CASE REPORT

Complete abolition of reading and writing ability with a third ventricle colloid cyst: implications for surgical intervention and proposed neural substrates of visual recognition and visual imaging ability

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SUMMARY

We report a rare case of a patient unable to read (alexia) and write (agraphia) after a mild head injury. He had preserved speech and comprehension, could spell aloud, identify words spelt aloud and copy letter features. He was unable to visualise letters but showed no problems with digits. Neuropsychological testing revealed general visual memory, processing speed and imaging deficits. Imaging data revealed an 8 mm colloid cyst of the third ventricle that splayed the fornix. Little is known about functions mediated by fornical connectivity, but this region is thought to contribute to memory recall. Other regions thought to mediate letter recognition and letter imagery, visual word form area and visual pathways were intact. We remediated reading and writing by multimodal letter retraining. The study raises issues about the neural substrates of reading, role of fornical tracts to selective memory in the absence of other pathology, and effective remediation strategies for selective functional deficits.

BACKGROUND

The current case is rare in the presentation of global incapacity to read and write with no language disruption or evidence of gross pathology. We are not aware of other reported cases presenting with *global* alexia and agraphia. Patients typically present with partial alexia/agraphia and show substantial lesions to the occipitoparietal regions and/or visual pathways (cf Déjérine¹). In addition, most reported cases of colloid cysts are postsurgery because the cyst has often grown relatively large by the time the patient presents to medical services, increasing the intracranial pressure, compressing the neural structures and requiring surgical intervention. Hence, almost all the cases reported in the literature, to our knowledge, show secondary effects on neural structures beyond displacement of the fornices due to the cyst size and location or as a result of surgery, or a combination of both. These may include: fornix transection, callosal tract disconnection, frontal atrophy, mamillary body atrophy, ventricular enlargement, hippocampal atrophy and evidence of gliosis.^{2 3} Thus, it is difficult to extrapolate the precise function of fornical tracts and surrounding structures from earlier postsurgical documented cases, and there are scant reported data revealing how a cyst abutting the fornix in an otherwise healthy brain might disrupt cognition. The present case

demonstrates severe memory dysfunction possibly as a consequence of white (fornical) matter tract disruption. Arguably, the nature of the deficit underlying memory, reading and writing deficits is visual (visual letter recognition and visual imagery ability). The findings challenge the assumptions that a specific visual word form area (VWFA) in the brain governs reading and writing ability. Thus, the case is medically rare, exceptional in the functional/clinical presentation and challenges current hypotheses regarding the neural substrates of reading/writing ability.

CASE PRESENTATION

The patient was aged 18 when he sustained a minor traumatic brain injury and neck injury in a road traffic accident. There was a post-traumatic amnesia of at least 1 h. Accident and emergency records show that he presented with neck injury and no loss of consciousness, although there was a period of confusion lasting up to 5 days following the injury where he reported loss of episodic recall for events. He was diagnosed with whiplash injuries. Imaging data showed a colloid cyst to the third ventricle abutting the fornix.

The patient reported that he had become progressively illiterate over a period of 2 months post-accident; he first became aware of it when letters on TV appeared alien 'like foreign symbols'. On referral, he reported with an inability to read or write (although he was able to draw), memory deficits, headaches, irritation and excessive levels of fatigue on exertion. There were no clinical signs of receptive or expressive dysphasia, word finding or comprehension deficits based on screening subtests from the Test for Reception of Grammar (TROG⁴ and Psycholinguistic Assessments of Language Processing in Aphasia (PALPA⁵); social use of language appeared normal. He presented with a profound inability to recognise, image or write letters based on free recall. He was unable to recognise or write any letter (with the exception of O and X, which he recognised as mathematical symbols) or words on the initial interview. When words were spelt aloud, he could correctly identify the word. He was unable to sign his name or write his address but could spell his name and address verbally. When asked to read a registration plate, he could only read the numbers. He performed arithmetic sums presented visually. Some difficulties

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were noted with the spelling of exception words (eg, sieve, dove), and correctly identifying irregular words spelt aloud on a letter-by-letter basis ('sow' interpreted as 'stitch' ie, 'sew'). He repeated letters aloud phonetically and commented that he could not 'see' the letter or word 'in his mind's eye'. He could recite the alphabet slowly forwards and backwards.

