

CASE REPORTS

Akinetic mutism—not coma

ANITA CHITTARANJAN SHETTY¹, JOHN MORRIS², PAUL O'MAHONY¹

¹Department of Geriatric Medicine, The University of Sydney, Westmead Hospital, NSW 2145, Australia

²Department of Neurology, The University of Sydney, Westmead Hospital, NSW 2145, Australia

Address correspondence to: Paul O'Mahony. Tel: (+44) 2084013616; Fax: (+44) 2084013620.

Email: paul.o'mahony@mayday.nhs.uk

Abstract

We describe a case of akinetic mutism mistaken as coma. A 77-year old lady presented with apparent unresponsiveness. In fact she responded when stimulated. She subsequently developed movements typical of clonic perseveration, erroneously treated as seizures. She also had features of a frontal lobe syndrome. Initial CT scan showed no abnormality; it was only after an MRI scan that a diagnosis of bilateral paramedian thalamic infarction was made. MRI scanning should be considered early in the investigation of patients with atypical neurological presentation.

Keywords: stroke, akinetic mutism, coma, elderly

A 77-year-old right-handed lady was brought to hospital 'unresponsive'. She had woken normally that morning to take her tablets but 3 h later her husband could not rouse her. The paramedics documented her Glasgow Coma Scale (GCS) as 5/15. Premorbidly, she had been functionally independent, with a history of hypertension, osteoarthritis, osteoporosis and glaucoma. She had never smoked or consumed alcohol. It was close to the anniversary of her son's death and three siblings had suffered fatal strokes.

Her GCS varied between 3 and 13. Vital signs were normal. She had bilaterally small pupils, sluggishly reacting to light and bilateral ptosis. When more alert, all limbs moved spontaneously. Reflexes were intact; plantar responses flexor. Speech was dysarthric. Examination was otherwise unremarkable. The emergency department (ED) physicians queried overdose, stroke and intracranial infection as a cause of perceived coma. Blood tests, electrocardiogram (ECG) and chest X-ray were normal. Blood alcohol and urine toxicology were negative. CT of the brain done within 4 h of presentation showed changes of small vessel disease. Cerebrospinal fluid examination was normal except for minimally raised protein. On day 3 when first seen by us, she lay still, with her eyes shut. She did not speak spontaneously. When stimulated, she opened her eyes, uttered a few slurred words obeying single step commands. Bilateral grasp reflexes were evident. Abnormal movements had developed, in the form of intermittent spontaneous (but also inducible) bilateral repetitive, rhythmical rubbing and scratching movements of

upper and lower limbs, which would persist for minutes. We prescribed phenytoin for suspected seizures. An electroencephalogram (EEG) showed diffuse slowing but no epileptiform activity. We considered an acute encephalopathy but other investigations (including biochemistry, glucose, blood gas and acid–base status, thyroid function and CT of pancreas) for this were negative. She required artificial hydration and feeding and total nursing care. As her condition continued to puzzle us, we requested an MRI scan, which showed bilateral acute paramedian thalamogeniculate infarction (Figure 1). Retrospective review of the initial CT brain scan found no early signs of these infarcts. Over the next month, the movements decreased and she spent more time with her eyes open, but needed enteral feeding. Despite intensive rehabilitation, she remained totally dependent requiring nursing home placement. At hospital presentation, some months later for enteral feeding tube replacement, her condition remained unchanged.

This patient had akinetic mutism not coma, with some of the classic features (apathy, akinesia, abulia, lack of initiative) of a frontal lobe syndrome due to thalamic disconnection. The key features were that while her eyes were often shut and she said nothing, she responded when stimulated. Moreover, she exhibited rhythmic, stereotyped, repetitive, inducible [1] movements of her limbs, which represent clonic perseveration [2]. This is the inappropriate repetition of an action, once initiated, in the absence of an ongoing cue; the ability to change in response to a new cue being preserved. The

