>95</td>
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<td>Performance IQ</td>
<td>76</td>
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<tr>
<td>Full Scale IQ</td>
<td>85 (low average)</td>
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<td>Estimated pre-morbid IQ</td>
<td>116 (high average)</td>
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Wechsler Memory Scale—Revised:

| Verbal Memory Index        | 102                  |
| Visual Memory Index        | 81                   |
| General Memory Index       | 93                   |
| Attention/Concentration    | 92                   |
| Delayed Recall Index       | 62                   |

Wisconsin Card Sorting Test: Untestable

| FAS word fluency            | F 15 (1 perseveration, 1 neologism) |
| A                            | 3 impaired                  |
| S                            | 16 (1 perseveration)        |

Cognitive Estimation: 1 (normal)
observations of MP did not suggest that the alien hand played any major role in determining her performance deficit. On blocks design, for example, she appeared to perform similarly whether or not the alien hand was physically restrained.

On frontal lobe tests, as one might expect, MP showed evidence of considerable impairment. She was completely unable to do the Wisconsin Card Sorting Task (WCST, Nelson modification, Nelson, 1976) and performance on FAS word fluency (Benton et al., 1983) was significantly impaired. However, cognitive estimation (Shallice and Evans, 1978) was normal. This discrepancy most likely reflects the heterogeneity of frontal lobe tests. Leng and Parkin (1988) recently proposed that cognitive estimation deficits are particularly indicative of orbito-frontal damage whereas several studies suggest that WCST is an indicator of dorso-lateral damage to the frontal cortex (e.g. Weinberger et al., 1986). MP's data are reasonably consistent with this position. Her lesion is infero-lateral, i.e. adjacent to dorso-lateral frontal cortex, and might therefore be expected to impair WCST performance more than a test sensitive to orbito-frontal lesions.

On the Wechsler Memory Scale, Revised (WMS-R; Wechsler, 1984) there was a marked discrepancy between her verbal and visual memory indices mirroring that between verbal and performance IQ. Indices of immediate function were in line with a general depressed level of function but delayed memory indicated a substantial impairment.

Additional Evaluation

Astereognosis

MP’s ability to identify objects by touch using both left and right hands was evaluated in two sessions separated by 2 weeks. In session 1 objects were presented inside a cardboard box with a hole in it. On session 2 an apparatus consisting of a plywood board with two holes and a curtain was used. MP was presented with a total of 10 objects to her left hand and 10 objects to her right hand in each session. Only one error occurred with presentation to the right hand. With her left hand MP correctly identified only one object correctly, a metal knife (she identified this correctly on two subsequent occasions but failed to identify a plastic knife). Her errors never bore any resemblance to the object in her hand (e.g. tobacco pipe = “tennis ball”). Analysis of video film showed that her manipulations in her left hand were extremely detailed even though, subjectively, she would sometimes query whether anything was there at all. MP often made use of her reasonably intact memory or common sense to try and guess what the item was (“is it that tennis ball again?”) and this had to be taken into account when designing her test procedures. A second test involved presentation of simple shapes which she then had to match to one of an array of pictures. MP identified all five presented to her right hand correctly but performed at chance when presented to the left hand.
Finger agnosia

This was evaluated briefly by touching each of MP's fingers one at a time while her hand was out of sight and asking her to say which one had been touched. The fingers of the right hand were identified perfectly but, even though she responded immediately to her left hand being touched, she failed to identify the correct finger on any occasion.

Obeying verbal commands

MP was asked to perform six verbal commands using her left hand. Each command contained two elements (e.g. pick up the white triangle and move it so that it is next to the blue square). MP performed only one of these correctly even though she was always able to perform the first component accurately. In contrast she had no difficulty performing this task with her right hand. MP's ability to indicate the answer to five simple arithmetic problems with her left hand was also briefly explored. Each sum was read out and MP was asked to point to the answer on a sheet of numbers in front of her. She achieved one correct response although on one occasion, when the left hand was fixed over an incorrect response, the right hand zoomed in and moved the responses sheet so that the correct answer fell directly under the left hand!

Writing and drawing

Left hand  MP found it extremely difficult to write with her left hand. She was only able to produce her name after a great effort. Figure 1 shows MP's attempt to draw a face left-handed which, on completion, she noted "was not very good".

