The cerebellar cognitive affective syndrome

Introduction

This article presents a case of cerebellar cognitive affective syndrome to remind the general physician of the profound effect that the cerebellum exerts over executive functioning, visuospatial cognition, personality and linguistic ability. Described in a variety of underlying cerebellar pathologies, it is important that this often overlooked syndrome is remembered, as the cognitive effects of cerebellar pathology can often be more disabling than the motor effects (Garrard et al, 2002; Maeshima and Osawa, 2007).

Discussion

These deficits, which are more traditionally associated with frontal executive dysfunction, illustrate features of the cerebellar cognitive affective syndrome. This syndrome is well described in patients with lesions confined to the cerebellum and is characterized by the following features (Schmahmann, 1998):

1. Disturbed executive function – deficient planning, abstract reasoning, working memory, verbal fluency
2. Impaired spatial cognition – visuospatial disorganization and poor visuospatial memory
3. Personality change – blunted affect and disinhibition
4. Linguistic difficulties – mild anomia, agrammatism.

Case reports from the 1830s suggesting that the cerebellum may exert control over non-motor function and behaviour were dismissed as anecdotal and it was not until the 1970s and 80s that the role of the cerebellum in non-motor functioning was more fully described. Schmahmann and Sherman (1998) reported 20 cases of isolated cerebellar pathology and summarized these dysfunctions in executive functioning, language, affect and spacial cognition as the cerebellar cognitive affective syndrome. The underlying cerebellar pathology described is varied and can occur with olivopontocerebellar atrophy, Freidrich's ataxia, following excision of tumours, stroke and chronic phenytoin intoxication (Desmond and Fiez, 1998; Schmahmann and Sherman, 1998).

Positron emission tomography and functional magnetic resonance imaging provide an anatomical substrate accounting for the role of the cerebellum in higher order processing. Feedforward and feedback connections exist between the cerebellum and cerebral cortical regions suggesting involvement in cognitive processes such as working memory, language, and implicit and explicit memory (Desmond and Fiez, 1998; Middleton and Strick, 1998; Paulus et al, 2004). Just as the cerebellum regulates rate, force, rhythm and accuracy of movement so it maintains cognitive function around a 'homeostatic baseline' regulating speed, capacity, consistency and appropriateness of mental or cognitive processes. Without this mental processes are imperfectly conceived, erratically monitored and poorly performed (Schmahmann, 1998).

Although physically the patient could have returned to work his ongoing difficulty with verbal generation and executive function has prevented this. Before his discharge from the stroke unit he was asked to give a short lecture to staff on the unit, which was video-recorded and then analysed with his therapists. Although he seemed confident about his ability to achieve this simple task, in reality he encountered marked difficulties. He was disablingly nervous, there was profound disorganization of material, questions were answered tangentially and he had difficulties with word finding and naming.

Conclusions

This case is presented to remind the general physician of the effect that the cerebellum exerts over cognition, affect and verbal function, now described in a variety of cerebellar pathologies, as in some cases this is more disruptive to recovery than the motor effects of the disease process. BJHM

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Case Report

A 62-year-old male university lecturer suffered a hypertensive cerebellar haemorrhage while on holiday. The haemorrhage was confined to the dentate nucleus and was managed conservatively with blood pressure control and physiotherapy before the patient was allowed to fly back to the UK. Six weeks after the haemorrhage on his return, he self presented to the emergency department, reporting confusion and word finding difficulty. On initial examination he was normotensive with a wide based gait, subtle right-sided past pointing and notable word finding difficulties in conversation. Abbreviated Mental Test Score performed by the admitting doctor was 10/10. Folstein Mini Mental State Examination was 29/30. He was admitted to the stroke unit for assessment.

During his admission despite rapid progress physically with physiotherapy, the team were surprised by the patient's blunted affect and apparent difficulty in verbal expression. Further neuropsychological assessment revealed profound abnormalities of language, affect and executive functioning.

He obtained only an average score using the Wechsler (1982) Abbreviated Test of Intelligence, which is less than expected given his premorbid high academic function. Using the graded naming test (Cambridge Cognition; McKenna and Warrington, 1983) he was only able to name four of 30 line drawings suggesting significant anomia. In the similarities test he exhibited original but not essential relationships. His score in the Hopkins Verbal Learning Test (Brandt, 1991) was well below the mean, again at odds with his high premorbid academic level. There was a marked slowness in generating words with numerous errors in conforming to simple rules in the letter verbal fluency test suggesting executive dysfunction. Similarly using the category fluency test the managed to name only 15 animals in 60 seconds, which is at the 25% level. Further assessment using the proverb test revealed concrete explanations with low scores in both his ability with accuracy and achievement.