INVESTIGATIONS

The patient completed a battery of measures of intelligence, memory, executive function and mood state to establish his functional profile of deficit and sparing. Test results are presented in table 1.

Table 1 Scores of the patient on neuropsychological tests of intelligence, attention and memory

Function tested	Score: centile
Intelligence (WAIS III ⁶)	
Full scale IQ	107 (68%)
Verbal IQ	110 (75%)
Performance IQ	99 (47%)
Verbal Comprehension Index	120 (91%)
Perceptual Organisation Index	109 (73%)
Working Memory Index	95 (37%)
Processing Speed Index	73 (4%)
Visuospatial perception (VOSP ⁷)	
Screening test	19/20
Silhouettes	26/30
Dot counting	10/10
Position discrimination	20/20
Number location	9/10
Cube analysis	10/10
Visual and auditory attention (TEA ⁸)	
Map search	
Symbols circled, 1 min	27 (1–5th)
Symbols circled, 2 min	45 (10–25th)
Elevator counting	7 (100)
Elevator counting with distraction	10 (75th)
AMIPB ⁹	
Verbal list learning total A1–A5	55 (25th)
Distractor list	6 (50th)
Target list after distractor	12 (25–50th)
Story recall immediate	31 (10–25th)
Delayed	17 (2.5–10th)
Retained	55% (<2.5th)
Figure recall copy	80 (75th)
Immediate	42 (<2.5th)
Delayed	28 (<2.5th)
Retained	66% (<2.5th)
Visual/verbal recall and recognition (Doors and People ¹⁰)	
Doors	18 (5th)
Shapes	13 (1–5th)
Verbal forgetting scores	36 (>90th)
Visual forgetting score	2 (25th)
	1 (50th)

AMIPB, Attention and Memory Information Processing Battery; TEA, Test of Everyday Attention; VOSP, Visual Object and Space Perception Battery; WAIS III, Wechsler Adult Intelligence Scale III.

Interpretation of test performance: general intelligence (Wechsler Adult Intelligence Scale III)

The patient's general intellectual functioning scores fell within the average range for full-scale, performance and verbal IQ indices.⁶ Conversion analyses of verbal and performance subtest scaled scores showed that digit span (SS=6), a verbal subtest, and digit-symbol coding, a performance subtest score (SS=5), were statistically significantly lower ($p=0.05$) than the mean score for verbal ($m=11.8$) and performance ($m=9$) subtests overall. Although the digit span subtest is verbally mediated and the digit-symbol is performance based, they require manipulation of abstract symbols, which may explain his significantly poorer ability on these measures compared with other subtests. The vocabulary (SS=17), picture completion (SS=15) and matrix reasoning (SS=13) scaled scores were statistically higher than the overall verbal and performance means indicating excellent knowledge of the meaning of words, holistic visual processing and problem solving with abstract visual symbols that do not have a visual structure similar to letters.

The score for the Verbal Comprehension Index fell within the high average range, whereas that for the Perceptual Organisation Index was borderline, with the Working Memory and Processing Speed Index scores ranging from extremely low to impaired. The Verbal (110) and Performance IQ (99) scores were significantly different ($p=0.05$) showing poorer ability on performance compared with verbal tasks. The Verbal Comprehension Index (120) score was significantly higher ($p=0.05$) than the Perceptual Organisation Index (109), Working Memory Index (95) and Processing Speed Index scores (75). The Processing Speed Index score was significantly lower than the Working Memory Index score, and both were significantly lower than the other index scores ($p=0.05$). His core profile indicated a general working memory, information processing speed and perceptual organisation weaknesses compared with verbal ability.

