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To cite this article: Mohammed Fadelalla, Avinash Kanodia, Mustafa Elsheikh, John Ellis, Vivien Smith & Kismet Hossain-Ibrahim (2019): A case of aneurysmal subarchnoid haemorrhage and superficial siderosis complicated by prospagnosia, simultagnosia and alexia without agraphia, British Journal of Neurosurgery, DOI: [10.1080/02688697.2019.1687848](https://doi.org/10.1080/02688697.2019.1687848)

To link to this article: <https://doi.org/10.1080/02688697.2019.1687848>



Published online: 02 Dec 2019.



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SHORT REPORT



A case of aneurysmal subarachnoid haemorrhage and superficial siderosis complicated by prosopagnosia, simultagnosia and alexia without agraphia

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ABSTRACT

A 42-year-old lady presented with acute aneurysmal subarachnoid haemorrhage and developed difficulty recognising faces (prosopagnosia), inability to process visual information in busy environments (simultagnosia) and difficulty to read (alexia). She was subsequently found to have superficial siderosis on MRI.

ARTICLE HISTORY

Received 19 July 2019
Revised 25 October 2019
Accepted 29 October 2019

KEYWORDS

Aneurysmal subarachnoid haemorrhage; prosopagnosia; simultagnosia; alexia without agraphia; superficial siderosis

Presentation

A 42 year old lady presented with sudden onset headache, seizure and decreased level of consciousness. On CT she was found to have diffuse subarachnoid haemorrhage (SAH) (Fisher grade IV) from a 6 mm anterior communicating artery aneurysm (Figure 1).¹ She had no neurological deficits and was WFNS grade 2.² The aneurysm was coiled. She developed blurring of vision in right eye at the time of the SAH and was found to have vitreous haemorrhage suggesting Terson syndrome. This did not settle and was treated with vitrectomy (3 months following her SAH). She also developed a small sub-inner limiting membrane bleed in the left eye which was treated conservatively. She developed headache and papilloedema with hydrocephalus on CT and was shunted 2 months after initial presentation. 7 months post-SAH (4 months after the vitrectomy), her visual acuity significantly improved from counting fingers in right eye to 6/6, and from 6/18 to 6/6 on the left but she continued to complain of considerable visual difficulties. These were initially difficult to define but after detailed evaluation, she had inability to recognise faces (prosopagnosia), inability to read (alexia without agraphia) and difficulty processing visual information in the presence of multiple afferent data (simultagnosia). The latter is due to a generalised impairment of capacity to integrate and consume multimodal parallel sensory information and therefore dislike busy, noisy places. In this case, the patient found it particularly difficult to recognise objects or read whilst she was moving but this was easier once she stood still.

Magnetic Resonance Imaging (MRI) 8 months following the initial presentation (Figure 2) revealed significant superficial siderosis which was fairly generalized, but most prominent in the interhemispheric fissure (around corpus callosum).

She remained under neuropsychological follow-up with visual cognitive tests that assessed verbal and visual memory, visual recognition, attention, visual perceptual skills (object and space), naming and executive functioning. The diagnosis of simultagnosia was made 5 months post-SAH based on clinical presentation and was found to be improving but not resolved on testing at 10 months following the haemorrhage. This was demonstrated in Cookie Theft tests (Table 1). Her visual perceptual abilities, visual confrontation naming, visual memory and recognition of different objects was accurate as seen by her performance on the Visual Reproduction test, Doors and People Test (Doors A), Boston Naming Test and Visual Object and Space Perception battery (Table 1). However, her recognition of faces was impaired as shown on the Warrington Recognition Memory for Faces Test (Table 1). The patient's recognition of objects within the same category (structurally similar) was also impaired as seen on the Doors and People Test (Doors B).

Discussion

Aneurysmal SAH may result in hemosiderin deposition over the surface of brain (superficial siderosis) but a combination of prosopagnosia, simultagnosia and alexia without agraphia have not been reported as complications of SAH or superficial siderosis.

