CASE REPORT

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Chiari Malformation (CM) type III surgical management in infant: a case report

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ABSTRACT

Background: Chiari Malformations (CM) is a collection of hindbrain abnormalities ranging from simple to complete agenesis of the cerebellum. Type III CM (CM III) is the rarest CM, with herniation of the cerebellum and brainstem into a posterior encephalocele and other intracranial anomalies. Reduction of the viable herniated brain, along with providing intracranial space and inadequate closure, often presents as the main problem. This case study aims to evaluate the Chiari malformation type III surgical management in infants.

Case Presentation: A 16-day-old male infant with an intact congenital mass in the occipital, with no neurological deficit. Radiological examination shows a defect in the occipital skull consisting of cystic non-contrast enhancing lesion and parenchymal tissue herniated through the defect. The cele excision and defect closure were performed, and viable neural tissue was found inside the sac. Blossom flower-like trim expanded the fossa posterior space and put the viable neural tissue inside the intracranial room. The defect was then closed with a dura, periosteal patch, and tension-free two-layered scalp closure. Post-operative follow-up presented a good outcome, and 1-year follow-up showed good recovery, with developmental delay, following developmental rehabilitation.

Conclusion: CM III is rare and has poor outcomes because neurological deficits and respiratory problems usually follow it. Operative management patient with CM III was presented with occipital encephalocele with no post-operative complication. Occipital bone reconstruction using a blossomed-flower-like technique could be considered to create adequate room for viable neural tissue.

Keywords: Chiari Malformation III, Occipital Meningoencephalocele, Surgery, Reconstruction.
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INTRODUCTION

Infants with Chiari Malformation (CM) type III have poor prognoses and usually present with respiratory failure, seizure, swallowing dysfunction, hypertonia, microcephaly, and/or amyotonia. Surgery is quite a challenge, often problematic from both technical and ethical points of view in CM.[1-3] Reduction of the viable herniated brain, along with providing intracranial space and inadequate closure, often present as the main problem.[2,3] CM type III is the rarest Chiari malformation, with herniation of the cerebellum and brainstem into a posterior encephalocele and other intracranial anomalies.[4] This is the most severe form of hindbrain herniation. Posterior encephalocele that occurred is regarded as a postneurulation disorder and is usually confined to the midline between the lambdoid suture and foramen magnum.[4] The neck of the sac is closely related to the venous sinuses, particularly the superior sagittal sinus, circular heterophilic, and occipital sinus, but rarely to the transverse sinuses. Size and consistency vary from large, potentially viable brain and passage vessels and large, primarily cerebrospinal fluid (CSF) to small with CSF only. The contents of the sac vary from the usual dysplastic neural tissue to the functional occipital lobe, brainstem, or cerebellum.[4,5]

We report a case of an infant with occipital defect, craniocervical junction incomplete cervical vertebrae I-3, and occipital cele with herniated cerebellar parenchyma. Those presentations are characteristic of Chiari Malformation Type III. The patient underwent a surgical procedure to remove the encephalocele along with the sac, dural closure, and modified occipital bone expansion to provide space for the viable neural tissue. This case demonstrates successful surgical management in Chiari Malformation type III.

CASE PRESENTATION

A case of a 16-day-old boy infant came to our Emergency Department with an intact mass on the head since birth without any size enlargement. There were no seizures, vomiting, or fever occurred. The patient was generally weak, with normal vital signs. Neurological examination showed spontaneous eyes opening with weak crying, and no movement deficits could be seen over the four extremities. Another neurological examination was within normal limits. The head circumference was 29 cm, with the great sinciput still open, flat, and without mass on palpation. The mass was located at the occipital with the

size of 15x13x3.5 cm, intact, tender, and light transillumination positive (Figure 1).

Head contrast Computed Tomography (CT) showed a defect of the calvaria in the occipital region along with a heterogenous mass herniated through the occipital consisting of a hypodense part suggestive of cerebral fluid and an isodense part suggestive of brain parenchymal (Figure 2). There was no sign of hydrocephalus. Bone window CT Scan showed lacunar consistency with defect at the occipital and suboccipital of 1.72 x 2 cm and widening of the first three cervical suggestive of spina bifida (Figure 3). Head magnetic resonance imaging (MRI) and Magnetic Resonance Venography (MRV) showed a hypointense cystic lesion through the occipital defect with a cerebrovascular fluid component in T1WI and hyperintense in T2WI; there was no contrast enhancement. There is herniation of occipital lobes through the defect, and there is no extrusion of the sagittal sinus through the defect (Figure 4). Echocardiography shows Ventricular Septal Defect (VSD) with a small perimembranous left to right Shunt without any clinical symptom.

