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ORIGINAL PAPER

Posterior Fossa Syndrome in an Adult Patient Following Surgical Evacuation of an Intracerebellar Haematoma

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Abstract The posterior fossa syndrome (PFS) consists of transient cerebellar mutism, cognitive symptoms and neurobehavioural abnormalities that typically develop in children following posterior fossa tumour resection. Although PFS has been documented in more than 350 paediatric cases, reports of adult patients with a vascular aetiology are extremely rare. In addition, the pathophysiological substrate of the syndrome remains unclear. We report an adult patient with PFS after surgical evacuation of a cerebellar bleeding. After 45 days of (akinetic) mutism, the patient's cognitive and behavioural profile closely resembled the "cerebellar cognitive–affective syndrome". A quantified SPECT study showed perfusional deficits in the anatomoclinically suspected supratentorial areas, subserving language dynamics, executive functioning, spatial cognition and affective regulation. We hypothesize that cerebello-cerebral diaschisis might be an important pathophysiological mechanism underlying akinetic mutism, cognitive deficits and behavioural–affective changes in adult patients with PFS.

Keywords Posterior fossa syndrome · Cerebellum · SPECT · Cerebellar cognitive–affective syndrome

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Introduction

Although the posterior fossa syndrome (PFS) is an aetiologically heterogeneous clinical condition that may develop following acute cerebellar damage, it mostly occurs after posterior fossa tumour surgery in paediatric patients. Transient mutism is considered as the core symptom of PFS, but associated neurobehavioural abnormalities and personality changes also typically develop after a short postoperative interval of relatively normal functioning. Despite extensive research, the pathophysiological substrate of PFS remains unclear (for a review see De Smet et al. [1]).

Although documented in more than 350 paediatric cases, PFS has only been described in 21 adults. In addition, PFS associated with vascular aetiologies is only reported in a very limited number of three adult cases [2–4]. We report the clinical and functional neuroimaging findings in an adult who presented with PFS following surgical evacuation of an intracerebellar haematoma. A technetium-99m-ethyl cysteinate dimer (Tc-99m-ECD) single-photon emission computed tomography (SPECT) study of the brain revealed perfusional deficits in the anatomoclinically suspected supratentorial regions.

Case Report

During a stay abroad, a 60-year-old right-handed civil engineer acutely developed severe headache with vomiting. On admission to a hospital a few hours later, the patient was conscious and cooperative. The clinical neurological examination only revealed bilateral dysdiadochokinesia and mild dysmetria, more pronounced on the left side. Tendon reflexes were brisker at the right than on the left side of

the body. Plantar response was flexor bilaterally. No speech abnormalities were found. CT scan of the brain showed a left cerebellar haematoma and obstructive hydrocephalus. Since consciousness progressively decreased, a large cerebellar blood clot was surgically removed on the day of admission and an external ventricular drain was installed. The patient was intubated and ventilated. Five days post-stroke, he was transferred to the department of neurology of ZNA Middelheim for further treatment. Sedation and ventilation were discontinued 10 days post-stroke due to bacterial infection (methicillin-resistant *Staphylococcus aureus*). A tracheostomy with a speech cannula was installed, but the patient did not produce any speech. Prefrontal-like inhibitory symptoms such as a complete lack of spontaneity, flattened affect, akinesia and apathy dominated the clinical tableau. Twenty days post-surgery, the patient started to move his limbs on verbal command. Mutism, however, persisted. Repeat CT of the brain revealed hyperdense lesions in the resection area and oedema. Ventrical volumes were normal. No supratentorial lesions were detected. After 45 days of complete mutism, verbal reactions could be evoked after stimulation. Speech was marked by ataxic dysarthria. The patient only responded by means of short two- to three-word utterances. Two months post-stroke, the neurological examination only revealed mild ataxia (balance disturbances and dysdiadochokinesia) reflected by a score of 5/30 on the Brief Ataxia Rating Scale [5]. Magnetic resonance imaging (MRI) of the brain showed sequela of a left posterior mid-cerebellar haemorrhage with chronic hemosiderin located in the midcerebellar and posterior cerebellar region (Fig. 1A–D). Quantified Tc-99m-ECD brain SPECT was performed 3 months post-surgery. Transaxial images with a pixel size of 3.56 mm were anatomically standardized using SPM and compared to a standard normal and SD image obtained from 15 normal ECD perfusion studies. Using a 31 ROI template, the Z-scores (SD) were calculated for each region. A regional Z-score of >2.0 is considered significant. In comparison to normal database findings, ECD SPECT results showed significantly decreased perfusion in the left cerebellar hemisphere (−2.58 SD) as well as in the right prefrontal lateral region (−4.35 SD), both medial prefrontal regions (right −6.32 SD, left −4.23 SD) and the right temporal lateral area (−2.12 SD; Fig. 2A–D).

