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Posterior fossa syndrome with a large inflammatory ponto-mesencephalic lesion

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ABSTRACT

Demonstration of a posterior fossa syndrome (PFS) in a 32-year-old male patient with clinically isolated syndrome which subsequently developed into relapsing-remitting Multiple Sclerosis.

The patient suffered from double vision, coordination problems including unsteady gait and atactic dysarthria, concentration difficulties, as well as adynamia and impaired decision making.

The patient clinically presented a cerebellar and dysexecutive syndrome. Cerebral magnetic resonance imaging (MRI) revealed a contrast enhancing ponto-mesencephalic lesion with a volume of 4.8 cm^3. Neuropsychological tests showed pronounced executive dysfunctions, reduced visuoconstructive skills, attentional deficits, echolalia, and non-fluent speech production.

After cortisone and plasmapheresis, the cerebellar syndrome improved but manual fine motor skills and executive dysfunctions persisted.

After three months, symptoms remitted except for a slight gait imbalance. After six months, neuropsychological tests were normal except for a moderate attention deficit. MRI revealed a clear regression of the ponto-mesencephalic lesion to a volume of 2.4 cm^3 without contrast enhancement.

This case report intends to provide an overview of the symptomatology and etiology of PFS and offers new insights into its pathomechanism demonstrating a pontine disconnection syndrome caused by a large demyelinating plaque.

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1. Introduction

Posterior fossa syndrome (PFS) is an aetiologically heterogeneous condition that may typically develop after various lesions of the cerebellum. It is characterised by several motoric, linguistic, cognitive and affective-behavioral abnormalities (Pollack, 1997). Most often it develops in children after posterior fossa tumor resection. More than 350 pediatric cases of PFS have been described, whereas it occurs very rarely in adults (21 cases) (De Smet & Marien, 2012). This discrepancy in prevalence may be associated with a higher frequency of posterior fossa tumors in children than in adults (54–70% versus 15–20%) (O' Brien et al., 2001). Indeed, Marien, De Smet et al., 2013 showed that up to 98.5% (196/199) of PFS cases in children are due to neoplastic etiology compared to 85.7% (18/21) in adults.

PFS due to vascular etiology is very rare in children (1.5%, 3/199) and has been reported in 14.3% of adult patients (Marien, De Smet et al., 2013).

According to present knowledge, patients with cerebellar stroke only developed PFS following surgical evacuation of the lesion. PFS with akinetic mutism and cerebellar cognitive affective syndrome (CCAS) in a 71-year-old patient after cerebellar haemorrhage without surgical treatment was described only once (Marien, Verslegers et al., 2013).

PFS presents a complex neurological dysfunction affecting motor and non-motor functions, including cognition, behavior, and language. Most often it develops after a short postoperative, symptom-free period with cerebellar mutism – which itself may be a dysexecutive mutism indirectly caused by a cerebellar lesion (authors' annotation) - that may last for a couple of days to several months followed by motor speech deficits (De Smet et al., 2007). Cerebellar mutism is frequently accompanied by decreased initiation of voluntary movements and swallowing difficulties (Siffert et al., 2000). Cognitive deficits include executive dysfunctions, visuo-spatial disorders, as well as attentional and concentration

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deficits (De Smet et al., 2009). Non-motor language symptoms consist of impaired verbal fluency, word-finding difficulties, agrammatism, and comprehension deficits (Aarsen, Van Dongen, Paquier, Van Mourik, & Catsman-Berrevoets, 2004; De Smet et al., 2009; Siffert et al., 2000). In addition, most patients present behavioral and affective disorders including adynamia, apathy, disinhibited behavior as a result of frontal lobe dysfunction (Pollack, 1997), as well as symptoms consistent with a CCAS (Schmahmann & Sherman, 1998).

The pathophysiological mechanisms of PFS are not yet clear. Several hypotheses have been established including the phenomenon of cerebello-cerebral diaschisis. This phenomenon is based on the idea of functional disruption of reciprocal pathways that connect the cerebellum with cortical areas associated with cognitive, behavioral, and affective regulation (Baillieux et al., 2006; Marien, De Smet, Paquier, & Verhoeven, 2010; Marien et al., 2009; Miller et al., 2010).

Marien, De Smet et al. (2013) described an adult patient with PFS after resection of an ependymoma in the posterior fossa. The preoperative phase was characterized by symptoms matching the diagnosis of CCAS. In the immediate postoperative phase, the patient showed prefrontal dysfunctioning and subsequently akinetic mutism that lasted for 12 days, as well as pathological laughing and crying. These impairments resolved whereas CCAS persisted during longterm follow-up. De Smet and Marien (2012) reported an adult patient developing PFS after surgical resection of an intracerebellar haematoma. After 45 days of cerebellar mutism, the patient showed symptoms of CCAS consisting of visuospatial and attentional deficits, reduced verbal fluency, as well as frontal-like behavioral and affective abnormalities. Single-photon-emission computer tomography showed significantly decreased perfusion in the anatomo-clinically suspected supratentorial regions in both case reports, proving the distant impact of the cerebellum on cognition and affective regulation. Consequently, authors suggested a crucial involvement of the phenomenon of cerebello-cerebral diaschisis in the development of PFS. Morris et al. (2009) evaluated a cohort of 26 children after resection of an embryonal tumor (13 patients developed PFS, 13 non-affected). In patients with PFS, magnetic resonance imaging (MRI) showed a more rostral position of the tumor in the 4th ventricle and more signal abnormalities in the superior cerebellar peduncles and midbrain compared to non-affected patients. They concluded that damage of dentato-thalamo-cortical pathways and functional disruption of white matter bundles in the superior cerebellar peduncles may contribute to the development of PFS.

