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Cerebellar mutism

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ABSTRACT

Cerebellar mutism occurs in about 25% of children following posterior fossa tumor surgery. It is usually accompanied by other neurological and behavioral disturbances. Mutism is transient in nature lasting several days to months and is frequently followed by dysarthria. In addition, impairment of language and other neuropsychological functions can be found after long term follow up in the majority of patients. The pathophysiological background of mutism may be higher speech dysfunction mediated by crossed cerebello-cerebral diaschisis which is frequently found during the mute period. Foremost injury to the bilateral dentatothalamocortical tract appears to be critical for the development of cerebello-cerebral diaschisis and subsequent mutism. Direct cerebellar injury is the likely reason for persisting deficits after the mute period. Minimization of injury to the dentatothalamocortical tract during surgery may be promising in the prevention of mutism. While there is no established treatment of mutism, early speech and rehabilitation therapy is recommended.

1. Introduction

Cerebellar mutism refers to speechlessness that evolves after cerebellar injury. Postoperative mutism in children following cerebellar surgery was first recognized in the 1970s (Hirsch, Renier, Czernichow, Benveniste, & Pierre-Kahn, 1979; Stein, Fraser, & Tenner, 1972). In 1985 Rekate et al. drew attention to this condition in an article presenting six cases of cerebellar mutism after posterior fossa surgery (Rekate, Grubb, Aram, Hahn, & Ratcheson, 1985). So far over 400 cases have been reported in the literature (Gudrunardottir, Sehested, Juhler, & Schmiegelow, 2011 for recent review). Cerebellar mutism occurs rarely isolated but often together with other neurological, emotional and behavioral disturbances. Emphasizing on the complex character comprising multiple symptoms many authors refer to cerebellar mutism as the cerebellar mutism syndrome or posterior fossa syndrome. Most frequently cerebellar mutism can be found in children after cerebellar tumor surgery (Fig. 1). However, it may also be present in other conditions affecting primarily the cerebellum or in adults (Ildan et al., 2002). Brain tumors represent the most common disease group of solid tumors in childhood and can be most frequently found in the cerebellum (Kaatsch, Rickert, Kühl, Schüz, & Michaelis, 2001). Mutism is a devastating side effect of tumor treatment and occurs with comparatively high incidence. Although it resolves spontaneously other symptoms like dysarthria and language disorders can persist. In addition, cognitive and behavioral disturbances occuring during the mute phase may also persist to some degree.

Direct cerebellar injury and remote neocortical dysfunction mediated by cerebello-cerebral diaschisis may contribute differentially to symptoms of cerebellar mutism over time. In particular, neuroimaging studies have increased our knowledge about the pathophysiology of cerebellar mutism and identified lesions of the dentate nucleus and its efferent output as a critical predictive factor for the latter development of mutism. The question why mutism manifests after a latency period remains unanswered.

2. Epidemiology

The incidence of cerebellar mutism syndrome after posterior fossa surgery in children ranges between 11% and 29% (Gudrunardottir et al., 2011). Recent prospective studies recruiting substantial pediatric patient numbers found an incidence of 27.7% (Catsman-Berrevoets & Aarsen, 2010) and 24% (Robertson et al., 2006). Mean ages were 6–7 years in these studies. However, neither sex nor age was predictive for the development of cerebellar mutism. Highest incidences were found following medulloblastoma surgery (40%) compared to pilocytic astrocytoma (16%) or ependymoma (4%) surgery (Catsman-Berrevoets & Aarsen, 2010). In a study with a smaller patient population pre-surgical language impairment was associated with post-surgical occurrence of mutism (Di Rocco et al., 2011). Based on case reports rare causes of cerebellar mutism syndrome comprise trauma (Fujisawa et al., 2005), stroke (Baillieux, Weyns, Paquier, De Deyn, & Mariën, 2007) and inflammation (Papavasiliou, Kotsalis, & Trakadas, 2004). Although exact figures are lacking in adults, postoperative cerebellar mutism syndrome is considerably less frequent and was present in roughly 1% (6 out of $n = 500$; Dubey et al., 2009).