Prosopagnosia results from either damage to the fusiform face area (located in the inferior temporal cortex in the fusiform gyrus) bilaterally or that of the right hemisphere as face identification seems to localize to the right.³

There are two forms of simultagnosia dorsal and ventral. The dorsal simultagnosia causes difficulty identifying more than one object at a time. It results from bilateral occipitotemporal cortex damage. In the ventral form, patients cannot recognize complex

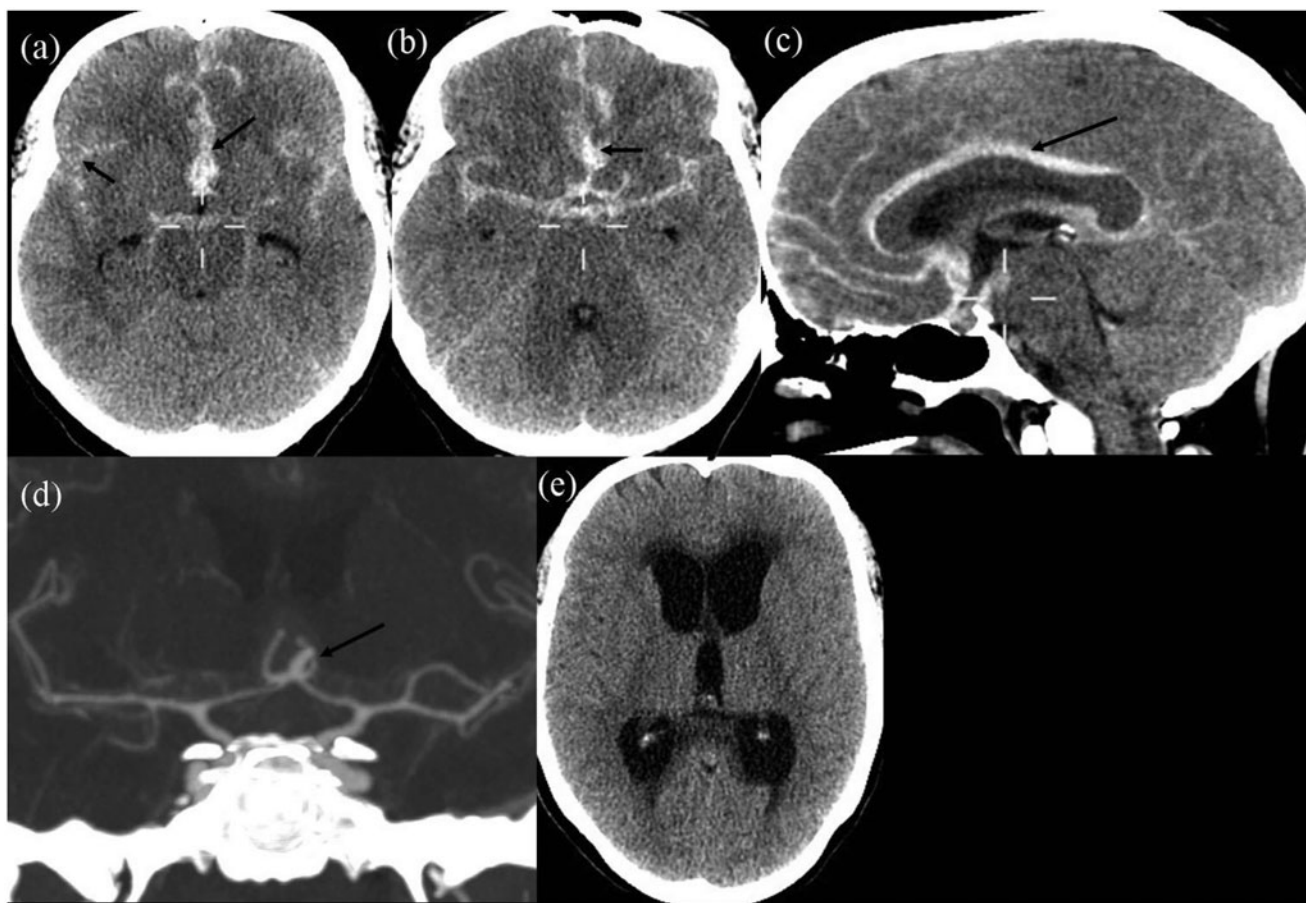


Figure 1. CT Scans. (a) and (b) Initial non-contrast CT axial scans showing diffuse SAH (black arrows). (c) sagittal CT reconstruction showing SAH (black arrow), quite prominent around corpus callosum. (d) CT angiogram showing ACom aneurysm (black arrow). (e) showing a subsequent CT scan showing hydrocephalus with periventricular interstitial oedema.

objects. These patients have lesions in the left inferior occipital area.⁴ Simultagnosia is known as part of the triad of Balint's syndrome, which includes optic apraxia and optic ataxia in addition to simultagnosia.³

Alexia without agraphia, also known as 'pure alexia', 'word blindness' and 'agnostic alexia, is known to be caused by lesions to the visual word form area in the left occipital lobe.⁵ In our case MRI ruled out any lesions in the fusiform gyri, occipital lobes and temporal lobes. The only prominent abnormality was widespread deposition of hemosiderin over the brain surfaces especially the corpus callosum and adjacent brain surfaces.

It has been reported that injury to the splenium of the corpus callosum and adjacent left occipital cortex can result in alexia without agraphia.⁶ The cause of that is the inability of signals to be transmitted to the dominant hemisphere through the corpus callosum from the right visual cortex. The injury to the left occipital lobe causes right homonymous hemianopia.³ This variable causation process has led to the generation of a new classification of pure alexia into disconnection alexia and cortical alexia.⁷