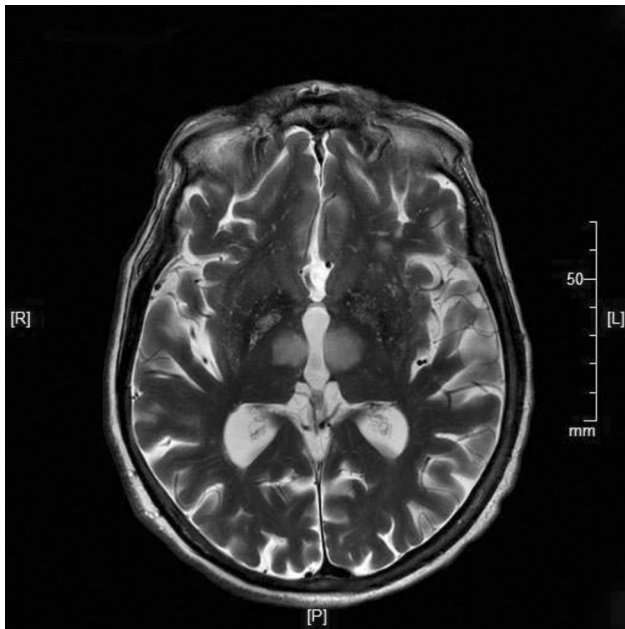


Figure 1. MRI showing bilateral paramedian thalamic infarction.

movements are commonly mistaken for tremor, chorea or seizure [1]. They may not appear for days after stroke onset as in our case and result from interruption of prefrontal connections at the thalamic level. Despite being uncommon (less than 0.6% of strokes), bilateral thalamic infarction presents a well-defined syndrome [2, 3]. Frequently reported features, which may fluctuate, include akinetic mutism, gaze palsies, dysarthria, hypersomnia and amnesia (thalamic dementia). We believe that the ‘ptosis’ documented by ED physicians was in fact a combination of hypersomnia with vertical gaze palsy.

Differentiating akinetic mutism from other disorders of consciousness is difficult, reflecting the complex underlying pathophysiology in such cases [4]. Thorough clinical assessment with appropriate investigation and consultation with neurologists will facilitate physicians in more accurate diagnosis of such cases.

The paramedian, inferolateral, tuberothalamic and posterior choroidal arteries all supply blood to the thalamus. The paramedian artery on each side normally arises from the basilar communicating artery. An anomaly of this, whereby both paramedian thalamic arteries arise from a basilar communicating artery on one side, either as a single trunk (this variant is called the artery of Percheron) or as two separate closely related vessels is most commonly identified in cases of bilateral thalamic infarction [5]. Occlusion of the single trunk will result in bilateral infarction. A genetic origin to the anatomical abnormality has been proposed [6], and our patient’s family history of stroke would support this. Diverse risk factors (and even none) have been described [3, 6]. The prognosis of the condition is often poor as in our patient and

seems independent of the presence or absence of ‘frontal lobe syndrome’ symptomatology [3, 6]. Bilateral paramedian thalamic infarcts can cause sudden onset of akinetic mutism and clonic perseveration, conditions which may be confused with coma and seizures. MRI scanning should be considered early in the investigation of patients with such an atypical presentation.

Key points

- Differentiating akinetic mutism from coma is difficult.
- Clonic perseveration is an unusual but well recognised sign which may be mistaken for seizures.
- Bilateral paramedian thalamic infarction is an uncommon but recognised cause of akinetic mutism and clonic perseveration.
- MRI scanning should be considered early in the investigation of patients with atypical neurological presentation.

Acknowledgements

We wish to acknowledge the patient and the next of kin to whom this case report refers.

Conflicts of interest

We declare that we participated in the preparation and writing of this manuscript and that we have seen and approved the final version. The article is original work, has not received prior publication and is not under consideration for publication elsewhere. We have no relevant conflicts of interest.

References

1. Fung VSC, Morris JGL, Leicester J, Soo YS, Davies L. Clonic perseveration following thalamofrontal disconnection: a distinctive movement disorder. *Mov Disord* 1997; 12: 378–85.
2. Liepmann H. Die perseveration. In: Liepmann H, ed. *Ueber Storungen des Handelns bei Gehirnkranken*. Berlin: Karger, 1905; 115–27.
3. Kumral E, Evyapan D, Balkir K, Kutluhan S. Bilateral thalamic infarction: clinical, etiological and MRI correlates. *Acta Neurol Scand* 2001; 103: 35–42.
4. Giacino JT. Disorders of consciousness: differential diagnosis and neuropathologic features. *Semin Neurol* 1997; 17: 105–11.
5. Percheron G. The anatomy of the arterial supply of the human thalamus and its use for the interpretation of the thalamic vascular pathology. *Z Neurol* 1973; 205: 1–13.
6. Reilly M, Connolly S, Stack J, Martin EA, Hutchinson M. Bilateral paramedian thalamic infarction: a distinct but poorly recognized stroke syndrome. *Q J Med* 1992; 82: 63–70.

Received 19 August 2008; accepted in revised form 05 January 2009