Clinical Case Reports

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Mercedes Benz pattern craniosynostosis associated with Chiari malformation type 1: A case report of a rare condition

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Introduction/Background:Craniosynostosis results from the premature fusion of cranial sutures and is classified as complex when it affects multiple sutures. The abnormal and rare fusion of the sagittal and bilateral lambdoid sutures is known as the Mercedes-Benz (MBC) pattern, responsible for up to 0.7% of craniosynostoses. It's characterized by flattening of the occipital bone and sub-lambdoid depression. Additionally, the Arnold-Chiari malformation, one of the anomalies associated, occurs due to the junction of deformities in the posterior fossa and rhombencephalon. Type 1 (CM1) is the most incident and less severe, represented by the pointed shape of the cerebellar tonsils.

Case presentation: Male patient, 11 months old, diagnosed with MBC associated with CM1, presenting with anterior turribrachycephaly and occipital brachycephaly. The diagnosis was made after the 10th month of life, with typical findings on computed tomography and cerebellar tonsil herniation (CTH) on magnetic resonance imaging (MRI). Subtle signs of neurodevelopmental delay were observed by the neurosurgeon, especially in language and motor skills. No history of cranial malformations were observed in the family. At 11 months, the patient underwent neurosurgery involving cranial parietal remodeling to correct craniosynostosis, along with posterior fossa decompression to address CM1.

Conclusion: Neurosurgical techniques like parietal remodeling and posterior decompression, despite their complexity, play a central role in addressing MBC and CM1. It's crucial to discuss this rare condition to ensure early diagnosis and treatment, facilitating an efficient approach and preventing potential later consequences.

Keywords: Craniosynostosis , Mercedes benz pattern ; Chiari malformation type 1; Neurodevelopment

INTRODUCTION

Premature closure of the bilambdoid and posterior parietal sutures, also known in the literature as the Mercedes Benz craniosynostosis pattern (MBC) - due to the similarities in the visualization of the fused sutures on Computed Tomography with the logo of the automaker Mercedes Benz - is a rare and complex pattern of craniosynostosis, with a prevalence of about 0.3-0.7% of all craniosynostosis [1,2].

Thus, in patients affected by this pattern, the development of anterior turribrachycephaly combined with mild brachycephaly is observed due to the flattening of the occipital bone and retreat of the bilamdoid sutures, leading to compression of the posterior fossa, compensated by the enlargement of the frontal bone[1].

Furthermore, the presence of multiple malformations in the central nervous system is a point of great attention in this type of clinical entity, highlighting among them, as being the most frequently anomaly associated with this condition, present in approximately 61% of MBC, the herniation of the cerebellar tonsils through the foramen magnum, typically known as Chari Malformation Type I(CM1) [3].

Therefore, the objective of this work is to describe a rare case of a 11-month-old child affected by MBC associated, concomitantly, with CM1.

CASE REPORT

A 11 months old male patient was born through vaginal birth in the maternity hospital, where developed generalized cyanosis and was immediately taken to the hospital's intensive care unit (ICU), being diagnosed with neonatal sepsis. During the period of hospitalization in the ICU, it was noticed that the newborn's cranial shape was unusual, presenting anterior turribrachycephaly and occipital brachycephaly

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The mother was able to get a consultation with the neuropediatrician when the child was 8 months old. A nuclear magnetic resonance imaging (MRI) was requested, which showed radiological changes suggestive of CM1(figure 1), such as herniation of the cerebellar tonsils through the foramen magnum, at approximately 1.3 centimeters below the MacRae line, also observing a reduction in the cerebrospinal fluid columns of the craniocervical junction. Furthermore, the MRI showed a considerable decrease in the dimensions of the posterior fossa, which is characterized by the absence of cisterna magna, low implantation of the confluence of the sinuses and partial obliteration of the cisterns at the base of the skull. With all these findings, the diagnosis of CM1 was reached.

