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CEREBELLAR MUTISM
SEREBELLAR MUTİZM

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Abstract

Cerebellar mutism has been a well-known clinical entity that develops in a subset of patients who have undergone resection of posterior fossa tumors. It is characterized by severely diminished or absent speech output as well as other neurological, cognitive, and behavioral impairments. Though increasing numbers of case reports and literature reviews that indicate the cerebellar mutism, the mechanism of occurrence and best therapeutic approaches are not established. This article reviews current status of this devastating complication with respect to epidemiology, anatomical substrate, pathophysiology, risk factors, treatment options, prognosis and prevention.

Keywords: Cerebellar, complication, mutism, posterior fossa, tumor

Özet

Serebellar mutizm arka çukur tümörlerinin çıkarılması sonrasında bir grup hastada geliştiği bilinen bir klinik tablodur. Bu tablo, konuşmanın bozulması veya hiç konuşamama ile birlikte diğer nörolojik, bilişsel ve davranışsal bozukluklar ile karakterizedir. Literatürde serebellar mutizmle igili vaka raporları ve derlemelerdeki artışa rağmen oluşum mekanizması ve etkin tedavi yaklaşımları henüz belirlenememiştir. Bu makalede, bu yıkıcı komplikasyonun görülme sıklığı, anatomik temeli, patofizyolojisi, risk faktörleri, tedavi seçenekleri, sonucu ve korunması açısından güncel durumu gözden geçirilmiştir.

Anahtar Kelimeler: arka çukur, komplikasyon, mutizm, serebellar, tümör

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1. Introduction

Cerebellar mutism (CM) has been defined as muteness following lesion of the cerebellum as opposed to the cerebrum or the lower cranial nerves. It is characterized by delayed onset, limited duration, and usually long-term linguistic sequellae. It occurs rarely isolated but often together with other neurological, emotional and behavioral disturbances. Cerebellar mutism most frequently occurs in pediatric population following surgical treatment of posterior fossa tumors (Gudrunardottir et al., 2011, Küper&Timmann, 2013). It can also be seen following trauma, vascular events, infection, pineal gland tumor removal of pineal gland tumors, or in adults (Baillieux et al., 2007, Ellis et al, 2011, Ersahin et al., 1997, Frassanito et al., 2009, Ildan et al., 2002, Papavasiliou et al., 2004). It was first anecdotally reported by Stein et al. in 1972, and later by Hirsch et al. and Pierre-Kahn et al. However, Yonemasu and Rekate et al. are generally considered the first who have reported this peculiar syndrome in more detail. Since 1985, more than 400 cases of CM have been described in the literature (Gudrunardottir et al., 2011, Küper&Timmann, 2013, Pitsika&Tsitouras, 2013).

2. Epidemiology

The incidence of cerebellar mutism after posterior fossa surgery in children is reported to range between 8%-39% in the recent literature (Pitsika&Tsitouras, 2013). Mean ages were 6-7 years in these reports. In adults, the incidence of postoperative cerebellar mutism is less frequent and was present in 1% of the reported cases. Brainstem involvement by the tumor, tumor type, midline location and preoperative language impairment were determined as risk factors for development of cerebellar mutism (Di Rocco et al., 2011, Law et al., 2012, Tasdemiroglu et al., 2011).

3. Pathophysiology

The exact reason for cerebellar mutism has not been agreed upon. Bilateral interruption of the dentato-thalamo-cortical pathway is suggested to be the main cause of cerebellar mutism. Interruption of this pathway results in cerebro-cerebellar diaschisis which has been described as a temporary functional deactivation of an intact brain region remote from the lesion area. Delayed onset and resolution of the mutism suggest that a secondary pathophysiological mechanism initiated by the tumor resection mediates the cerebellar mutism. Proposed mechanisms involve cerebellar perfusional disturbances, postoperative edema, transient dysregulation of neurotransmitter release and functional disruption of the white matter bundles containing efferent axons within the superior cerebellar peduncles (Gudrunardottir et al.,

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2011, Ozgur et al., 2006, Pollack et al., 1995, Puget et al., 2009).

4. Clinical features

Cerebellar mutism has three characteristic features. First, cerebellar mutism is not present directly after surgery, but develops within a time interval of hours to several days after the surgical intervention (Robertson et al., 2006). Second, 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 and can occasionally be rapid and complete (Gelabert-González&Fernández-Villa, 2001). Finally, after the mutistic phase, symptoms of motor speech and language impairments, cognitive, emotional and behavioral disturbances remain to various extends.

5. Treatment

There is no established treatment modality exists for cerebellar mutism. Pharmacological and speech therapy has been used to reverse the symptoms of mutism. Bromocriptine, zolpidem and fluoxetine were found to beneficial in sporadic cases but have not been systematically assessed (Akhaddar et al., 2012, Caner et al., 1999, Shyu et al, 2011). But there is a complete lack of trials that explore the efficacy of both pharmacological and speech therapy during the recovery phase.

6. Prevention

Preventive strategies to protect dentate nucleus and superior cerebellar vermis can be effective for reducing the incidence of cerebellar mutism. Several surgical strategies are recommended to avoid from this complication. Piecemeal removal of the tumor, access to the tumor without splitting the vermis by telovelar approach and short-lasting retraction of the cerebellar vermis were reported to provide significant advantages in the prevention of cerebellar mutism (Aguiar et al., 1995, Frassanito et al., 2009, Mussi&Rhoton, 2000).

7. Prognosis

Prognosis is variable in patients suffering from cerebellar mutism. The duration of symptoms after surgery seems to be correlated with functional prognosis. If the symptoms persist for more than four weeks, patients will have a high risk of suffering language dysfunction at postoperative 1st year (Robertson et al., 2006). Patients have improved quality of life if they receive pharmacological, speech therapy and individual educational support with psychiatric examination. There are no reports in the literature in regards to a recurrence of cerebellar mutism with subsequent surgeries.

8. Conclusion

Over the past 30 years, more than 400 cases of mutism and associated behavioral and personality changes have been reported after the removal of posterior fossa tumors. Advanced neuroimaging techniques could contribute to identification of high-risk patients preoperatively and allow for more effective surgical planning that should focus on maximal tumor resection with minimal risk to important neural structures. Properly designed multicenter trials are needed to provide stronger evidence regarding effective prevention of cerebellar mutism and the best therapeutic approaches for such patients with a combination of pharmacological agents and multidisciplinary speech and behavior augmentation.

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