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Reversible cerebral vasoconstriction syndrome

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ABSTRACT



Reversible cerebral vasoconstriction syndrome (RCVS) is a relatively rare, non-progressive angiopathy frequently heralded by severe thunderclap headache. It is characterised by vasoconstriction of cerebral arteries which usually resolves within three months of onset. Transient focal neurological signs may occur, and persistent deficits associated with haemorrhagic comorbidities have been reported in a small percentage of individuals. In this paper we report the case of RH, a 36-year-old woman who presented at a university teaching hospital in Sydney with a clinical history and radiological evidence consistent with RCVS. There were no haemorrhagic events during the course of her illness, and vasoconstriction resolved within a few days, following treatment with verapamil. Neuropsychological evaluation 16 months later revealed significant deficits in autobiographical memory, verbal and non-verbal new learning and active delayed recall, cognitive flexibility, abstraction and (to a lesser extent) immediate attention span and information processing speed. RH's case was unusual because the "trigger" for RCVS (Ear, Nose and Throat surgery) has not been previously reported, and because despite there being no haemorrhagic complications during the course of RCVS and no subsequent radiological abnormalities, she had significant cognitive impairment. To date, persistent neuropsychological deficits have not been recognised as a feature of RCVS.

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KEYWORDS Angiopathy; RCVS; neuropsychological impairment

Introduction

Some neurological conditions are relatively rare but clinicians working in neurology settings of large tertiary referral hospitals would, over the years, have a good chance of coming across at least one individual with the condition. Reversible cerebral vasoconstriction syndrome (RCVS) is such a condition. RH, a woman who was 36 years old when the clinical data described in this paper were collected, is such an individual. In 2014 she was admitted to a major teaching hospital in Sydney, Australia, under one of the authors (GH) with symptoms which eventually led to a diagnosis of RCVS. It was also a rare case because the potential "trigger" for the condition was unusual,

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and cognitive sequelae such as those she demonstrated have not, to our knowledge, been reported in the literature.

Reversible cerebral vasoconstriction syndrome (RCVS)

The most frequent presenting symptom of RCVS is a severe thunderclap headache often followed by recurrent severe thunderclap headaches. The peak of pain can occur within a minute of onset with the pain lasting minutes to days. Nausea, vomiting, sono and photophobia may also occur, and the headache can recur, with an average of four attacks over the next four weeks. Focal neurological deficits can be seen, the most frequent being visual disturbance, but hemiparesis, sensory change, dysarthria and dysphasia are also described. Persistent deficits are rare, and suggest either an ischaemic stroke or haemorrhage rather than RCVS (Mehdi & Hajj-Ali, 2014).

The onset of symptoms may be spontaneous, although the headache is often associated with some exertion, especially sexual activity, bathing, or straining with exercise (Valenca, Andrade-Valenca, Bordini, & Speciali, 2008). RCVS has been described in association with a myriad of medical conditions, but over half occur in the post-partum period (Bogousslavsky, Despland, Regli, & Dubuis, 1989), or in association with serotonergic or adrenergic drugs, including both prescribed (selective serotonin reuptake inhibitors, alpha sympathomimetics, and acute migraine treatments) and recreational substances (cocaine, amphetamines, LSD and marijuana) (Calabrese, Dodick, Schwedt, & Singhal, 2007; Ducros et al., 2007). Other conditions associated with RCVS include migraine, eclampsia and pre-eclampsia (Dukes & Vieth, 1964), and as a result of neurosurgical procedures (Hyde-Rowan, Roessmann, & Drodkey, 1983). The early literature reports of this condition ascribed various other syndromic names including “postpartum angiopathy”, “drug induced angiopathy” and the Call-Fleming syndrome, but it is now generally accepted that the term introduced by Calabrese et al. in 2007 is the preferred descriptor.

The mean age of onset is 42.5 years with a female to male ratio of approximately 2:1 (Ducros et al., 2007; Ducros et al., 2010) but RCVS can also occur in patients in early adolescence (Chen et al., 2010b; Singhal et al., 2011). Prevalence and incidence are unknown, but the condition is reasonably rare and probably remains under-diagnosed (Calic, Cappelen-Smith, & Zagami, 2015; Chen, Fuh, & Wang, 2010a). The latter might be, at least partly, due to the lack of well-standardised diagnostic criteria.