Visuospatial perception: Visual Object and Space Perception Battery (VOSP)

The patient showed intact visual perceptual and visuospatial abilities on subtests indicating that the low-level perceptuospatial deficits were not driving poor scores on the other measures.⁷

Visual and auditory attention: the Test of Everyday Attention (TEA)

He showed impaired selective visual attention in the first minute of the task.⁸ Performance improved in the second minute, falling within the borderline impaired range. He performed within normal ranges on auditory attentional subtests (elevator counting task).

Attention and visual and verbal memory: the Attention and Memory Information Processing Battery (AMIPB)

He performed within the low average range for immediate verbal recall for the story recall subtest of episodic memory, and at borderline to impaired range for immediate recall for the figure recall subtest.⁹ He showed impaired delayed recall for verbal and visual information and an impaired level of retention over time for verbal and visual material on the story recall and figure recall subtests (figure 1).

Figure recall drawings revealed poor retention of visual information from immediate copy with picture present to immediate and delayed copies without target stimulus present for figural features of the image indicating severe disruption to visual memory.

Figure 1 (A) Figure recall from Attention and Memory Information Processing Battery (AMIPB): copy with picture present. (B) Immediate copy with picture present. (C) Delayed copy without original picture.

Verbal and non-verbal visual recognition and recall: the Doors and People test

He performed at the impaired range on the Doors test, a non-verbal visual recognition task, and the People test, a task requiring acquisition of the name of four face stimuli across three trials.¹⁰ He performed at ceiling on the Shapes test requiring four simple shapes to be copied and reproduced after a delay of several seconds, which was in contrast to impaired figure recall performance shown on the AMIPB.⁹

To summarise, performance on the intelligence and memory measures revealed impaired information processing speed often seen after a head injury. He also showed an impaired working memory, poor perceptual organisation and performance-based IQ compared with verbal ability, impaired visual attention, impaired retention of complex verbal and visual material over time, impaired immediate retention of visual material, impaired ability to recognise visual stimuli amid distractors after a brief delay, impaired ability to match visual stimuli (faces) with corresponding verbal labels and intact ability to copy and remember simple visual shapes. Scores on the Hospital Anxiety and Depression scale (HADS¹¹) reflected a mild degree of anxiety and depressive symptoms in the preceding week.

Imaging data

Images were obtained on a 3 Tesla Philips Achieva scanner (Philips Medical Systems, Best, The Netherlands). The sequences obtained were axial and coronal TSE T2 (TR 2067, TE 80); axial and coronal proton density (TR 2067, TE 17.78); T1 volume (TR 10.18, TE 4.691, flip angle 8, 0.8 mm isotropic voxel size). The skull base demonstrated some congenital asymmetry with underdevelopment of the right petrous temporal bone. As a result of this, there was some asymmetry in the shape of the overlying temporal lobes. The brain regions were otherwise normal. There was no evidence of any visible contusional damage to the lingual (medial occipitotemporal) gyrus or the fusiform (lateral occipitotemporal) gyrus on either side. Other areas commonly affected by contusional damage were also normal. Gradient echo and susceptibility weighted imaging are very sensitive to the presence of haemosiderin, a lasting product of intraparenchymal haemorrhage. There was no evidence of any haemosiderin staining in any region of the brain. The only abnormality was the presence of a small (8 mm maximal diameter) colloid cyst at a typical location at the anterior end of the third ventricle. This was not obstructing the foramina of Monro and there was no lateral ventricular

Figure 2 MRI showing the location of a colloid cyst in the third ventricle in the coronal, sagittal and axial planes.

dilation; however, the cyst splayed the columns of the fornix (figures 2 and 3).