![Figure 1. MP's attempt to draw a face left handed.](image-url)
Right hand  Figure 2a shows that MP wrote normally with her right hand, and that she was able to draw simple two-dimensional shapes and a clock without difficulty (Fig. 2b,c). However, a cube proved somewhat more difficult (Fig. 2d). Her response to the command “draw a face” (Fig. 2e)
drew a surprising first response followed by a somewhat impressionistic second attempt (Fig. 2f). Her husband considered both responses totally untypical of her premorbidly. Her attempt to draw a building (Fig. 2g) was also peculiar. She explained that it was the type of building shown briefly to link scenes in the TV series Dallas. Her attempt to draw her own house (Fig. 2h) was also unusual in that she seemed unable to incorporate the third dimension.

**Long-term follow up**

MP was interviewed and assessed briefly 14 months after her aneurysm. The alien hand sign had gradually receded during the recovery period but was still manifest to a small degree. When operating the dishwasher, for example, the left hand would counteract the button pushed by the right hand. Some impairment in the initiation of complex sequences of actions was also reported. Left astereognosis was still severe in that only one object out of 10, a ball, was identified with the left hand. Left apraxic difficulties had, however, ameliorated. Writing with the left hand had improved and drawing a face now produced something with a facial configuration. Drawing with the right hand now allowed better incorporation of the third dimension (see Fig 3). The ability to respond to verbal commands with the left hand was greatly improved and MP indicated the correct answer to eight simple arithmetic problems with her left hand.

**Discussion**

MP suffered a ruptured aneurysm of the ACoA accompanied by vasospasm affecting the left anterior cerebral artery. Neuroradiological findings indicated infarction in the genu of the corpus callosum and frontal lobes and a number of distinct neuropsychological sequelae were noted. Stenhouse et al. (in press) have recently emphasized the importance of vasospasm in determining outcome from ruptured ACoA aneurysms and the details of the present case reinforce this point. Evidence of callosal disconnection was manifest by left astereognosis, agraphia, and apraxia, and finger agnosia (see below). In addition there was a prominent alien hand sign, problems with movement initiation, impaired performances on frontal lobe tasks, and substantial memory loss.

Following the classic work of Liepmann (1908) a disconnection account of astereognosis, agraphia, and apraxia of the left hand argues that these deficits all arise from the inability of right hemisphere sensory and motor processes to communicate with left hemisphere language centres (Geschwind and Kaplan, 1962). Thus astereognosis arises from the failed transmission of sensory information from the right to left hemisphere, agraphia from an inability to transmit information about graphic-motor movements from left to right, and apraxia in the same terms by arguing that disconnection prevents the transmission of verbally determined arm movements. However, the fact that MP was able to follow some verbal commands indicates that
this disconnection may not have been total. This can be attributed either to some degree of language function in the right hemisphere or some residual interconnectivity between the hemispheres. The latter interpretation gains support from the anterior nature of MP's lesion. The fibres of the corpus

Fig. 3. MP's left handed writing and attempt to draw a face at 14 month follow up (A and B). Right handed drawing of cube and house at follow up (C and D).
callous cross directly from right to left and, in general terms, centres concerned with sensory and motor programming lie anterior to those concerned with language so there is a possibility that some callosal fibres concerning language functions may have survived. This latter point is also supported by the recovery noted in MP’s ability to respond to verbal commands with the left hand. Moreover, the fact that astereognosis did not remit significantly is consistent with an anterior gradient in MP’s lesion.

Interpretation of the “alien hand sign” is more problematic. One possibility is that these counterproductive arm movements are due to independent control of the left hand by the right hemisphere which, due to callosal disconnection, is now freed from the normal domination by the left hemisphere. Gazzaniga (1970) directly attributed alien hand behaviour to control of the left hand by the right hemisphere:

Case I . . . would sometimes find himself pulling his pants down with one hand and pulling them up with the other. Once, he grabbed his wife with his left hand and shook her violently, while with the right trying to come to his wife’s aid in bringing the left belligerent hand under control. Once, while I was playing horseshoes with the patient in his backyard, he happened to pick up an axe leaning against the house with his left hand. Because it was entirely likely that the more aggressive right hemisphere might be in control, I discreetly left the scene—not wanting to be the victim for the test case of which half-brain does society punish or execute. (Gazzaniga 1970, p. 107)

Attribution of alien hand behaviour to callosal disconnection is complicated by the fact that this sign, unlike the triad of left astereognosis, agraphia and apraxia, is only a persistent feature of callosal patients who have additional extra-callosal lesions (see below). Two possibilities arise: alien hand depends on a conjoint lesion of the corpus callosum and one or more additional structures, or that callosal damage is an incidental correlate of damage to some other structure(s) which itself is directly responsible for alien hand behaviour.