The patient then was planned for Cele excision and defect closure. After general anesthesia, the infant was put in a prone position with the head flexed and supported by a horseshoe headrest. The chest was supported using a bolster. We kite some part of the encephalocele dome around the normal skin and elevate it with tissue forceps to facilitate skin preparation. The maneuver was carefully done to avoid interfering with the basal brainstem function. A vertical incision on the dome of the encephalocele was done, followed by blunt plane dissection to identify the dural sac, taking care not to break the sac until the neck of the sac and the bone defect are exposed (Figure 5).

The periosteum was preserved and the skin retracted while adequate hemostasis was done. The dome of the dural sac was incised and decompressed by CSF release. The content of the sac looked reddish, suggesting visually viable neural tissue, and was preserved. The skull defect was enlarged using the craniotome. We made a blossomed-flower-like trim to expand the posterior fossa (Figure 4) and reduce viable neural tissue to the center. The dura

Figure 1. Clinical presentation of Occipital Meningoencephalocele, intact, and transillumination test positive, indicates it has a large amount of Cerebrospinal Fluid (CSF).

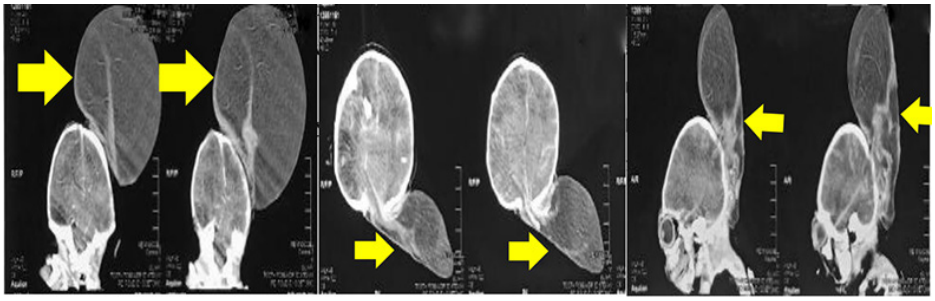

Figure 2. Head CT Scan with contrast Shows encephalocele dominated with CSF and a small amount of brain parenchyma.

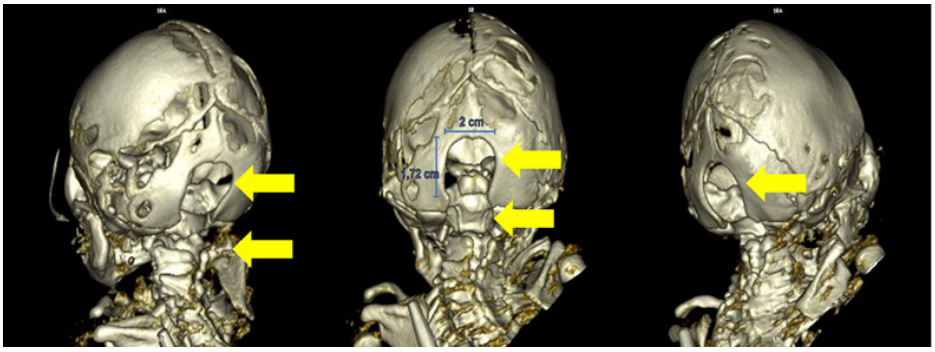

Figure 3. CT 3D Reconstruction shows the lacunar cranial bone and a defect in the lower part of the Occipital about 2x1,7cm, and incomplete development of Cervical I-III

was then closed primarily utilizing healthy dura mater. The periosteum was used for closure, covering the skull and the dura mater. The excessed skin was trimmed and tension-free and two-layered scalp closure was performed. The dressing was then applied to the wound (Figure 5).

The early postoperative examination showed stable condition in the neonatal intensive care unit (NICU) with ventilator support and spontaneous breathing mode. Neurological examination showed spontaneous eye opening and no neurological deficit on four extremities. Another neurological examination was within normal limits (Figure 5).

A 1-year follow-up was done; the patient was 1 year old, with excellent growth, with Global Developmental Delay. We collaborated with the Pediatrician and Rehabilitative Department to catch up on the developmental delay.