TREATMENT

The therapy intervention approaches were designed as follows: the rote rehearsal method required him to study each randomly selected target letter by concentrating on the shape of the letter and any distinctive figural features. The speech therapist presented the letter and enunciated the sound of the letter three times for approximately 1 s/letter to reinforce learning. He completed both types of training within each session in a counterbalanced order over 10 sessions conducted over a 3-week period. For the multimodal procedure, he was instructed to study the letter in detail and to think of an alternative stimulus beginning with the letter that he associated with the letter shape, for example, 'm' reminded him of mountains because of the up and down strokes, 's' reminded him of a snake because of its curved shape and 'e' reminded him of an eye. This strategy created a phonemic link between the letter and the personal mnemonic prompt to enhance relevance and optimise learning. We also devised verbal descriptions of letter features, for example, 'c' described as an incomplete circle with a section missing on the right hand side, 'l' described as a long vertical stick. The letters were reproduced in enlarged format. We asked him to trace the shape of the letter while the therapist described visual features of the letter and reminded him of the personal mnemonic prompt he had generated for the letter. On the third presentation, he was asked to generate a word beginning with the chosen letter (different from the mnemonic word generated earlier to aid recognition) to strengthen semantic word–letter associations. In this way, multisensory (visual, auditory and tactile/kinetic) associations were made for each letter. We anticipated that this might prove more effective than the rote rehearsal method because that relies predominantly on visual/phonemic integration alone, while the multimodal method synthesised multisensory information to enhance learning and depended on mnemonic cues specifically generated by and personal to him.

Figure 4 shows the accuracy of the ability to select target letters to a verbal prompt, and to recognise and name target letters across 10 therapy sessions in the rote rehearsal intervention. The letter 'i' was recognised as familiar and named correctly after 1 s following 10 treatment sessions of the rote rehearsal method. Although four letters of this set (w, p, q and i) were considered to be familiar to him, 'n', 'v' and 'w', and 'p' and 'q' were regularly confused. The average time to name each letter on session 10 was 5 s, although only one response was accurate.

Figure 5 shows accuracy scores for letter selection familiarity and naming across 10 sessions using the multimodal procedure. Plotted data show that the complete letter set was identified, named and rated as familiar by him following the 10 treatment sessions. The mean response time for letter naming was 5 s/letter, the same as for the rote rehearsal set, possibly indicating that although the multimodal letters had been learnt, identification was not automatic as with fluent reading. However, results of the initial intervention suggested that a multimodal approach to letter learning was more effective than rote learning in facilitating relearning of letters and this approach was used for the 12 letters of the alphabet not yet studied in the second treatment block.

OUTCOME AND FOLLOW-UP

Results graphed in figure 6 show that the 12 remaining letters of the alphabet were remediated after 10 additional therapeutic sessions using the multimodal intervention for letter relearning.

DISCUSSION

Complete inability to recognise a single letter of the alphabet is rarely seen in alexia cases and when it does occur, the ability to write usually remains intact.¹ Abstract representation of visual letters/words has been termed the 'visual word form'.^{12 13} The assumption is that recognition of the 'word form' enables fluent reading to occur across font, case, colour and size.^{12 13} The term *agraphia* denotes individuals who have lost the capacity to write with reading ability generally intact; such cases are rare although more frequently documented than combined alexia

Figure 3 Intact left visual word form area region of interest in the coronal, sagittal and axial planes; imaging data show an intact mid-fusiform gyrus on the left with the region of interest identified by cross hairs.

Figure 4 Results of rote rehearsal letter remediation intervention, familiarity, selection and letter naming scores. Target letters: n, p, v, q, i and w.

and agraphia in the same individual as reported here.¹⁴ Miozzo and Caramazza¹⁵ reported the case of an 84-year-old woman who sustained a large left posterior brain lesion. She was impaired at reading letters and numbers, and naming pictures, objects and colours. However, performance accuracy for naming tactile stimuli was at ceiling, indicating intact access to non-visual word information. Agraphia indicates impaired writing when the limb movements are intact.¹⁴ This type of impairment is associated with pathology affecting a region in the left parietal cortex designated the VWFA, and thought to mediate letter recognition and letter imagery ability.^{16 17}

The fornix is the major fibre tract connecting the hippocampal formation to the basal forebrain and medial temporal regions associated with recall and episodic memory.^{18 19} Cysts of the third ventricle abutting the fornix are often undetected until their presence causes increased intracranial pressure resulting in dilation of ventricles and compacting of surrounding structures. Some authors report that ablation of the anterior column of the fornix does not induce memory dysfunction.²⁰ Others suggest that removal of a colloid cyst may cause anterograde amnesia and systematic disruption to episodic recall, although memory deficits are also seen prior to cyst removal.² Poreh *et al*¹⁸ reported impaired delayed visual recall on the Rey complex figure test (similar to the figure recall task used here) and severe visual and verbal memory deficits postfornicectomy (see also Vann *et al*²¹).