MP’s impairments are consistent with either position because she has extra-callosal damage. A non-callosal account of alien hand can only be unambiguously supported by appearance of alien hand in the absence of callosal damage, or where callosal damage, although present, cannot be invoked to explain the impairment. McNabb et al. (1988) report three right handed patients with dominant anterior cerebral artery infarction who presented with alien hand sign in addition to features of callosal disconnection. The remarkable feature of these cases, and two earlier patients reported by Goldberg et al. (1981), is that alien hand behaviour was executed by the right hand. In terms of disconnection theory the left hand should always exhibit aberrant behaviour because the right hand will remain under the control of the dominant left hemisphere.

To account for their findings McNabb et al. adopted the earlier view of Goldberg et al. by arguing that the alien hand sign was entirely non-callosal in origin and that the likely cause of alien hand was damage to the supplementary motor area (SMA). The SMA is located in the mesial frontal cortex and its function has been the subject of a number of investigations.
Studies in non-human primates have indicated that SMA neurons become active during movement preparation and the execution of some movements (Brinkman and Porter, 1979; Tanji and Kurata, 1982). In humans damage to the SMA has resulted in an initial aphasic and motor disturbance which ameliorates to one affecting only alternating movements of the hands (Laplane et al., 1977; Schell et al., 1986; Nagafuchi et al., 1991) and an impairment in the initiation of voluntary movement (Damasio and Van Hoesen, 1980).

Nonetheless, it remains the case that the alien hand sign has not been observed in any patient in which callosal damage can be ruled out. Thus in studies of left alien hand callosal plus extra-callosal damage are both present (e.g. Brion and Jedynak, 1972; Barbizet et al., 1978; Beukelman et al., 1980; Watson and Heilman, 1983; Levin et al., 1987). Goldberg et al. (1981) account for the temporary left-sided alien hand in “pure” commissurotomy patients as a consequence of their operation which involved retraction of the non-dominant hemisphere and possible temporary disruption of the right SMA. However, it remains the case that these patients also had callosal disconnection and this may therefore have been critical in determining the appearance of alien hand. Returning to our case, MP, her impairments in initiating movement and immediate post-operative course during which total aphasia was observed, are consistent with SMA damage. In relation to this we assessed MP on the alternating hand movement task of Laplane et al. (1977) which is considered indicative of SMA damage. She performed this normally but, because this test was done much later (Nov. 1990), and at a time when her alien hand and movement problems had also ameliorated somewhat, interpretation of this normal performance is ambiguous.

Turning to instances of right alien hand, the only patient of McNabb et al.’s series with radiological evidence had a callosal lesion. Goldberg et al. only had positive evidence for left mesio-frontal damage in their two patients but the authors concede that “damage to the corpus callosum cannot be ruled out in the two cases reported” (p. 685). The most likely conclusion, therefore, is that the alien hand sign requires callosal damage plus extra-callosal damage involving the SMA. It is known that the SMA is bilaterally activated during response initiation in man (Roland et al., 1980). Integrity of the corpus callosum may therefore be essential for normal SMA function. However, the manner in which this conjoint lesion determines the exact pattern of alien hand behaviour has yet to be explained.

Before leaving the issue of SMA function we should also note that the SMA may also be implicated in skilled motor acts. Watson et al. (1986) report two patients with left mesial infarctions involving the SMA in which bi-lateral ideomotor apraxia was a primary feature. It is therefore possible that MP’s apraxic difficulties with both hands may have been exacerbated by left SMA damage although the fact that bilateral deficits were not present could undermine this interpretation.

MP’s memory impairment was disabling but not so severe as to warrant the description amnesia. This is in accord with the general level of reports concerning memory loss following ACoA aneurysms. Poorer visual memory
is most probably attributed hemispheric disconnection. MP’s drawings indicated a lack of spatial information which can be attributed to an inability to transmit right hemisphere based spatial knowledge to the left hemisphere. This problem could also undermine visual memory tasks on WMS-R because these have a strong spatial component and, because MP was responding right-handed, this information would not have been available. A specific deficicy in the transmission of spatial information might also account for some of MP’s marked performance IQ deficit but here the possibility of deficits stemming from the frontal lesion cannot be ruled out.

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References


ALIEN HAND