The mother was advised to seek a pediatric neurosurgeon to manage the condition. The neurosurgeon observed a neuropsychomotor developmental delay, especially in language and motor skills, such as inability to crawl and stand, in addition to making few sounds and words. A skull computed tomography was requested, which showed the bilateral early closure of the lambdoid sutures and the posterior portion of the sagittal suture(figure 1), characterizing a typical pattern of a rare craniosynostosis known as MBC.

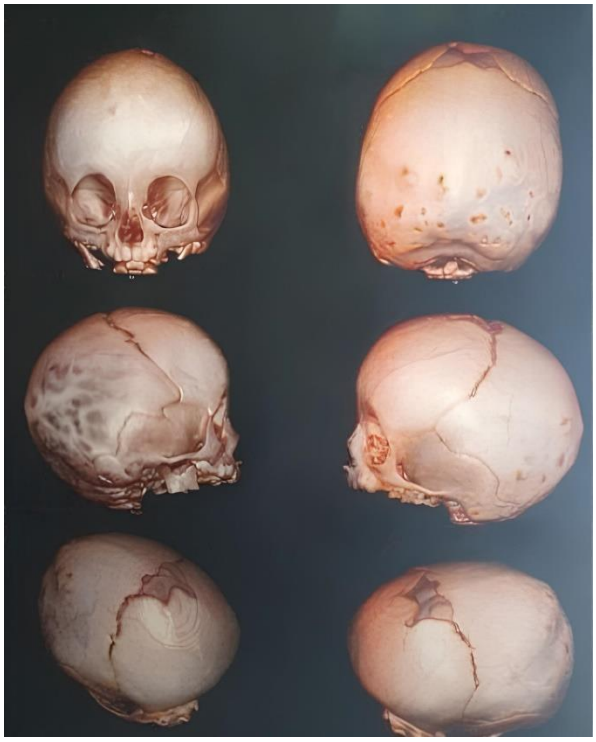

Figure 1 - 3D reconstruction of computed tomography showing anterior turribrachycephaly and occipital brachycephaly.

When asked, the mother denies ingesting alcohol or any type of teratogenic substance during the gestational period, as well as any history of cranial malformations in the family.

At 11 months, the child underwent the surgery, in which cranial parietal and occipital bone remodeling to correct the craniosynostosis, associated with decompression of the posterior fossa to correct CM1, by unblocking the foramen magnum.

For the surgical procedure, the child was placed in the prone position, undergoing a bicoronal zigzag incision from the coronal suture to the vicinity of the lambdoid suture (figure 2). The dura mater meninges were not opened, considering that as it was a bicoronal incision, there would be no possibility for its reconstruction. Using surgical drills with a high degree of rotation, bilateral sagittal craniotomies were performed, as well as coronal incisions were made in the parietal and occipital bones, in order to allow greater enlargement of the skull. Excisions and fragmentations were also performed on the median sagittal bone crest, with its parts being repositioned in the midline and fixed from the anterior to the posterior portion of the craniectomy, thus enabling a better aesthetic result.(figure 3)

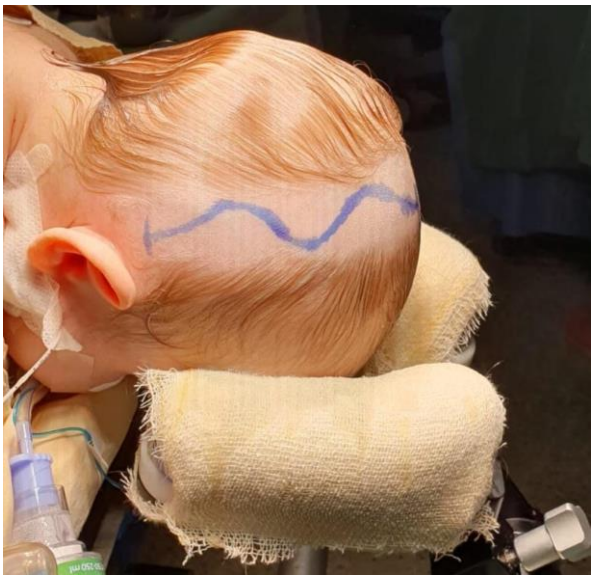

Figure 2 - Operative medical photographs of children showing turricephaly, brachycephaly, and sublambdoid depression in addition to the bicoronal demarcations for craniectomy.