Neurological complications and comorbidities can occur during the acute phase of RCVS, usually within days of symptom onset. Recurring thunderclap headaches are very common, up to 100% in some case series (Singhal et al., 2011). There may be focal neurological signs which are transient in most cases. Persistent (i.e., > 24 hour duration) focal neurological deficits may occur in approximately 30% and 2% of individuals with and without haemorrhagic complications, respectively (Ducros et al., 2010). However, the true prevalence of cerebral stroke/non-aneurysmal haemorrhagic events in these patients is unclear. Low rates of ischaemic stroke (7.8%) are reported in some case series, but no other type of haemorrhagic event (Chen et al., 2010b). By contrast, in the series of 89 patients reported by Ducros et al. (2010), approximately one third had at least one haemorrhagic complication, with chronic subarachnoid haemorrhage being the most common (30%). Variable rates of posterior reversible

encephalopathy syndrome (9.1–38%) have been reported (Chen et al., 2010b; Singhal et al., 2011).

The radiological appearance of RCVS is of vasoconstriction of cerebral arteries with resolution occurring within three months. Calabrese et al. (2007) proposed the first diagnostic criteria, which included clinical findings but also the presence of multifocal segmental cerebral artery vasoconstriction demonstrated via direct (catheter) or indirect angiography.

The course of RCVS is not progressive, and tends to be short. The thunderclap headaches resolve within three weeks but vasoconstriction may persist for up to three months. The majority of patients recover well, and vascular abnormalities reverse spontaneously or following treatment with calcium channel blockers such as nimodipine. There are, however, non-reversible consequences of RCVS, but permanent neurological deficits are fairly rare (3–8%) (Ducros et al., 2010; Chen et al., 2010b). Haemorrhagic complications and stroke occur in 34% of cases (Ducros et al., 2010). Long-term prognosis is poorer for these patients than for those without these complications. At six-month post-acute phase, none of the individuals without haemorrhagic/stroke complications had significant deficits, compared with 27% of those who did. By this time eight patients had not yet returned to work due to persistence of severe deficit or asthenia.

Neuropsychology of RCVS

It is possible that during the acute phase some patients with haemorrhagic complications might be referred for neuropsychological assessment if there is concern about cognitive impairment, such that it becomes a clinical management issue. We have found no information regarding the cognitive status of individuals reported by Ducros et al. (2010) who had haemorrhagic/stroke complications and experienced significant persistent deficit after recovery from the angiopathy. There has been, to date, little attention focused on the neuropsychological sequelae of RCVS. To our knowledge, only one study has specifically documented impairment of cognitive function post-recovery from RCVS (Smigielski, 2014). The case was that of a middle-aged woman who had an episode of RCVS, diagnosed on the basis of biopsy and magnetic resonance angiogram (MRA), and which was triggered by a high altitude ascent. Neuropsychological evaluation 18 months after revealed impairment of perceptual organisation and visuospatial neurocognitive abilities, whereas verbal intellectual, executive and other cognitive functions were unimpaired.

The case of RH

Background

RH is a right-handed woman who was 36 years old at the time of assessment. She was born in Namibia and her mother tongue was Afrikaans but she also spoke English. She was educated in South Africa (six years of primary and five years of secondary school), and had been studying forensic psychology for one year, but discontinued when she applied for residency in Australia. She had been living in Australia since 2012 or 2013 (she was unsure about the date; see below), and worked as an accounts payable officer. She has been married for 11 or 12 years (she was also unsure about this; see below) and had a daughter aged 15 years and a son aged 9 years.

Medical course

At the end of August 2014, RH presented with sudden onset of severe headache associated with nausea, vomiting and neck stiffness, with onset while vomiting. A week before, she had undertaken nasal sinus surgery, and several days before presentation had had a repair of a cerebrospinal fluid leak at the apex of her nose, both procedures under general anaesthesia.

Her past medical history included migraines, which were associated with bifrontal headache and visual aura, but were not as severe as the current headache. She also had a slow transit bowel syndrome, and cervical spondylosis.

On examination, RH was afebrile with normal reflexes, tone, power, coordination and speech, but with a stiff neck and clearly in pain. Initial diagnoses of meningitis or sub-arachnoid haemorrhage were excluded by a lumbar puncture. Cerebral computerised tomography (CT) scan was normal, as was the magnetic resonance imaging (MRI) of the brain parenchyma (Figure 1), but the MRA showed irregularity of the proximal intracranial vessels, raising the possibility of vasculitis.

