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Posterior fossa tumor resection in children: a case report and the role of intraoperative neurophysiologic monitoring

Charles Kondageski[1], Humberto Kluge Schroeder[1], Antonio Cesar de Melo Mussi[2], Cezar Massaru Guiotoku[1], Marcela de Moraes Barros Sousa[1], Jean Costa Nunes[3]

[1] Hospital Santa Catarina de Blumenau. Blumenau, Santa Catarina, Brazil

[2] Celso Ramos Governor's Hospital, Florianópolis, Santa Catarina, Brazil

[3] Neurodiagnostics Brazil, Florianópolis, Santa Catarina, Brazil

✉ Charles Kondageski, MD

e-mail: chskki@yahoo.com.br

Available at:
http://www.archpedneurosurg.com.br/

Introduction/Background: Surgery plays a crucial role in the surgical management of posterior fossa tumors in children, but maximizing the resection while preserving function remains challenging. Along with other tools available to the surgeon, intraoperative neuromonitoring aims at improving safety during these complex interventions. This case report highlights the role of real-time intraoperative neuromonitoring during surgery and its impact on the surgeon's decision-making process.

Case report: We present the case of a 10-year-old boy with a fourth ventricle medulloblastoma, who underwent microneurosurgical resection under intraoperative neuromonitoring. The use of neurophysiological monitoring yielded relevant information during the tumor dissection of the floor of the fourth ventricle, the most crucial step of surgery, thus helping the surgeon to change tactics to minimize long-term neurological deficits.

Conclusion: In combination with meticulous microneurosurgery techniques, modern anesthetic regimens, and ultrasonic aspiration, intraoperative neuromonitoring adds up to the surgeon's armamentarium for increasing safety and improving outcomes following surgery for pediatric patients bearing posterior fossa tumors.

Keywords: posterior fossa tumors, children, medulloblastoma, intraoperative neuromonitoring

INTRODUCTION

Posterior fossa is the most common site for pediatric central nervous system tumors in children, while medulloblastomas, astrocytomas, and ependymomas are the most prevalent histological types [1,2]. Management of these cases is challenging, due to the tumor location and its biology, posing major risks and obstacles for surgery and adjuvant therapies, especially in pediatrics [1,3,4,5]. Long-term survival rates, nowadays, stand at around 70% of all patients, but, for certain subgroups, sonic hedgehog patients (group 3) as an example, the prognosis is worse, with a 10-year survival rate of 40% to 50% [6]. Even though surgery plays a central role in the management of these lesions, it carries a high risk of permanent disability and poor quality of life [3,4,5]. Various surgical tools, such as the microscope, ultrasound aspirator, neuronavigation, fluorescence guidance, and intraoperative neurophysiological monitoring (IONM) assist the neurosurgeon with the task of achieving the safest maximum resection on these particular cases [7]. The aim of this case report is to bring awareness to the complexities of performing surgery in the posterior fossa of children and the potential of increasing surgical efficiency and safety by using adjunct surgical tools such as IONM.

CASE REPORT

A 10-year-old boy presented with a 3-week history of progressive convergent strabismus, associated with morning nausea episodes and gait ataxia. Magnetic resonance of the brain disclosed a heterogenous space-occupying lesion centered in the fourth ventricle, measuring 4.9 x 4.7 x 4.3 cm, with lobulated contours and apparent extension through the foramens of Magendie and Luschka. There was no clear cleavage layer with the surrounding walls of the ventricle, and, in addition, the lesion exerted regional mass effect, dislocating the brainstem anteriorly. The mass presented a predominant low signal on T1, high signal on FLAIR and T2, and peripheral areas of diffusion restriction, suggesting hypercellularity. In addition, there was an enlargement of the supratentorial ventricles (Figure 1). Further imaging of the entire neuraxis did not display other lesions.