The causes of visual agnosia are variable but commonly include stroke and degenerative disorders.³ Occipital arteriovenous malformations have also been reported to cause pure alexia and prosopagnosia.⁸

Superficial siderosis is known to cause neurological disorders. In the infratemporal compartment, it can affect cranial nerves, the commonest being the seventh and eighth. The commonest symptoms described are hearing loss and ataxia.⁹

Superficial siderosis is usually diagnosed on MRI, the most sensitive sequence being susceptibility weighted images (SWI). Cortical superficial siderosis, which is the supratentorial form of hemosiderin deposition, may result in focal neurological deficits. In a study of cortical superficial siderosis, cognitive impairment was more frequent in patients with disseminated cortical superficial siderosis when compared to those with focal superficial siderosis.¹⁰

In a study that was done to assess prevalence of superficial siderosis following singular acute subarachnoid bleed, it was found that superficial siderosis develops in around half of patients. In the same study it was concluded that old age and the volume of SAH were associated with a higher incidence.¹¹ The authors noted that all patients in the study with *supratentorial* SAH developed cortical superficial siderosis compared to only 36% of patients with *infratentorial* SAH.

Superficial siderosis has also been found in cases of traumatic SAH. It was radiologically detected in around 97% of patients with traumatic SAH 6 weeks after the initial injury. There was also a correlation between the site of the subarachnoid bleed and that of the superficial siderosis.¹²

Superficial siderosis tends to happen preferentially at sites of greater blood deposition with less CSF flow. It is also more prominent in patients with high Fisher grade SAH.¹³ In our case the SAH resulted from a ruptured ACom aneurysm with a significant blood load over the corpus callosum and