Treatment with empiric antibiotics, and analgesics (oxycodone, pregabalin, and tramadol) was used as initial management, and corticosteroids (prednisolone 50 mg in reducing doses) was initiated after the MRA suggestion of vasculitis. A cerebral angiogram (Figure 2) confirmed the irregular narrowing of the anterior cerebral and middle cerebral branches (M2), which improved after the administration of 10 mg verapamil intra-arterially (Figure 3). The reversibility of the narrowing suggested the condition was RCVS as opposed to vasculitis, which does not show reversibility. Oral calcium channel blocker (verapamil 40 mg b.d.) was commenced. On discharge home 10 days after admission, the headache was settling, and she had no focal neurological signs. Repeat brain MRI two days before discharge revealed no evidence of haemorrhage/stroke after the resolution of RCVS (Figure 4).

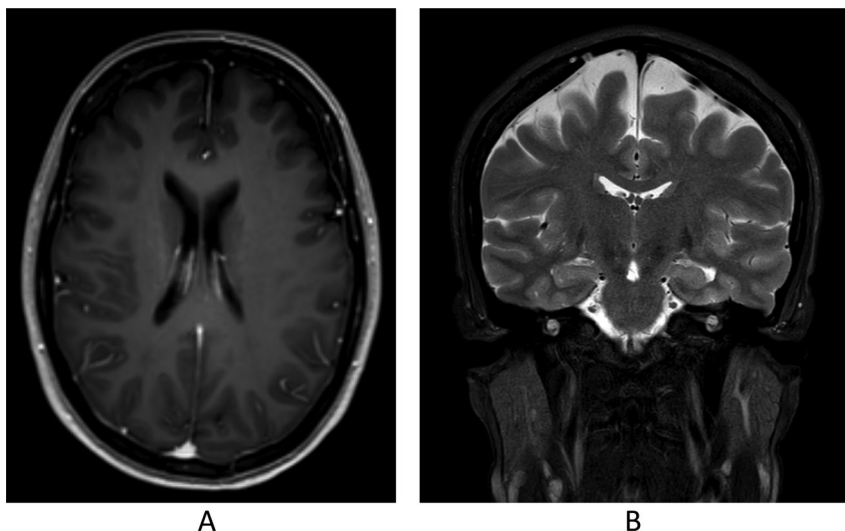


Figure 1. MRI two days after admission. A = axial (gradient echo T1 sequence with gadolinium); B = coronal (T2 spiral).

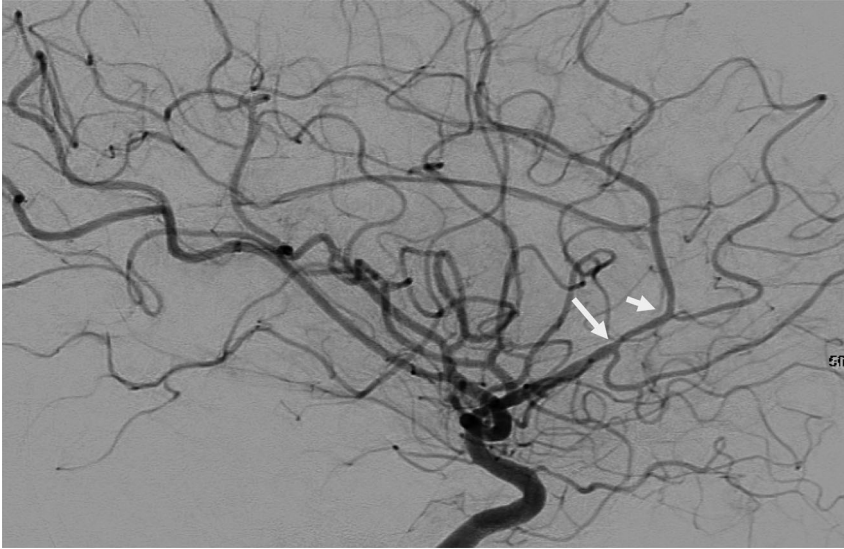


Figure 2. Cerebral angiogram, left internal carotid artery lateral view, pre-verapamil, showing vasoconstriction of the vessels (arrowed).