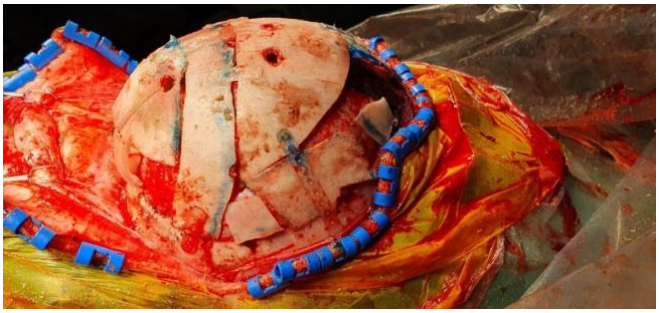

Figure 3 - Posterior craniectomy with remodeling of occipital and parietal bones. Foramen magnum decompression was also done.

After the surgical intervention, the patient remained under hospital care in the institution's ICU, aiming for more detailed post-operative monitoring.The day after the surgery, a computed tomography scan of the skull was performed, where it was already possible to notice an improvement in the shape and symmetry of the skull, without the presence of synostoses.There were no complications during the post-operative period and the child was discharged the same week after surgery (Figure 4).

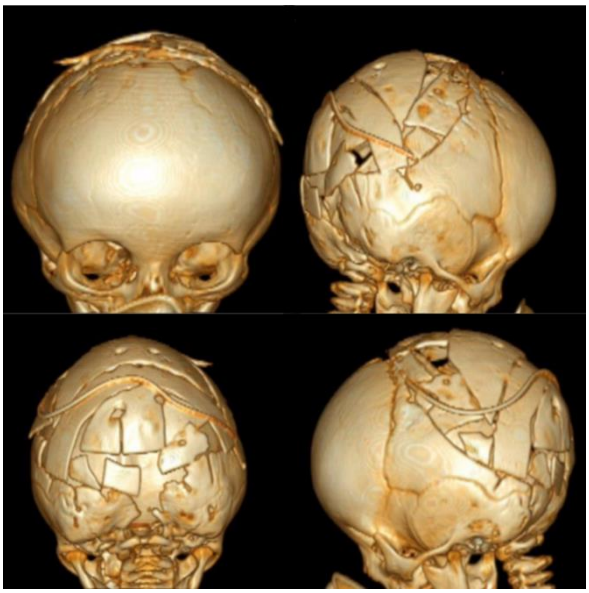

Figure 4 - 3D reconstruction of computed tomography the day after surgery.

The child attended the neurosurgery outpatient clinic for follow-up 3 weeks after surgery, where not only a significant improvement in the aesthetics of the child's cranial shape was observed(figure 5), but also a good development in the child's motor skills and language, with a much more active crawling and walking movement, in addition to a good improvement in the child's formation of sounds and first sentences, continuing work with speech therapy for language development. Also, it was advised to continue going to the outpatient clinic for follow-up every three months, for continuous evaluation.

Figure 5 - Follow-up 3 weeks after surgery.

DISCUSSION

The MBC, first reported by Neuhäuser et al.[4] in 1976 and later coined with the name "Mercedes-Benz Syndrome" by Moore et al. with 3 patients in 1998[5], is a craniofacial dyssynostosis characterized by the synostosis pattern of bilateral lambdoid and sagittal sutures, with minimal correlation with other syndromes[1,6]. Since then, few studies have been published on the subject, but it has been observed that its prevalence is higher in males, appearing in a ratio of 3:1 compared to females[1,6].