At 3.5 months post-stroke, the patient started to show more initiative, but verbal output was still severely reduced. Speech was characterized by ataxic dysarthria, including imprecise consonants, distorted vowels, irregular articulatory breakdown, monopitch, slow rate and excess and equal stress. Language was formally investigated by means of the Comprehensive Aphasia Test-NL [6, 7] which revealed no evidence for an underlying aphasic syndrome. Visual confrontation naming (Boston Naming Test, [8]) was

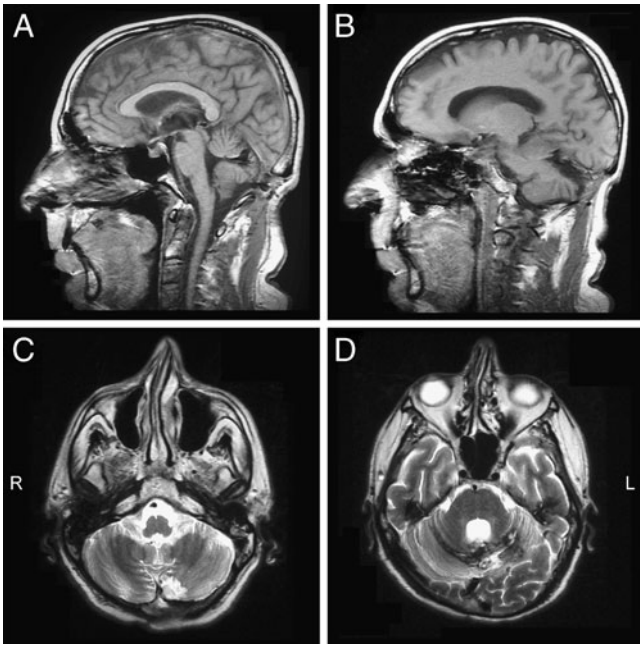

Fig. 1 Brain MRI sagittal FSE T1-weighted slices (**A**–**B**) revealing sequela of a left posterior mid-cerebellar haemorrhage and axial FSE T2-weighted slices (**C**–**D**) disclosing post-stroke changes with chronic hemosiderin located midcerebellar and posteriorly

normal. Cognitive screening by means of the Mini-Mental State Examination [9] was deficient (21/30; Table 1). The Wechsler Adult Intelligence Scale-III [10] was marked by a significant discrepancy of 23 IQ points between a normal verbal IQ of 104 and a depressed performance IQ of 81. Deficient scaled scores of ≥2 SD were found at the performance level for the subtest digit symbol substitution. Immediate and recent memory were impaired (Repeatable Battery for the Assessment of Neuropsychological Status) [11]. Disruption of cognitive flexibility and conceptual organisation of goal-oriented cognitive strategies was indicated by defective results on the Wisconsin Card Sorting Test [12]. The patient did not succeed to complete any category within 128 trials (<pc 1). In comparison to an age- and education-matched control group, he obtained a standard score of 76 (pc 5) for perseverative responses, which places him in the mildly to moderately impaired range. A strong tendency to perseverate was confirmed by a defective score on the Frontal Assessment Battery [13].

Discussion

Following surgical evacuation of an intracerebellar haematoma, this patient presented with akinetic mutism for 45 days and "prefrontal"-like behavioural disturbances, matching a diagnosis of PFS. When mutism receded, cognitive and affective changes were found including visuospatial and attentional deficits, impaired frontal plan-