Several weeks later, RH represented with a flare of her headache, a repeat MRI scan and MR-angiogram showed persisting, but improved, irregularity of the anterior intracranial vessels, but no parenchymal abnormality. The subsequent headache pattern

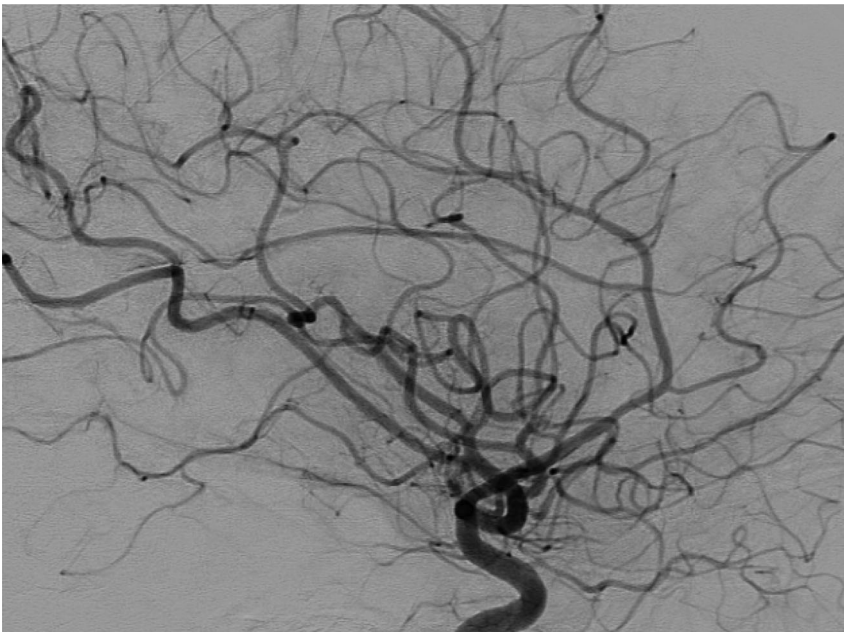


Figure 3. Cerebral angiogram of the left internal carotid artery after the intra-arterial administration of verapamil, showing resolution of the areas of narrowing previously noted in [Figure 2](#).

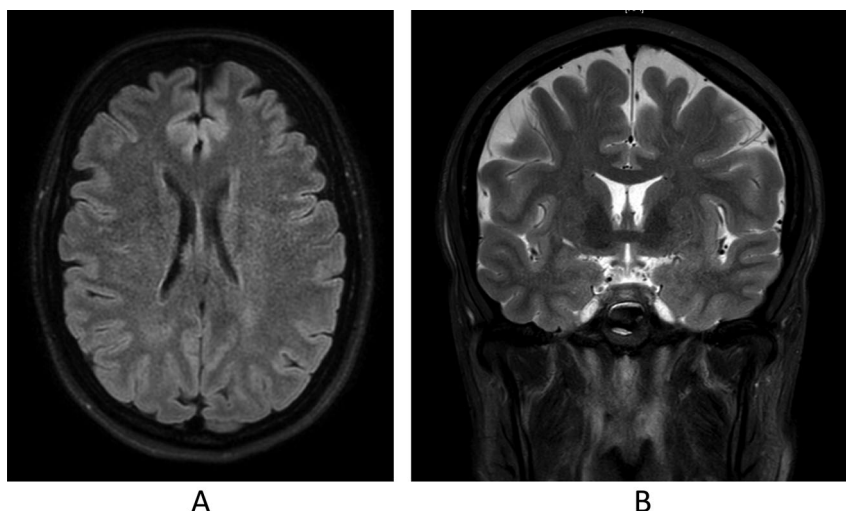


Figure 4. MRI following resolution of vasoconstriction and two days before discharge from hospital. A = axial (3D Flair); B = coronal (T2 spiral).

was of recurrent flares, most of which were similar to her usual migraine pattern. The headaches occurred every 6–8 weeks, were not severe, and required simple analgesia for their management (paracetamol as required). Prior to the neuropsychological assessment, she had been headache free for 8 weeks. Calcium channel blockers were weaned after 6 months as they contributed to her slow transit colonic disorder; beta blockers, amitriptyline and topiramate were trialed but poorly tolerated.