He was then operated on in a prone position and under intraoperative neuromonitoring. Initially, a right parietal

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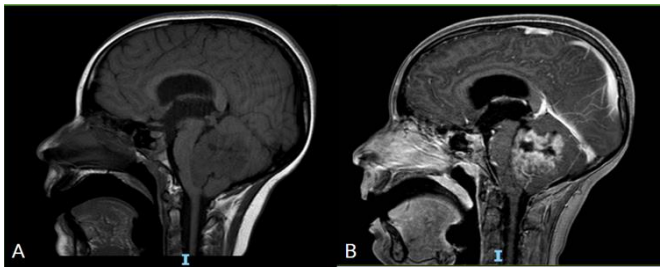

Figure 1 - Preoperative MRI images depicting a space occupying lesion within the fourth ventricle. A) Pre contrast sagittal T1-weighted image; B) Post contrast sagittal T1-weighted image

external ventricular drain (EVD) was placed for later cerebrospinal fluid (CSF) drainage. Surgery was performed through a standard suboccipital craniotomy and resection of the posterior arch of C1. Just before a "Y" shaped durotomy, 25 mL of CSF was drained through the EVD. At the subdural space exposure, a small portion of the lesion was noticeable at the level of the enlarged Magendie foramen. Surgery went on with a telovelar approach under microscopic technique, exposing most of the fourth ventricle and the lesion within. Resection started with a debulking achieved with ultrasonic aspiration and, after a good reduction in its size, the surgeon worked all around the lesion, dissecting it from the surrounding tissues when possible. While the dissection was performed at the level of the facial colliculus on the left, the neurophysiology team gave a warning regarding abnormal left mentalis electromyographic activity, concurrent with a decrease in the amplitude of the corticobulbar potentials recorded from the mentalis muscle (see IONM findings below). The surgeon stopped resection at that moment, and when resuming with the resection, the abnormal IONM findings returned. After a brief discussion with the entire team he then decided to leave a portion of the lesion in relation to the floor of the fourth ventricle (Figure 2). The other motor, auditory and sensory potentials remained as expected, with no warning criteria met according to the neurophysiologist.. After revising the hemostasis, dura was primarily closed as well as the other surgical layers. The patient was awakened from anesthesia, extubated, and then transferred to the pediatric intensive care unit. The external drainage was kept open at the 20 cm height from the tragus for 24 hours and then removed after 72 hours.

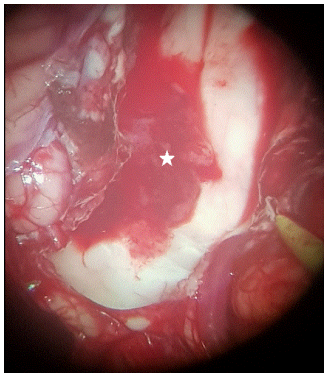

Figure 2. Intraoperative photograph showing the tumor on the floor of the fourth ventricle during dissection (white star)

On the first postoperative day a left facial palsy, House Brackmann grade 4, was noticeable, as well as a persistent convergent strabismus. Post operative MRI obtained on the second day after surgery disclosed a decrease in the ventricles size and a subtotal resection of the lesion, with a residual layer of approximately 2 cm3 on the floor of the fourth ventricle (FIgure 3). On the third postoperative day, the patient developed speech arrest, hypotonia and motor incoordination, all compatible with akinetic mutism. Pathology analysis disclosed a medulloblastoma WHO grade 4, with molecular analysis defining a non-WNT/non SHH, group 4, while CSF was clear of pathological cells. The patient was then submitted to craniospinal radiation (36 Gy neuroaxis + 18 Gy posterior fossa boost) followed by initiation of the COG chemotherapy protocol. After 4 weeks the mutism symptoms started to subside and at the last follow up (5 months after surgery) the child fully recovered his speech skills, facial function and extrinsic ocular motility, though there still remained some mild ataxia and moderate dysgraphia. He´s currently under the chemotherapy protocol and at his last MRI scans there was no evidence of the disease.

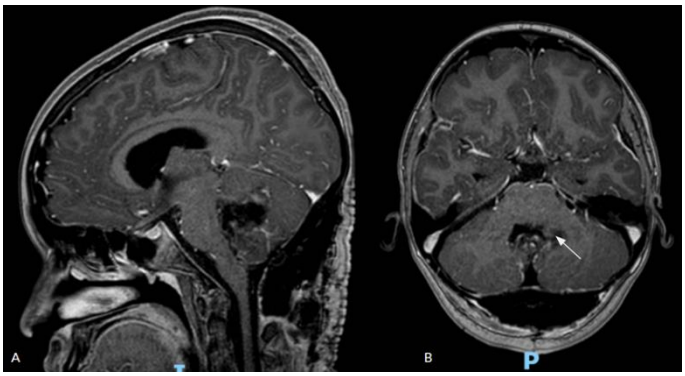

Figure 3 - Early postoperative MRI images. A) Sagittal view, B) Axial view. White arrow: resection cavity with a small residue within the fourth ventricle

