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Cerebellar mutism syndrome after surgical resection of posterior fossa neoplastic lesions

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Abstract

Cerebellar Mutism Syndrome (CMS) is a well-described clinical entity that complicates surgeries for posterior fossa tumours; more so in children than adults. This review focuses on the current understanding of CMS, its incidence and risk factors. Incidence showed a variable range in retrospective studies due to variety of definitions. Risk factors can be classified as either modifiable including surgical technique, or non-modifiable which include tumour related factors. A positive correlation has been associated between tumour pathology, brain stem invasion and size of tumour at time of presentation with development of CMS.

Keywords: Pediatric brain tumors, cerebellar mutism syndrome, posterior fossa syndrome

Introduction

Cerebellar mutism syndrome (CMS), often used interchangeably with posterior fossa syndrome (PFS), is not an uncommon complication, following surgical resection of tumours located in the cerebellum or fourth ventricle.[1] Cerebellar mutism was initially described as 'akinetic mutism' by Daly and Love in 1958 after the removal of a cerebellar tumour in a child. Historically, it comprised of complete loss of speech that was not associated with cranial nerve palsies, long tract signs or alteration of consciousness; and typically occurred within few hours to several days after posterior fossa tumour surgery.[2] However, CMS is now used to describe a broader clinical condition which comprises of transient speech impairment, decreased muscle tone, irritability, ataxia and psychosis mostly in children and young adults, usually following surgery for posterior fossa tumours.[3]

The incidence of CMS has been reported in 8% to 32% children, however, there is significant discrepancy in this number because of high variability of definitions of CMS used by different authors.[4] Patients show inconsistent recovery in a variable amount of time, with some regaining the lost function completely, but most having some residual dysarthria, ataxia or intellectual disability. CMS is commonly believed to be caused by destruction of the reciprocal cerebello-cerebral pathway and bilateral dentate nuclei. This disrupts dentate-thalamo-cerebral tract, which impairs corresponding areas of the cerebral cortex.[4] Risk factors are mostly non-modifiable and include larger tumours (>5cm in diameter) located in the midline, brainstem invasion and pre-operative language deficits.[4] It is commonly associated with medulloblastoma (40%), ependymomas (20%) and astrocytomas (15%).[4]

Owing to advancement in surgical techniques, and early

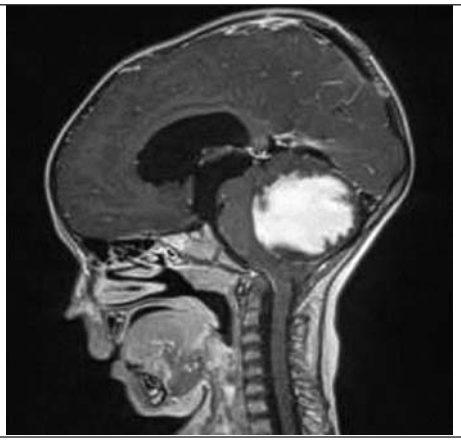

Figure 1A: Pre-operative T1 post-contrast sagittal section of MRI brain of a 3 years old boy showing a homogenously contrast enhancing lesion in the posterior fossa, causing hydrocephalus and significant mass effect on brainstem.

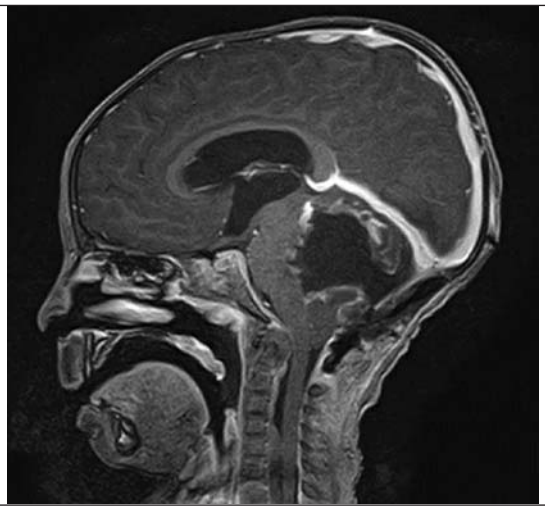

Figure 1B: PPost-operative T1 post-contrast sagittal section of MRI brain of the same patient showing resection cavity filled with fluid, and interval reduction in mass effect on brainstem and cerebellum as well as hydrocephalus.

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diagnosis of posterior fossa tumours due to wider availability of magnetic resonance imaging (MRI), there has been an increase in posterior fossa surgeries and attempts for gross total resection, as one of the most important prognostic indicators in patients with posterior fossa tumours is extent of surgical resection. This theoretically increases the risk of developing CMS. Herein, we have reviewed the existing English language literature on CMS after surgical resection of posterior fossa brain tumours.

Review of Evidence

We searched 'cerebellar mutism syndrome', akinetic mutism' and 'posterior fossa syndrome' on PubMed and Google scholar database. Despite a high reported incidence, the condition has not been very commonly studied. Robertson et al., published a large prospective multicenter study between 1994 and 2000, and studied 450 patients who had undergone craniotomy and resection of posterior fossa medulloblastoma.[5] They reported incidence of CMS in 107 patients (24%) within two months from surgery, and classified it into severe (43%), moderate (49%) and mild (8%) using a CMS survey that they had specifically designed for this study. The only significant risk factor with a positive correlation with CMS was brainstem invasion, whereas radical surgical resection was not a significant risk factor. They also found variable extent of recovery in most patients, but fewer patients attained complete recovery up to 1 year after surgery. Korah et al., in their retrospective series reviewed 63 children who had undergone surgical resection of medulloblastoma.[6] Their median follow-up time was 7 years, and incidence of CMS was 29% (18 patients). This study reported younger age, gross total resection, midline location of tumour and brainstem invasion to be statistically significant predictors of CMS.

Another case series of 32 children who underwent resection of cerebellar tumours was published by Kotil et al., who reported the occurrence of CMS in 10 children (32%).[7] They found larger tumour size and midline location to be significant risk factors for developing CMS. Although CMS or PFS have been predominantly described to occur in children, some cases in adults have also been reported. In 2019, Shamov et al., reported post-operative CMS in a 25 years old female patient after resection of 4th ventricular medulloblastoma, which was the eighth such case in literature.[8] They attributed excessive use of bipolar cautery and ultrasonic aspirator in close proximity to the cerebellum to be possible causes.

Cobourn et al., had performed a retrospective review of 65 patients who had undergone surgical resection of posterior fossa tumours at their center, and studied relation between important surgical steps and CMS.[9] They reported incidence of CMS in 7 of their patients (10.8%) and found degree of cerebellar retraction and incision of vermis to be significant risk factors for CMS. In the light of their results, they recommended preference of telo-velar approach over trans-vermian approach for posterior fossa tumour resection. In another recent study, Dhaenens et al., had retrospectively reviewed MRI scans of 121 children with posterior fossa tumour who underwent surgery, and attempted to design and validate a preoperative risk prediction model for post-operative CMS.[10] Twenty six percent of their patients had developed CMS. In their radiological risk prediction model they included Evan's ratio, distance and angle between basilar artery and tumour and depth of invasion/compression and area of contact between brainstem and the tumour. Their proposed model had a sensitivity of 97% and specificity of 84% in pre-operative prediction of CMS.[10]

Conclusion

The review of available literature shows CMS/PFS to be an important complication following posterior fossa tumour resection. Important modifiable risk factor may include surgical technique that avoids manipulation of vermis. However, more common factors responsible for this condition include tumour location, pathology and size.

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