Neuropsychological assessment

Following discharge from hospital after her bout of RCVS, RH was, and continues to be, reviewed frequently (approximately every 3 months) by one of the authors (GH). The focus of these reviews was to monitor her recovery from RCVS, repeat brain MRIs and manage her headaches. She noted cognitive difficulties soon after discharge from hospital and voiced concerns about it at her first follow-up review, two months later. Because she continued to report concerns with cognition, she was subsequently referred for neuropsychological assessment in February 2016, 16 months after the diagnosis of RCVS. Up until the time of her assessment, RH remained in good health, brain imaging consistently demonstrated no additional acute intracranial pathology, no de novo neurological conditions were evident and her headaches were well controlled with paracetamol.

After discharge from hospital RH had continued working full-time, looking after her husband and two young children, and moved house in the first year. Soon after discharge, she began to notice increasing difficulty with memory, whereas she reported that she used to have a photographic memory. For instance, RH was an avid reader, and she was having difficulty remembering what she had read by the time she reached the bottom of a page. When she resumed reading a book, she had difficulty remembering where the story was up to when she had last read the book (usually the previous day), and had to re-read the last few pages to refresh her memory.

She also had difficulty remembering the names of and details about the characters in a book and details of the story line soon after finishing the book. She had never experienced these difficulties before. She also had difficulty recalling recent events. For instance, the week prior to the assessment, she had been on a cruise, but could not recall details of what had occurred.

Apart from problems with anterograde memory, RH also reported that she was having difficulties with memory for remote events, and had lost many autobiographical memories across her entire lifespan. For instance, she was able to recall (but was unsure) the day, month and year of her wedding, could recall (albeit with some difficulty) that it had taken place in a small church on a hot day, but could not recall who was there, what was on the menu, what her wedding song was, and so forth. Similarly, she had great difficulty recalling (or was uncertain about) when she learned to speak English, when she finished high school, or when she emigrated to Australia. When asked by her children about past events in their lives, RH often could not recall them. She felt she could not engage with her children the way she wanted to and was very distressed by this. She also felt that she had been very “moody and emotional” since her illness in 2014, with her mood fluctuating and cycling between happy, sad and angry.

In addition to her memory problems, RH reported that she was finding it difficult to perform multiple tasks, and this was having a negative impact on her work performance. She had had long-standing plans, prior to the episode of RCVS, to study criminology at university and not long before her neuropsychological assessment she had enrolled in a course. She had also intended to continue with her usual employment as well as look after her family. She was very keen to “get my brain back” and to do this course but feared and felt that she would not be able to cope.

The results of RH’s neuropsychological assessment are shown in [Table 1](#). Her interactions during the assessment were appropriate, and she was cooperative throughout the session, engaging adequately in and exerting suitable effort on all tasks administered. She was orientated to time, place and person, could not name the current Prime Minister, but could name the previous one.

Surprisingly, RH’s estimated premorbid intellectual level (as measured by the Shipley Institute of Living Verbal Scale) was at the 8th percentile, which is well below average ability. This might not have been an accurate reflection of RH’s “true” cognitive level but, arguably, a test-related artefact. The Shipley is, in essence, a vocabulary test. On the one hand, despite RH’s apparent good command of English, her vocabulary may be less extensive than in her mother tongue (Afrikaans); hence her poor performance.

Alternatively, RH’s poor performance on the Shipley might have indicated that she was losing knowledge of word meanings. Her expressive language was fluent and free of paraphasic errors or neologisms. Yet, semantic fluency (Animal Naming) was borderline impaired which suggested that access to semantic knowledge store might be impaired, or that the “word knowledge store” had degraded. This, in conjunction with RH’s extensive impairment of autobiographical memory, points to such a possibility. This would also contribute to her very poor performance on the Shipley. Moreover, her performance on cognitive domains that were spared (e.g., planning) also suggested a higher level of ability than the 8th percentile. Basic reading and basic writing skills were spared.

RH did not impress as being depressed or emotionally blunted; her mood was reactive and impressed as euthymic. On a self-rated screening measure of mood and affect

Table 1. Summary of RH's neuropsychological test results.