Intraoperative monitoring procedures and findings

IONM procedures

All recordings were obtained by a neurophysiologist using a Cadwell Iomax device (Cadwell Labs, USA). Total intravenous anesthesia (TIVA) was administered throughout the procedure, including the use of propofol and remifentanil. Intravenous rocuronium was only administered for intubation purposes and no inhalation agents were used. The intraoperative neurophysiological monitoring was multimodal, including the following techniques: free-run electromyography (EMG), somatosensory evoked potentials (SSEPs), transcranial motor evoked potentials (TcMEPs), corticobulbar evoked potentials (CbMEPs), and brainstem auditory evoked potentials (BAEPs). While under general anesthesia and just before positioning the patient, the electrodes were placed as

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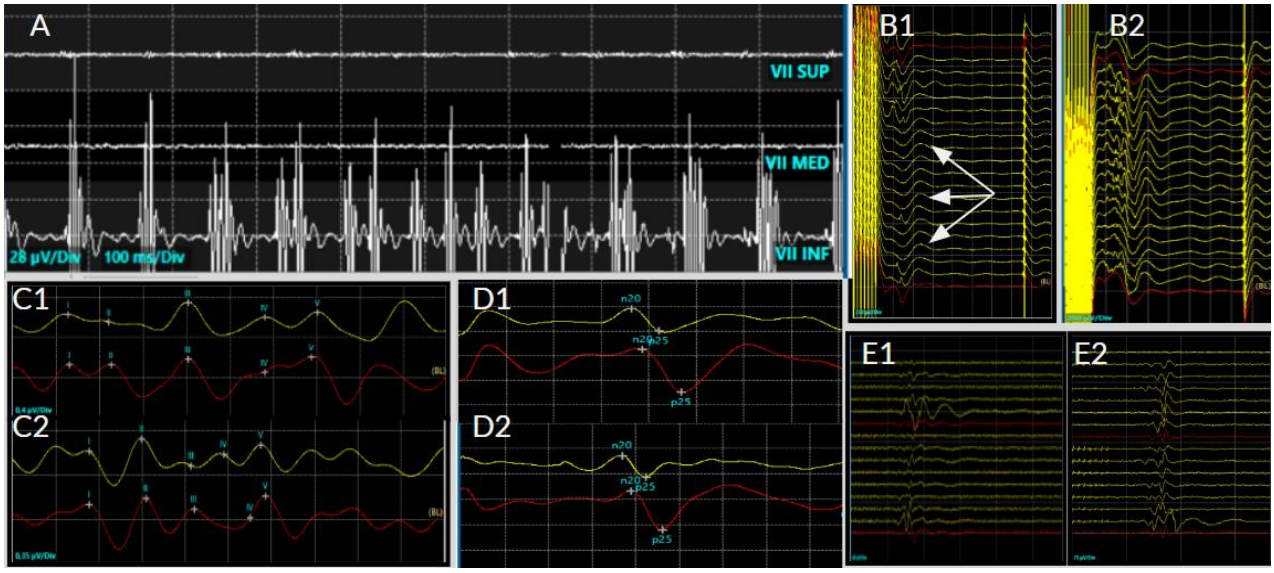

Figure 4 - A) Free-run electromyography displaying abnormal neurotonic discharges in the left mentalis muscle; B1) Left CbMEPs demonstrating a relevant decrease in the amount of phases e decreased amplitudes in the left mentalis muscle (triple white arrow), B2) Right CbMEPs; C1/C2) Left and right BAEPs; D1/D2) Left and right SSEPs and E1/E2) Left and right MEPs registered throughout the procedure.

follows: corkscrews (Spes medica, Italy) in the scalp at Fz, Cz', C3', C4', M3, M4, M6, M0, A1, A2, twisted paired stainless-steel subdermal (Spes medica, Italy) within the following muscles: lateral rectus of the eye, orbicularis oculi, orbicularis oris, mentalis, endotracheal laryngeal electrode, trapezium, tongue, abductor digiti minimi, and abductor hallucis all bilaterally, as well as a pair of stimulating surface electrodes (Spes medica, Italy) over the ulnar nerves bilaterally at the wrists. Bite block was used as usual. After the patient was positioned in ventral decubitus, baseline recordings were obtained for all IONM modalities according to the standard stimulation and registration parameters as described elsewhere [8,9,10].