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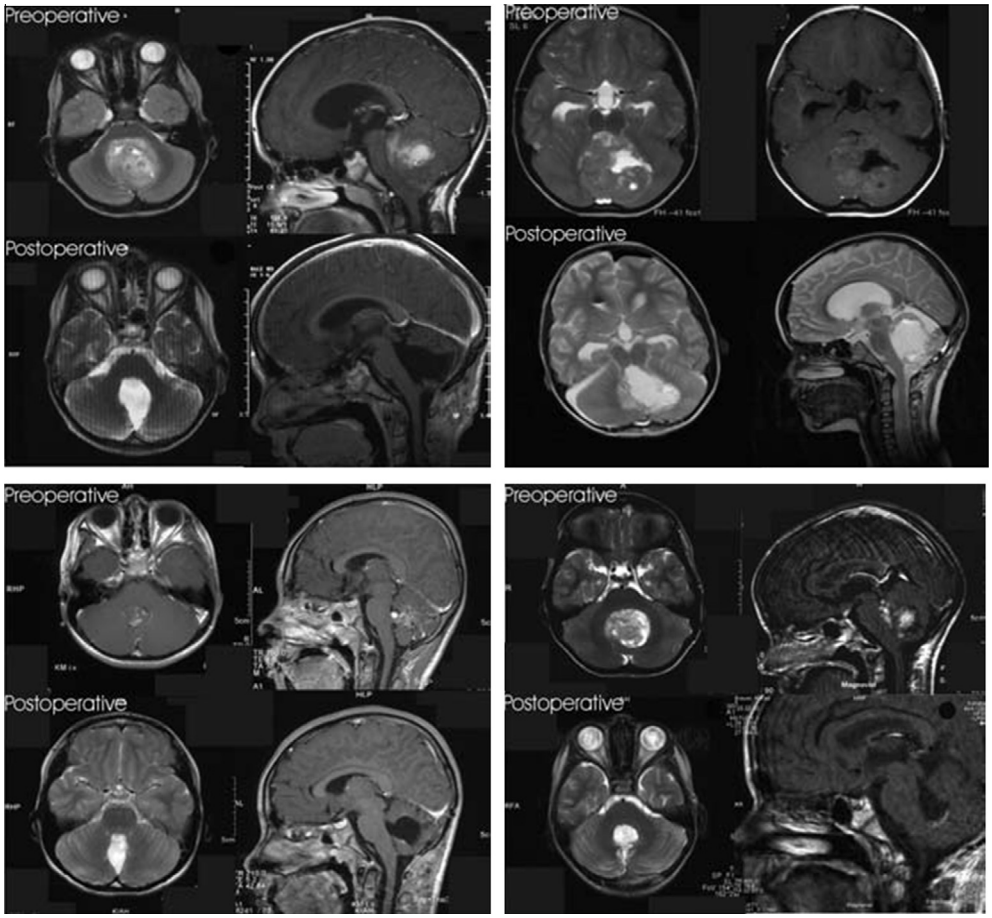

Fig. 1. Individual lesions of children with mutism based on Fig. 2 in Ozimek et al. (2004), used with kind permission of Springer Science + Business Media.

3. Clinical presentation and time course

3.1. Time course

The time course of cerebellar mutism syndrome has three characteristic phases. Firstly, cerebellar mutism is not present directly after surgery, but develops within a time interval of hours up to several days after the surgical intervention (Robertson et al., 2006; Wells, Walsh, Khademian, Keating, & Packer, 2008). Secondly, mutism is always transient. The duration is variable, lasting from a few days to several months (Ildan et al., 2002). Recovery of mutism occurs spontaneously. Finally, after the mutistic phase symptoms of motor speech and language disorders, cognitive, emotional and behavioral disorders remain to various extends.

3.2. Mutistic phase

During the mutistic phase high-pitched crying is the only form of vocalization (Robertson et al., 2006). Various concomitant neurological symptoms may be present suggestive of cerebellar and/or brainstem injury. These comprise ataxia, involuntary eyelid closure, pyramidal tract signs, horizontal gaze paralysis, cranial nerve palsies and oropharyngeal dyspraxia (Kirk, Howard, & Scott, 1995; Wells et al., 2008). Besides, cortical blindness has been reported (Catsman-Berrevoets & Aarsen, 2010).

Behavioral and emotional disturbances most frequently encompass emotional lability, apathy and autistic like behavior (Catsman-Berrevoets & Aarsen, 2010; Pollack, Polinko, Albright, Towbin, & Fitz, 1995). Bizarre personality changes may manifest as forced laughing or crying. Also, loss of bladder and bowel control is often found (Catsman-Berrevoets & Aarsen, 2010).

3.3. Postmutistic phase

Riva and Giorgi found that mutism dissolves in different ways (Ozimek et al., 2004; Riva & Giorgi, 2000). (1) Mutism is followed by dysarthria without signs of higher language disorders (mutism and subsequent dysarthria, van Dongen, Catsman-Berrevoets, & van Mourik, 1994). (2) Mutism is followed by a language disorder without dysarthria. (3) Mutism is followed by behavioral disturbances. The three subtypes are not mutually exclusive and may depend on lesion localization as outlined below.

This view is not unchallenged since other authors proposed that dysarthria is a general feature occurring in virtually every patient with resolving cerebellar mutism (De Smet et al., 2007). In their meta-analysis reviewing 167 cases of post-surgical mutism the vast majority ($n = 165$, 98.8%) of patients were found to exhibit dysarthria. Authors were concerned that dysarthria might have been missed in the remaining two patients because of the presence of predominant behavioral disorders or lack of spontaneous speech in these patients.

Differences in evaluation criteria and study populations may explain inconsistent findings of previous studies. It would be helpful to define universal criteria on how to evaluate speech and language outcome for future studies.

3.3.1. Dysarthria

A detailed analysis of post-mutistic speech features revealed that classical signs of ataxic dysarthria according to Darley, Aron-