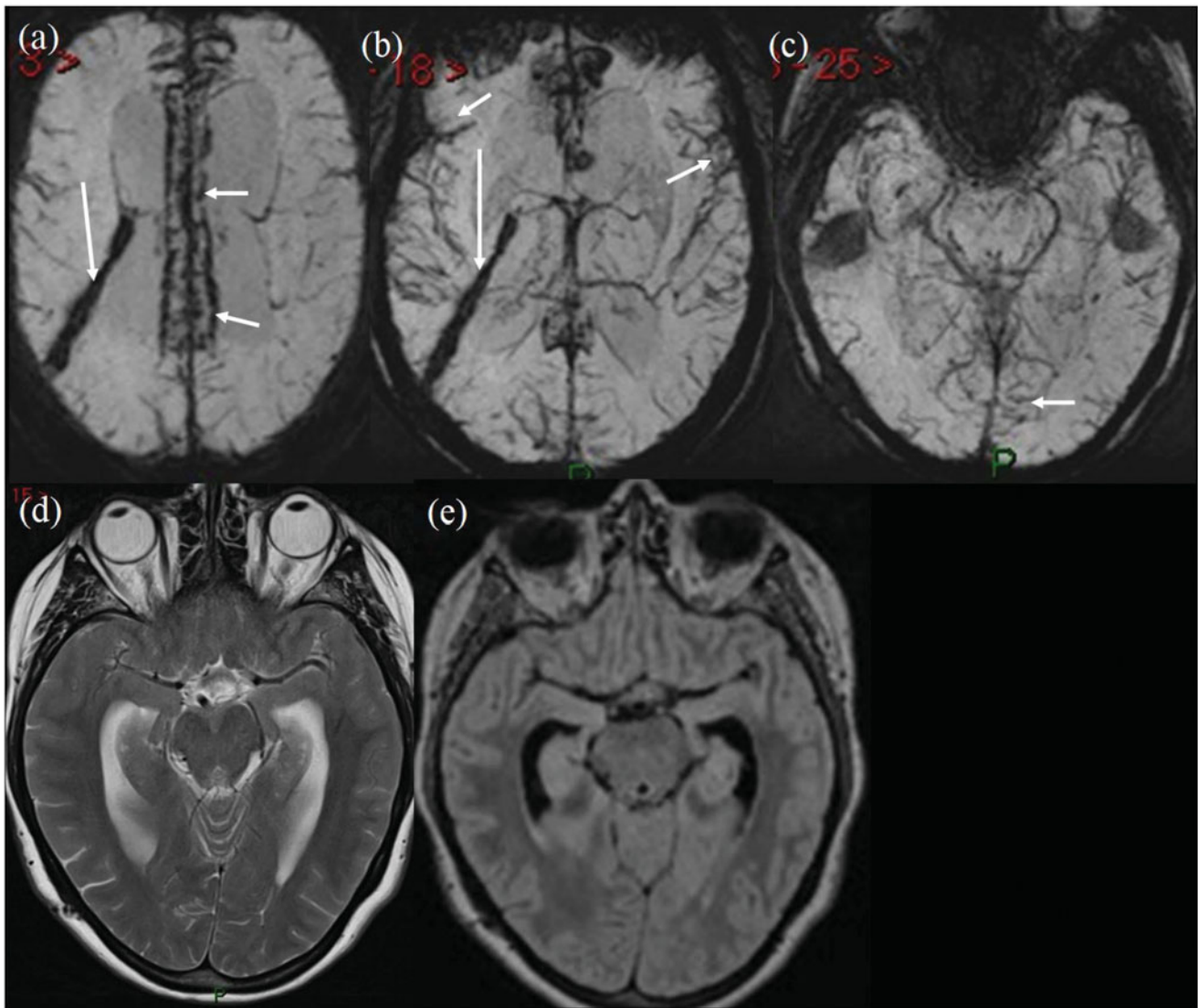


Figure 2. MRI. (a–c) Susceptibility-weighted Imaging (SWI) showing presence of haemosiderin deposition (short white arrows), this is particularly prominent in (a) around corpus callosum. Long white arrows in (a) and (b) show blood products along the shunt tract. (d) T2-weighted image and (e) FLAIR image, showing normal appearances of temporo-occipital lobes with no gross parenchymal changes.