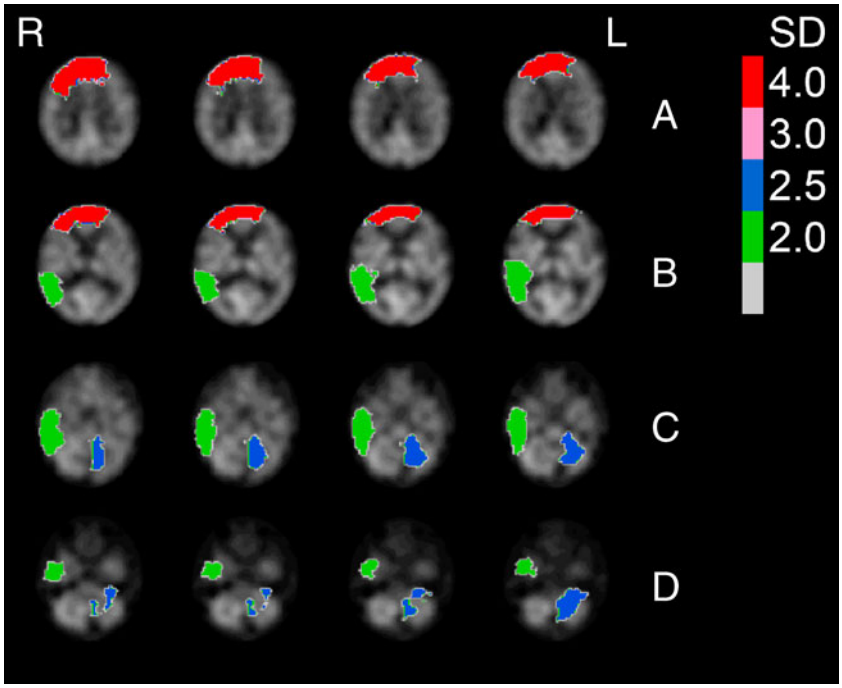

Fig. 2 Quantified ECD-SPECT showing a hypoperfusion at the supratentorial level (rows **A-D**) bilaterally in the medial prefrontal lobes, the right prefrontal lateral region and right temporal lateral area. In addition, a hypoperfusion was found infratentorial level (rows **C-D**) in the left cerebellar hemisphere

ning and problem solving, memory problems, reduced verbal fluency, decreased language dynamics and frontal-like behavioural problems such as apathy, behavioural and verbal inhibition, loss of facial expressions and a withdrawn attitude. This symptom complex strongly resembles the "cerebellar cognitive–affective syndrome" (CCAS), consisting of executive, visuospatial, affective and linguistic symptoms [14]. Indeed, recent studies not only showed that PFS and CCAS share overt semiological resemblances but may also be considered as cerebellar-induced phenomena, ranging on a continuum with different degrees of severity and symptom duration [15, 16].

To the best of our knowledge, only three adult patients have been reported with postoperative mutism following posterior fossa surgery for a vascular pathology (for a review see Mariën et al. [17]). Coplin et al. [2] described a 47-year-old man who developed mutism immediately after a vermian haematoma was evacuated. The 53-year-old patient of Dunwoody et al. [3] became mute 1 day after surgical treatment of a vermian arteriovenous malformation (AVM). SPECT demonstrated decreased perfusion in the entire left cerebral hemisphere. Idiaquez et al. [4] reported a 20-year-old man with a cerebellar haematoma and AVM who became mute 2 days after partial resection of the vermis and the right cerebellar hemisphere. Since none of these patients became mute before surgery, it appears that surgical damage to the cerebellum induced mutism.

Several hypotheses and risk factors have been advanced to explain the pathophysiological mechanisms involved in the PFS (for a review see De Smet et al. [1]). During the past years, the hypothesis of cerebello-cerebral diaschisis, reflecting the functional impact of a cerebellar lesion on distant cortical areas [18, 19], has attracted increased attention to explain cognitive and affective disturbances following posterior fossa lesions [4, 20]. Dunwoody et al. [3] already indicated that the PFS in their patient was not solely the result of local damage to the cerebellum but probably due to some distant hemispheric disturbance induced by the injury. However, the authors doubted whether the phenomenon of cerebello-cerebral diaschisis could be considered an important mechanism implicated in the pathophysiology of cerebellar mutism. Our findings seem to confirm that surgical cerebellar damage in adult patients may induce remote functional disturbances at the supratentorial level. In our patient, marked functional disruption of the structurally unaffected prefrontal brain regions was reflected by quantified SPECT findings showing a significant decrease of perfusion in the right prefrontal lateral region and both medial prefrontal areas. This observation might indicate disruption of transmission of excitatory impulses from the lesioned cerebellum to the prefrontal supratentorial regions crucially implicated in the regulation of cognitive, affective and behavioural processes. The dentatorubrothalamic tract, originating in the dentate nucleus, is reciprocally connected with the thalamus, which in turn is also connected to the prefrontal lobe, including the supplementary motor area (SMA) [21] known to be crucially involved in the planning and initiation of motor actions, including speech production [22]. Dysfunction of the SMA, as a possible consequence of cerebellar-induced deafferentiation, might in turn be considered to cause a range of behavioural and cognitive deficits among which is