The condition manifests early with turricephaly and brachycephaly – just like the child studied in this case (figure 1) –, and dolichocephaly is also a manifestation that can be visible. Additionally, deformities in the posterior fossa are associated, such as flattening of the occipital bone and sub-lambdoid depression[1,3]. Furthermore, a phenotypic difference is observed due to the action of the driving force acting on the skull, generating short (occipital narrowing) and long (dolichocephaly) phenotypes with volumetric and craniometric variations. While the long phenotype does not show significant differences in volume distribution or craniometric measures, the short phenotype exhibits an increase in the volume of the anterior and middle fossae, along with a decrease in the volume of the posterior fossa, an increase in the occipital bossing angle, and a decrease in the occipital bone angle[7].

Chivoret et al. reported 39 cases of sagittal bilambdoid synostosis in 2018, in which 31 of these patients had it in its isolated form, typically known as the Mercedes Benz pattern. Of these, 61% of patients had cerebellar tonsillar hernia, a condition similar to this case report[3].

The Chiari malformation Type 1 (CM1), also referred to in the literature as a condition associated with MBC, is defined as the herniation of cerebellar tonsils by at least 5 mm in adults and 3 mm in children below the foramen magnum. This is the most common of the 4 types of Chiari malformations, present in approximately 0.6% and 0.9% of the general pediatric and adult population, respectively[10,11]. The association between the development of these two pathologies occurs for multiple reasons. It may be an acquired malformation resulting from the fusion of lambdoid sutures in the first months of life, where neural growth is exacerbated, leading to the underdevelopment of the posterior fossa, or/and it may result from increased intracranial pressure secondary to venous hypertension if the craniosynostosis impairs its drainage[12]. Furthermore, this condition may or may not occur in association with hydrocephalus, which was not the case of this patient.

The diagnosis of MBC usually occurs, on average, around the 23rd month of a child's life, with Computerized Tomography (figure 1) being the most commonly used method in literary documentation[9]. However, our patient was diagnosed earlier than the global average reported in the literature, which seemed to accelerate the effects of the therapy, as motor and verbal language improvements were made in a short period of time.

About treatment the literature shows that there are two main types of surgery that are used in a phenotype similar to our patient (short phenotype): remodeling of the parietal bone and posterior decompression. For CM1, the foramen magnum is cleared[9]; this three techniques were performed successfully in the present case (figure 3).

As for postoperative results, a significant improvement in the cephalic aesthetic result was observed in the patient, on the day after surgery, by means of computed tomography, with a much more symmetrical and defined cranial shape, without any presence of synostosis, which corroborates the previous results found in the literature for this surgical procedure[9]. In addition, another point that deserves to be highlighted is the improvement of the patient's cognitive function and neuropsychomotor development, which showed significant gains in language and motor skills, reinforcing the importance of not only the aesthetic and psychosocial aspects, but also the functional aspects of corrective craniosynostosis surgeries for cases of MBC[1].

CONCLUSION

Despite not being associated with high morbidity and mortality in the pediatric population, the association of these two clinical entities has been shown to be well associated with dysfunctions in childhood neurodevelopment.It is relevant to mention that surgical interventions including remodeling of the parietal bone and posterior decompression, are of essential contribution not only to aesthetic aspects but also in improving the quality of life for these individuals, playing a significant role in the progression of a favorable prognosis in these patients.

ACKNOWLEDGMENTS

DISCLOSURES

Ethical approval

Ethics Committee approval was not required for this work, as it solely involves a case description without any patient intervention. However, we have obtained written consent from the responsible party for the patient's case report to be published in the journal. Furthermore, we ensure that this study was performed in line with the principles of the Declaration of Helsinki.

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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Artificial Intelligence

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CONTRIBUTIONS

-**Leonam de Oliveira Silva**: Conceptualization, Investigation, Methodology, Project administration, Writing – original draft, Writing – review & editing
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