Table 1. Neuropsychological results.

Cognitive domain	Test	Scores	Percentile
General Intelligence	WTAR FSIQ	101	
Memory	WMS-IV		
	Logical Memory I	14 (scaled score)	
	Logical Memory II	14 (scaled score)	
	Visual Reproduction I	7 (scaled score)	
	Visual Reproduction II	7 (scaled score)	
	Visual Reproduction Recognition	6/7 correct	(51–75%ile)
Attention/Processing Speed	WAIS-IV		
	Digit Span Forwards	7	
	Digit Span Backwards	5	
	Trail Making Test		
	Trails A		10%ile
	Trails B		10–20%ile
Language	Semantic fluency		25–50%ile
	FAS fluency		50–60%ile
	Boston Naming Test	54/60 correct	
Visuospatial	Line Bisection test	Accurate	
	Copying simple shapes	Accurate	
	Balloons Test	40/40 correct	
	Rey Complex Figure	31/36 correct	(29–40%ile)
	Doors and People Test		

(continued)

Table 1. Continued.

Cognitive domain	Test	Scores	Percentile
	Doors Test A (between category)	11/12 correct	(25–50%ile)
	Doors Test B (within category)	5/12 correct	(5%ile) ^a
	Warrington recognition memory for faces	38/50 correct	(5–10%ile) ^a
	VOSP		
	Incomplete letters	20/20	
	Silhouettes	28/30	
	Dot counting	10/10	
	Number location	10/10	
	Cube analysis	10/10	
	WMS-III		
	Spatial span forwards	5	
	Spatial span backwards	5	
	WAIS-IV		
	Block design	8 (scaled score)	
	Boston diagnostic Aphasia examination		
	Cookie theft picture	Accurate	

WMS-III: Wechsler Memory Scale – Third UK Edition (WMS-III-UK).

^aImpaired. WTAR FSIQ: Wechsler Test of Adult Reading – UK edition. WMS-IV: Wechsler Memory Scale—Fourth UK Edition (WMS-IV-UK). WAIS: Wechsler Adult Intelligence Scale—Fourth UK Edition (WAIS-IV-UK). VOSP: The Visual Object and Space Perception.

the adjacent cortex in the initial CT brain (Figure 1), which was also the subsequent site of maximum haemosiderin deposition.

We could find no prior reports of any relationship between superficial siderosis and any of the visual symptoms mentioned here. It is not known whether haemosiderin deposition over the corpus callosum can result in its dysfunction, potentially resulting in disturbance of signal transmission between the brain hemispheres. In absence of any other pertinent cause, we assume that it is the cause for this patient's presentation of alexia without agraphia. Given that superficial siderosis can cause a variety of cortical dysfunctions, we think it is reasonable to assume that the other symptoms of prosopagnosia and simultagnosia could be due deposition of hemosiderin on adjacent temporo-occipital lobes.

We also considered that possibility of ventricular dilatation as a potential cause of the abovementioned visual symptoms. We did come across a case of normal pressure hydrocephalus, associated with transient prosopagnosia, topographical disorientation, and visual objective agnosia; this was, however attributed to the periventricular white matter ischaemic changes rather than ventricular dilatation.¹⁴ We found no other association described between hydrocephalus and given visual symptoms and we could not think of any mechanism that could explain the same. We therefore believe that ventricular dilatation was incidental in the context of given visual symptoms.

Conclusion

We report the first case of visual cognitive difficulties in a patient with SAH and subsequent superficial siderosis. Cognitive visual impairment ought to be considered in patients where Terson Syndrome post-SAH does not improve and managed with ophthalmology and neuropsychology support.

Acknowledgements

The authors are grateful to the patient described in this case report for consenting to discussion and publication of their case history

Disclosure statement

The authors report no conflict of interest

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