IONM findings

During the approach (skin incision and soft tissue dissection, craniotomy, durotomy, and telovelar dissection), no abnormal recordings were found. At the time of the tumor resection from the fourth ventricle floor, frequent EMG discharges were recorded in the left mentalis muscle (Figure 4A). At the same time, a progressive decrease in the amplitude of the CbMEPs potentials was noticed in the left mentalis (Figure 4B1). This warranted changes in surgical maneuvers, such as stopping dissection at that time, while irrigating the surgical field with warm saline solution. After the abnormal EMG subsided, dissection resumed, until new EMG discharges appeared again, and the facial CbMEPs were eventually not recordable. Surgery was stopped again, and saline solution was used another time. Mean arterial blood pressure was within normal values, as well as body temperature measured from the rectum. After 5 minutes, facial CbMEPs started to recover to 20% to their original amplitude (Figure 4B1). The attending surgeon, at that time, then decided to stop resection, leaving a small tumor residue within the floor of the fourth ventricle in proximity to the left facial colliculus. There were no changes seen in the TcMEPs (Figures 4E1 e 4E2) and BAEPs (Figures 4C1 e 4C2) during the surgical procedure, while the SSEPs were decreased in amplitude by 50% but with latencies maintained bilaterally (Figure 4D1 e 4D2). The potentials were recorded until the end of the dural closure. Electrodes and the bite block were removed at the end of surgery.

DISCUSSION

Posterior fossa tumors are most prevalent in pediatric patients when compared to the adult population, and still remain a challenge to the multidisciplinary team, despite the latest developments in surgery, anesthetics, neurophysiology, pathology, radiotherapy, chemotherapy, and neurorehabilitation [1,2,4,5,6,10]. Outcomes following treatment for these patients have been much improved, but, nonetheless, for some histological subtypes and clinical presentations, prognosis still remains dismal [4,6]. When the quality of life is considered beyond survivorship, children and their families carry the burden of neurological disabilities and systemic issues related to the lesion itself, surgery, and adjunct therapies [11].

Surgery is essential for the management of posterior tumors since prognosis is directly affected by the amount of lesion that is removed or left behind [4,5]. Surgical resection of these tumors is complicated by the tumor location and its vicinity, which includes delicate, highly functional, and vital vasculo-nervous tissues, such as the brainstem, spinal cord,

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cranial nerves, cerebellum, and the vertebrobasilar vascular system. Not rarely, those tumors adhere and invade the surrounding tissues, making resections potentially hazardous [1,3,4,5]. In the present case report, there was a suspicion according to the preoperative MRI imaging, and confirmed by intraoperative findings, that the lesion could be invasive/highly adherent to the fourth ventricle walls, including its floor.

Over the course of the last few decades, many different surgical tools have been added to the surgeons' armamentarium, such as the microscope, neuronavigation, ultrasound imaging, ultrasonic aspiration, fluorescence-guided surgery (FGS), and intraoperative neurophysiologic monitoring (IONM) [7,9,12,13]. These technologies and techniques, when combined, help the neurosurgeon with the complex task of resecting a posterior fossa lesion at its entirety, whenever achievable, and with the maximum safety possible [3,6,7].

Intraoperative neurophysiological monitoring (IONM) techniques have emerged as diagnostic tools that make it possible to assess the functional integrity, in real time, of some of the central and peripheral nervous system pathways, and thus aid the surgeon and anesthesiologist to protect those areas by promptly modifying the techniques when necessary [4,8,12]. Along with the above mentioned surgical tools, it promotes higher rates of total resections while turning them safer [9]. To increase the sensitivity and specificity of the IONM, multiple monitoring modalities need to be combined, such as somatosensory evoked potentials (SSEPs), motor evoked potentials (MEPs), corticobulbar motor evoked potentials (CbMEPs), brainstem auditory evoked potentials (BAEPs), free-run electromyography (EMG), and triggered EMG [8,9,13]. According to the surgery being performed, its risks, and its goals, the techniques are chosen in agreement with the whole team [8,9,10,12]. For this particular case, IONM was discussed with the team and tailored to cover the nervous system structures that are anticipated to be under risk. Namely, EMG, TEMG, CbMEPs, BAEPs, MEPs, and SSEPs were elected, as well as electroencephalogram and train of four to assist with sedation depth and neuromuscular block status. Warning criteria have been defined for most of those tests (most often an increase in latencies and a decrease in amplitudes) [8]. Whenever these criteria are met, this information is taken into account by the entire team and then combined with the surgeon's and anesthesiologist's views so that surgical maneuvers and anesthesiological techniques are revised. If necessary, surgical manipulation at a given site is suspended, and then actions are taken to try to restore the potentials, which, in turn, should improve the patient's outcome [3,8,9,12].