Table 1 Neurocognitive test data 3 months post-surgery

Test	Test score	Mean	SD
Mini-Mental State Examination	21/30	29	1.3
Intelligence			
Standard Progressive Matrices	109	100	15
Wechsler Adult Intelligence Scale-III			
Full-scale IQ (FSIQ)	92	100	15
Verbal IQ	104	100	15
- Vocabulary	16	10	3
- Similarities	10	10	3
- Arithmetics	7	10	3
- Digit span	10	10	3
- Information	12	10	3
- Comprehension	10	10	3
Performance IQ	81	100	15
- Picture completion	6	10	3
- Digit symbol substitution	3	10	3
- Block design	9	10	3
- Matrix reasoning	9	10	3
- Picture arrangement	8	10	3
Repeatable Battery for the Assessment of Neuropsychological Status			
Visuospatial/constructive index	89	100	15
Immediate recall index	69	100	15
Language index	89	100	15
Concentration index	60	100	15
Delayed recall index	71	100	15
Language			
Boston Naming Test	50/60	55	3.21
Verbal fluency			
Semantic generation	37	60.4	7.14
Animals, 1 min	11		
Transportation, 1 min	8		
Vegetables, 1 min	8		
Clothing, 1 min	10		
Executive Functioning			
Wisconsin Card Sorting	0 cat. (128 trials)		

akinetic mutism. Miller et al. [20] suggested that bilateral damage anywhere along the proximal efferent cerebellar pathway (pECP) may result in PFS. They showed that in comparison to the children who did not develop PFS, the group of patients with PFS presented a consistent pattern of bilateral surgical damage to the pECP. In addition to pECP lesions, an association was found with significant perfusion alterations predominantly affecting the frontal regions. This finding is consistent with the phenomenon of cerebello-cerebral diaschisis. In addition, several studies have shown that left cerebellar damage results in typical right hemisphere dysfunctions, whereas right cerebellar lesions frequently lead to typical left hemisphere symptoms [16, 19]. This pattern of crossed functional connectivity between the cerebellum and the supratentorial association areas has been documented in several studies [23, 24]. In this respect, the hypoperfusion affecting the medial prefrontal regions bilaterally is rather unexpected given the absence of a perfusional deficit in the right cerebellar hemisphere. However, perfusional deficits affecting both prefrontal lobes have been reported after lesions confined to the cerebellar vermis [15]. Although it did not reach statistical significance (−1.45 SD), a relative hypoperfusion involving the vermis was found in our patient as well. An alternative explanation may be related to the phenomenon of 'trans-callosal' or 'transhemispheric diaschisis', which is reflected

by a hypoperfusion in the cerebral hemisphere opposite to the functionally disrupted region [25, 26]. Possible mechanisms of transcallosal diaschisis include disrupted input from the afferent fibre pathways [27], depressed excitatory transcallosal connection fibres [28], or transcallosal fibre degeneration [29]. Still another explanation might be found in the superficial siderosis. Van Harskamp et al. [30] investigated cognitive and social impairments in patients with superficial siderosis, which typically affects the cerebellum and which is caused by a deposition of hemosiderin in the superficial layers of the central nervous system. They found that superficial siderosis may be associated with a distinct pattern of cognitive and social abnormalities, including mild deficits in speech production, visual recall memory, executive function and difficulties in interpreting the behaviour of others (theory of mind (ToM)). Based on these results, the authors suggested a link with the CCAS. However, they also noted that the behavioural changes in their patients may be caused by supratentorial rather than cerebellar pathology [30]. Superficial siderosis is probably not the cause of the neurobehavioural deficits in our patient since he did not present with the typical symptoms of superficial siderosis including hearing loss, ataxia and anosmia; his neurobehavioural symptoms were rather related to apathy and inhibition than to ToM, and the cerebral cortex was not affected. Therefore, we suggest that in this patient, post-mutism cognitive and affective symptoms, including a range of executive dysfunctions, disrupted visuospatial skills, attentional deficits, frontal-like behavioural disturbances may be correlated with the perfusional deficits in the anatomoclinically suspected prefrontal and right temporal cortical areas which subserve executive processing, behavioural–affective processes and spatial cognition. As a result, our findings seem to show that the phenomenon of cerebello-cerebral diaschisis may be crucially implicated in the development of cognitive, affective and behavioural deficits in adult patients after surgery for (vascular) cerebellar pathologies.

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