Test	Score	%ile	Test	Score	%ile
Shipley Institute of Living Verbal Scale	23	8	COWAT Adjusted Total Score	24	4-5
			Category Naming Animals	13	<10
DASS					
Depression	1	n.a.d.	Similarities (WAIS-III) Age SS	5	5
Anxiety	1	n.a.d.			
Stress	8	n.a.d.	Tower of London		
			Test Total Correct	6	68
			Total Move Score	10	84
Orientation (WMS-R) Total Score	13	21	Total Initiation Time (sec)	89	85
Mental Control (WMS-R) Total Score	14	2.3	Total Execution Time (sec)	242	26
			Total Time (sec)	251	42
Symbol-Digit Modalities Test (SDMT)			Total Time Violations	0	66
Oral	39	7	Total Rule Violations	0	55
Written	36	1-2			
Digit Span (WAIS-III) Age SS	4	2			
Maximum Forward Span	3	<1	Wisconsin Card Sorting Test – 64 (WCST)		
Maximum Backward Span	3	4	Categories	2	11-16
			Trials to Complete 1 st Category	27	6-10
			Failure to Maintain set	0	–
Hopkins Verbal Learning Test (HVLТ)			Learning to learn	–4.63	>16
Trial 1	5	8	Total Number Correct	35	–
Trial 2	6	2	Total Number of Errors	29	5
Trial 3	8	2	Perseverative Responses	15	12
Σ1-3	19	2	Perseverative errors	14	9
Delayed Recall ²	6	3	Non Perseverative Errors	15	8
Recognition Hits	6	–	% Conceptual level responses	24	3
Recognition False Positives	0	–			
% retention	75	12			
Brief Visuospatial Memory Test (BVSMT)					
Trial 1	3	4	Stroop Test Adjusted Word Score	68	2
Trial 2	5	1	Adjusted Colour Score	48	1-2
Trial 3	7	1	Adjusted Colour-Word Score	24	1-2
Total Recall	15	1	Interference Score	–4.14	27-35
Learning	4	54	Judgement (NAB) Total Score	12	8
Delayed Recall	3	<1			
Precent retained	57	<1			
Recognition Hits	2	<1			
Recognition False Alarms	0	>16			
Recognition Discrimination Index	2	>16			
Recognition Response Bias	0.10	>16			

DAAS = Depression, Anxiety and Stress Scale; WMS-R = Wechsler Memory Scale Revised; WAIS-III = Wechsler Adult Intelligence Scale III; COWAT = Controlled Oral Word Association Test; NAB = Neuropsychological Assessment Battery; n.a.d. = no abnormality detected.

Bold type denotes poor or impaired performance.

(Depression, Anxiety and Stress Scale) she did not endorse symptoms suggestive of abnormality, despite having reported at initial interview that she had been experiencing marked mood and emotional fluctuations.

There was mild-to-moderate impairment of information processing speed, although processing accuracy was unimpaired (Digit-Symbol Modalities Test). RH's slow processing speed was also reflected in all but the interference score of the Stroop Test.

Some of RH's poorest performances were on measures of new learning. She demonstrated a modest learning curve for verbal and visuospatial information but her acquisition capacity was severely impaired in both instances (Hopkin's Verbal

Learning Test: Σ Trial1–3 = 2nd %ile; Brief Visuo-Spatial Memory Test: Σ Trial1–3 = 1st %ile). Active recall after a 20–25 minute delay was markedly impaired (particularly for visuospatial information). RH's impaired performance on recognition memory measures for either type of information suggested an underlying encoding deficit. RH's performance in the memory domain strongly suggested bilateral temporal lobe dysfunction.

RH's scores on the Tower of London Test (ToLT) reflected her highest performance on any of the tests administered. RH was able to solve just over half of the test items in the required number of moves (ToLT Correct score = 68th %ile). There was no evidence on other measures derived from the test that RH's ability to plan and to generate and successfully implement problem-solving strategies was impaired. It is worth noting that Execution time, whilst in the normal range for her age-group, was RH's lowest performance on the test. This might have reflected her relatively mild speed of processing deficit.

Her ability to inhibit familiar/automatic but task inappropriate responses was also unimpaired (Stroop Test Interference Score = 27–35th %ile). By contrast conceptual flexibility was notably impaired. RH's ability to formulate conceptual set was borderline impaired. She sorted only two categories on the Wisconsin Card-Sorting Test (WCST), a performance bordering the impairment range for her age group. She required more than average number of trials to sort the first category, and had difficulty formulating the sorting concept (WCST Conceptual-level responses = 3rd %ile). Subtle perseveration was also evident. Her ability to formulate verbal abstract concepts was mildly impaired (Similarities: Age SS = 5th %ile) and judgement was borderline impaired (Neuropsychological Assessment Battery: Judgement = 8th %ile).