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Large bladder stone masquerading as foreign body

A 78-year-old man with hypertension and diabetes mellitus was brought to the emergency department having suffered fever and disturbances in consciousness for 2 days. On examination, his vital signs were as follows: body temperature 38°C, heart rate 116 beats/minute, respiratory rate 22/minute, and blood pressure 145/72 mmHg. Abdominal examinations were all negative. Urinalysis showed pyuria and bacteriuria with calcium oxalate crystals, and laboratory tests revealed leukocytosis with a left shift.

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Plain abdominal film showed a radiographic opacity within pelvic area (*Figure 1a*). Pelvic ultrasonography showed a hyperechoic lesion with strong post-acoustic shadow in the urinary bladder (*Figure 1b*). Cystostomy and lithotripsy confirmed the presence of a large bladder stone (*Figure 2*). His consciousness improved after treatment and he was discharged 1 week later uneventfully.

There have been several reports describing a pelvic radio-opacity of such size and its origin, including foreign body (Kasmani and Irani, 2010), calcification of pelvic tumour (Burkill et al, 2009) and Meckel's diverticulum with enterolith (Gibney, 1991). Rapid bedside ultrasound could facilitate subsequent management and reduce unnecessary workup, such as computed tomography. BJHM

Burkill GJC, Allen SD, A'Hern R et al (2009) Significance of tumour calcification in ovarian carcinoma. *Br J Radiol* **82:** 640–4
Gibney EJ (1991) Diverticulum of a Meckel's diverticulum containing a stone. *Postgrad Med J* **67:** 692–4
Kasmani R, Irani F (2010) Opacity in the Pelvis. *Am J Med Sci* **340:** 154

Figure 1. a. An abdominal film showed a round and homogenous density mass at the centre of the true pelvis. A central line is set through the left femoral vein. b. Pelvic ultrasonography revealed a highly echogenic substance inside the urinary bladder with strong post-acoustic shadow.

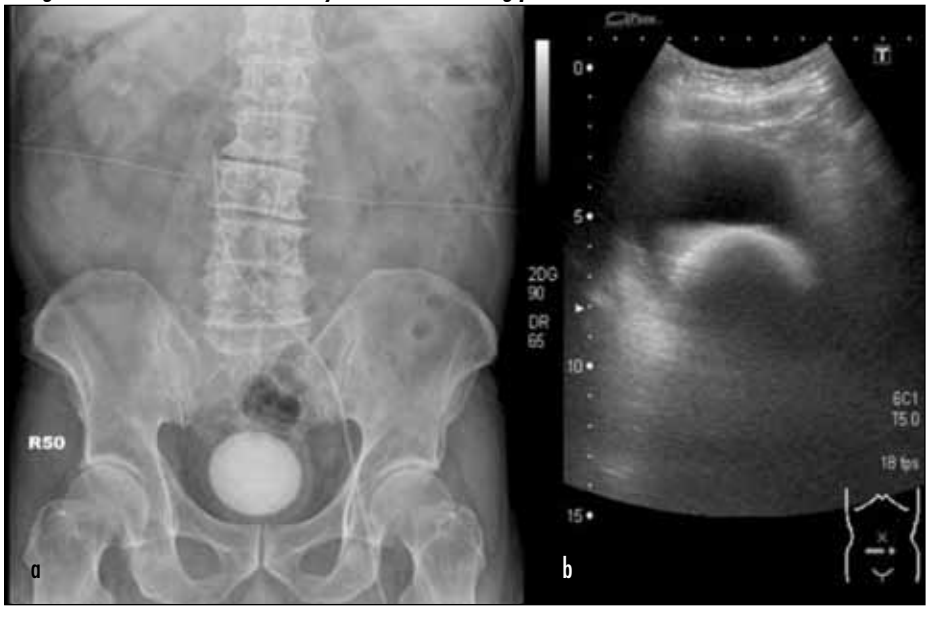

Figure 2. A bladder stone measuring 5 cm was retrieved after cystostomy. There is a multitude of crystals concentrating on the stone surface, as can be seen shedding on the plate.