During the microdissection of the lesion from the floor of the fourth ventricle in the present case, intraoperative neuromonitoring yielded warnings from abundant free-run EMG discharges in the mentalis muscle in combination with an abrupt decrease in the amplitude of the CbMEPs registered in the same muscle (Figure 4A and 4B1). This information was translated as a warning criterion that infers some sort of dysfunction and the likelihood of persistent facial palsy, and so, the surgeon stopped surgery for a while, irrigating the surgical field with warm saline solution. After the potentials started to recover, dissection was resumed, but ultimately had to be stopped due to further reductions in CbMEPs and EMG discharges within the left mentalis. The surgeon then decided to leave a small portion of the tumor in the fourth ventricle floor.

Despite the ever-growing development of new techniques and their widespread use, it should be taken into account that IONM currently is not able to assess all neural pathways that may be under risk at a given surgical site and time, as well as that it carries sensitivities and specificities that are not 100%. So while largely adding safety to procedures, IONM tests may portray false positives and false negatives that may affect outcomes, varying according to the type of surgery, the experience of the entire team, and IONM techniques being performed [9]. Most often, during posterior fossa surgery, several neural pathways and structures can be assessed with IONM techniques, such as the motor pathways to the limbs, corticobulbar pathways, including facial and lower cranial nerve function, ocular motricity function, cochlear nerve function, and somatosensory pathways. Several techniques, in isolation or combined, are employed to inform the surgical team about the integrity of those structures, but each one of those assessments carries different sensitivities and specificities with regards to predicting function in the medium term. As an example, the corticobulbar MEPs have a sensitivity of 89% and specificity of 99% when considering a 65% decrease in amplitude [13]. For the procedure reported here, despite meticulous telovelar approach to the fourth ventricle and the several described IONM techniques employed, the patient developed postoperative cerebellar mutism, from which he eventually fully recovered after 40 days. Although still not validated as a formal technique to assess the neural pathways related to cerebellar mutism, recent work demonstrates IONM approaches, such as cerebello-cortical stimulation protocols, that have potential to address this issue in the future [14].

CONCLUSION

Surgery is the mainstream treatment for posterior fossa tumors in children, with regards to tumor control and the obtention of tumor samples. The use of intraoperative neuromonitoring during surgery in this current case promoted a change in the surgeon's strategy, in which resection was terminated to avoid damage to the facial nerve pathways. Despite its limitations, multimodal

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intraoperative neuromonitoring, in combination with the other surgical tools, including meticulous microdissection techniques and an ultrasonic aspirator, is an indispensable tool to help the surgeon achieve the maximum, and the safest, surgical resections as possible in this group of patients. The development of new intraoperative neuromonitoring techniques will certainly add tools that will enhance safety for the complex procedures involving the posterior fossa in children.

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DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Authors declare that this work is exempted from ethics committee authorization.

Consent to participate

The patients gave consent to use their information and images for research purposes. *Consent for publication*

The patient gave consent to use his information and images for publication.

Conflict of interest

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper."

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CONTRIBUTIONS

-**Charles Kondageski**: Conceptualization, Data curation, Formal Analysis, Methodology, Writing – original draft, Writing – review & editing
-**Humberto Kluge Schroeder**: Data curation, Methodology, Writing – review & editing
-**Antonio Cesar de Melo Mussi**: Writing – review & editing
-Cezar Massaru Guiotoku: Writing – review & editing
-Marcela de Moraes Barros Sousa: Writing – review & editing
-Jean Costa Nunes: Writing – review & editingWriting – review & editing

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