Discussion

In the context of a relatively rare neurological syndrome, RH's case is even more unusual for two reasons. First, ENT surgery is an unusual trigger for the disorder, indeed we could find no previous references to this in the literature. Second, although she has otherwise recovered well from RCVS without haemorrhagic complications, she has significant cognitive impairment in the absence of either radiological abnormalities or severe, persistent functional deficit.

Although clearly occurring in the context of RCVS, the precise aetiology of RH's cognitive deficits is unclear. It is possible that they are a direct, permanent consequence of the vasoconstriction that had cleared over a year prior to her assessment, given that she experienced cognitive difficulties immediately after the episode of RCVS. Moreover, RH had: (1) relatively mild impairment of immediate attention span and information processing speed; (2) selective impairment of executive functions; (3) severe, non-material-specific impairment of new learning and active delayed recall; (4) significant autobiographical memory deficit; (5) possible impairment of semantic memory. Such a neuropsychological profile suggests both frontal lobe and bilateral temporal lobe dysfunction. This is congruent with the location of the segmental vasoconstriction shown on RH's MRA, that is, anterior cerebral and Sylvian segment of the middle cerebral artery.

However, if there had instead been predominant involvement of the posterior vessels and/or posterior reversible encephalopathy syndrome (PRES) had been a complication, a different pattern of cognitive deficits might have emerged. The case

reported by Smigielski (2014) tenuously supports this notion. The impairment reported in that case (perceptual organisation and visuospatial functions, but not verbal intellectual, executive and other cognitive functions) suggests involvement of posterior cerebral areas. Memory was not specifically mentioned by Smigielski, and there was no information about the specific location of vasoconstriction or whether or not PRES was a complication, but the study was reported in an abstract and, unfortunately, no further detail was provided.

Admittedly, RH's visuospatial and perceptual organisation functions were not examined, so there might have been impairment in this domain. Notwithstanding, RH's neuropsychological deficits on testing were congruent with her reported difficulties with recollection of events and information in day-to-day life, and difficulties multitasking at work.

If RH's cognitive deficits are associated with, but not directly caused by, vasoconstriction, their neuropathological basis is unclear. Brain MRI showed no radiological evidence of haemorrhage/stroke at the onset of RCVS or immediately after its resolution. Moreover, since RH's recovery from RCVS there has been no radiological evidence of further cerebral pathology or *de novo* neurological conditions that could cause her neuropsychological impairment. Furthermore, following the episode of RCVS, RH's headaches had been relatively mild and controlled by simple analgesia, and prior to the neuropsychological assessment she had been headache free for eight weeks. Migraine would, therefore, be an unlikely factor influencing her test performance. Besides, prior to the RCVS she had a long-standing history of migraines (which were more severe than the headaches she had post-RCVS) and cognitive difficulties had never been an issue then.

It is also extremely unlikely that RH's cognitive deficits can be attributed to the cerebrospinal fluid (CSF) leak which occurred after she underwent nasal sinus surgery. First, the CSF leak occurred and was repaired prior to the onset of RCVS, so it was no longer active or an issue. Second, cognitive deficits following a CSF leak are unlikely unless there has been subsequent infection of the CSF and/or meninges. This did not happen in RH's case.

RH's case may be exceptional and neuropsychological deficit may be a rare long-term outcome of what has been considered to be good recovery from RCVS without haemorrhagic/stroke complications. On the other hand, RH's cognitive impairment might be a salient instance of a deficit that might be more prevalent than previously suspected in these patients. We have found only one other case reported in the literature (Smigielski, 2014). The question then is whether or not RH's cognitive deficits represent a characteristic pattern of impairment. Possibly not.

RH resumed full-time work and continued to look after her family after leaving hospital and has continued to meet the challenges that her cognitive deficits impose on day-to-day life with determination. She became aware of her cognitive difficulties soon after discharge, and has felt that there has been no deterioration over time. We hope to be able to review her within approximately one year after her first assessment in order to investigate this possibility. At the time of writing RH had plans to enrol full-time, in a criminology course at a university in Sydney, but had deferred doing so after the neuropsychological assessment. She had also planned to continue working full-time and look after her family while she studied. At a subsequent recent review with one of the authors (GH), RH was counselled to consider part-time studies when she took up her enrolment, which she agreed to do.

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Disclosure statement

No potential conflict of interest was reported by the authors.

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