The current study presents the rare case of a patient who exhibited global alexia and agraphia after a mild head injury. Structural imaging data revealed a third ventricular colloid cyst abutting the fornix sufficiently to displace the fornical columns. There was no additional pathology or ventricular dilation. The

presence of the cyst may be incidental to his clinical presentation, but this seems unlikely; the slow emergence of functional deficits over several weeks postinjury suggests the unfolding of a pathological process. It is possible that the mild head injury was a catalyst for the development of the cyst, but we have no way of confirming whether it was present *before* the mild head injury.²² Neuropsychological testing revealed general widespread memory deficits across visual and verbal domains (worse for visual stimuli) that disproportionately impeded reading and writing abilities, slowed processing speed, and impoverished story and (complex) figure recall. Arguably, his visual memory problems took the form of a general visual imaging and visual processing deficit that abolished visual invariance (ie, the stable capacity to recognise letters across different fonts and viewpoints, etc). Several researchers have theorised that clinical features of alexia and agraphia are subtended by visual imaging deficits or a general visual processing deficit, although the mechanism is thought to be different for each condition.^{12 14-16} We are not aware of any theory proposing that the same structural impairment might drive both conditions post neuropathology, possibly reflecting few reported cases of complete alexia and agraphia in the same individual. He reported that presently, several years after the accident, he is only able to write cursively (typing is relatively fluent) by imaging letters that he was trained on. He claims that he is unable to vary the visual representation of the training letter by changing the font, colour or size of the image. He was able to draw a generic house on command possibly by utilising a verbal description of a house, but he was unable to draw *his* house. He appeared to have lost the capacity to 'free image' letters and likely other stimuli (his house) indicated by neuropsychological test results that required a type of visual 'fluency'.

Figure 5 Results of multimodal letter remediation intervention, familiarity, selection and letter naming scores after 10 sessions. Target letters: g, r, s, l, h and b.

Figure 6 Results of multimodal letter remediation intervention for the remaining untreated 12 letters of the alphabet, familiarity, selection and letter naming scores after 10 sessions. Target letters: z, j, e, y, a, k, u, c, f, m, t and d.

To conclude, the distinction between alexia and aphasia at functional and structural levels might be broadly considered an artefact of the conceptualisation of these disorders. Alexia and agraphia cases are seldom reported in a manner that evaluates the potential for shared functional deficits affecting reading and writing. Our case showed general memory impairments on functional tests that predominantly affected visual recognition memory and imaging ability. Our case suggests that there might be some global visual mechanism shared across alexia and agraphia that at present we can only conceptualise as 'free' visual imaging whereby a representation is self-generated rather than exactly reproduced from a previously seen item and that accounts for perceptual invariance in visual recall. The findings raise questions about the purported neural substrates of reading and writing, the general role of fornical connectivity to memory, the importance of visual recognition memory and visual imaging to reading and writing, and the efficacy of multimodal methods in reading retraining.

Learning points

- Displacement of fornical tract connectivity can induce visually mediated functional deficits.
- Gross grey matter lesions are not necessary for reading and writing deficits to be present after head injury.
- Fluent reading and cursive writing abilities may depend on a type of visual 'fluency' for abstract letter manipulation.
- Multimodal rather than rote rehearsal training can remediate profound reading and writing deficits.

Contributors LAB and NM designed the treatment approach implemented in therapy sessions by KG. LAB reviewed the relevant literature and wrote the article in conjunction with NM. CAJR devised the imaging protocol and interpreted the imaging data.

Competing interests None.

Patient consent Obtained.

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