The purpose of this paper is to report about neurobehavioral and neuroradiological findings in a 32-year-old male patient with a ponto-mesencephalic lesion matching the diagnosis of a clinically isolated syndrome (CIS) (Polman et al., 2011) and a pontine disconnection syndrome with PFS. According to today's knowledge this is the first case of PFS resulting from a non-surgical, inflammatory lesion located in the ponto-mesencephalic brainstem.

2. Case report

2.1. Presentation as PFS and diagnosis of CIS

Our study complies with the Declaration of Helsinki. Written informed consent was obtained.

A 32-year-old man was hospitalised in our department of neurology for evaluation of progressive ataxia affecting gait and coordination of extremities, double vision, and perioral hypesthesia. In addition, he noticed fatigue, concentration difficulties, and a disability to estimate distances properly. His girlfriend reported behavioral and affective changes, mainly characterised by adynamia, decreased motivation, social withdrawal, and impaired expression of will and decision-making.

Clinical examination on first admission revealed a ponto-mesencephalic syndrome with bilateral internuclear ophthalmoplegia, trochlear nerve palsy on the left side with a compensatory head tilt to the right and Bielschowsky phenomenon, up-beat-nystagmus, hearing impairment on the left, and positive Romberg test. Evaluation of head-impulse test and vestibular ocular reflex suppression were not possible due to patient's malcompliance. Cerebellar signs included atactic dysarthria, gait ataxia, mild bilateral dysdiadochokinesia, and unprecise finger-to-nose test on the right side. Tetrahyperreflexia without Babinski signs suggested pyramidal involvement. The patient also showed a dysexecutive syndrome with echolalia, non-fluent speech production, and attention deficit. Moreover he suffered from agraphia and apraxia. Personal medical history was unremarkable; family history was positive for bipolar disorder. The patient had an educational level of 12 years and worked in training management.

Cerebral MRIs (Fig. 1A–D) showed a large non-space-occupying ponto-mesencephalic lesion with a volume of 4.8 cm^3 and contrast enhancement in the periphery. Two further periventricular FLAIR and T2-hyperintensive, contrast-naïve, supratentorial lesions were found (Fig. 1K and L). MRI spectroscopy could not clearly distinguish an inflammatory from a neoplastic process. Single voxel spectroscopy and chemical shift imaging revealed a slightly increased Choline/Creatine ratio (2.58) and small lactate double-peaks indicative for an inflammatory process. CSF analysis showed slight mononuclear pleocytosis (5/µl), elevated protein content (0.9 g/l), normal lactate and glucose, and was negative for oligoclonal bands and neoplastic cells. Serologies against HIV, hepatitis B and C virus, and *Treponema pallidum* were negative. Vasculitis screening (antinuclear antibodies and anti-dsDNS-antibodies), limbic antibodies (NMDA-receptor, AMPA-1-receptor, AMPA-2-receptor, LGI-1, CASPR-2, GABA-receptor B1/2), and serum protein electrophoresis were unremarkable. Neuropsychological testing revealed pronounced executive dysfunction, reduced visuoconstructive skills, attentional deficits, and speech problems with word finding difficulties, echolalia, and non-fluent speech production (Tables 2 and 3). The patient's cognitive performance in the Montreal Cognitive Assessment (MoCA) test showed visuospatial and executive dysfunctions as well as attentional and speech deficits (22/30 points) (Table 1).

A demyelinating process was assumed and CIS diagnosed based on the MRI lesions, CSF pleocytosis, and exclusion of other inflammatory or neoplastic causes (Polman et al., 2011).

Initial treatment with high dose i.v. methylprednisolone therapy (1 g/d) had no effect on clinical symptoms. Repetition of i.v. methylprednisolone (2 g/d for 3 days) again did not improve the patient's symptoms. Subsequently, he underwent plasmapheresis (7 cycles, 1.5 l plasmavolumes exchanged per cycle). With a delay of few days, gait ataxia, ocular motility disorder, and double vision clearly improved, whereas manual fine motor skills and dysexecutive functions remained unchanged. His girlfriend however reported a definite improvement of adynamia, motivation, and decision making.

2.2. Follow-up during 6 months

Follow-up MRI after 3 weeks showed a slight increase of the ponto-mesencephalic lesion volume and a regression of contrast enhancement (Fig. 1E and F).

The patient was transferred to a neurorehabilitation unit where a temporary worsening of gait ataxia and speech problems was noticed. An MRI 6 weeks after the first one documented no change in lesion volume and contrast enhancement.

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