DISCUSSION

CM III is the rarest Chiari malformation type, a congenital defect identified by herniation of the cerebellum and brainstem into a posterior encephalocele. This operation aims to remove the sac, reduce the viable herniated brain, and have watertight dural closure with

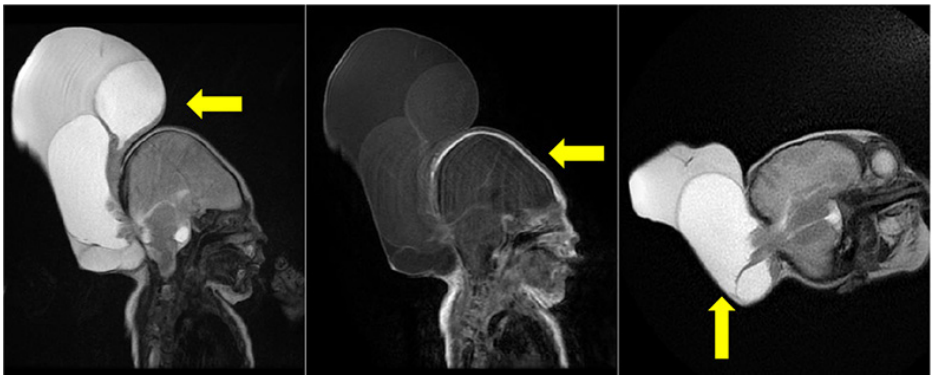

Figure 4. Head MRI and MRV showed a mass dominated with CSF, herniated cerebellum and no extrusion of the superior sagittalis sinus.

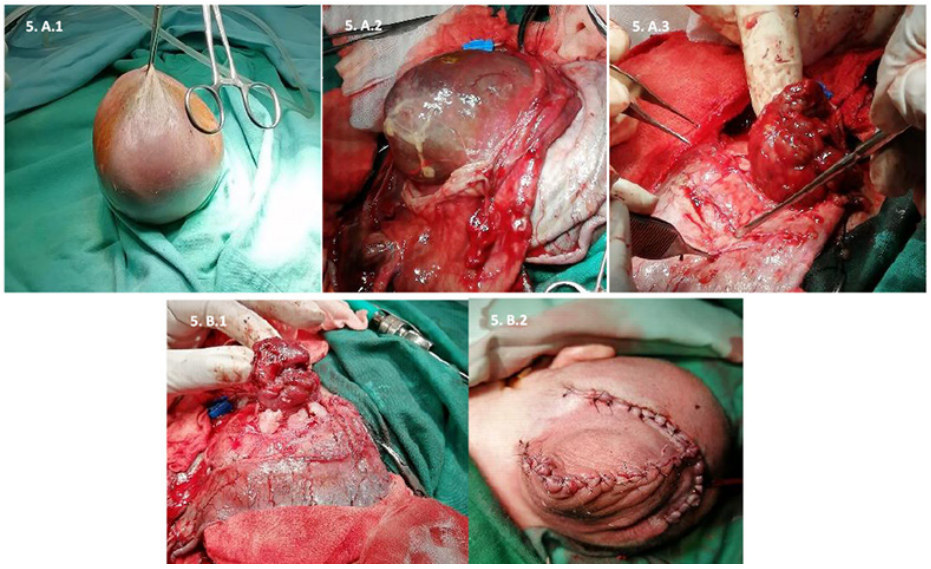

Figure 5. Operative step. (**5.A.1)** Prone position, some of the sacs changed; a Vertical incision was made, continued with blunt dissection to identify the dural sac until we visualized the neck of the sac and normal dura, followed by decompression of the sac (**5.A.2**). Neural tissue was visually evaluated, showing reddish appearance suggesting of viable neural tissue (**5.A.3**); Bone defect was trimmed perpendicularly, circularly in a *blossomed-flower* fashion to expand the occipital vault (**5.B.1);** Non-tension skin closure (**5.B.2)**

adequate healthy skin cover.[6] The main clinical presentation of this patient is microcephaly with a frontal-occipital circumference of 29 cm. An intact meningoencephalocele could be seen at the occipital region size of 15x13x3.5 cm. Vital signs were within normal limits, and there was no sign of neurological deficit. These are good provisions before the operation. MRI and MRV of the head revealed the meningoencephalocele was mostly comprised of CSF with some part of the cerebellum parenchyma. We could also see part of the brainstem herniated through the foramen magnum. There is no sign of sinus involvement inside the sac. Head CT Scan with 3-dimensional reconstructions showed a bone defect of the occipital and a widening of the posterior part of the first three cervical bones as a sign of spina bifida. This space provided room for the herniated brain stem and fossa posterior parenchyma, so the pressure around the structure did not increase, which could lead to neurological symptoms.

Surgery for this patient is a challenge. Surgery was done under the services of a well-experienced neuropediatric surgeon and neuro anesthesiologist.[6,7] The surgery's primary goal includes removing the sac, reducing the viable herniated brain, and watertight dural closure with adequate healthy skin covering. Careful preoperative planning was made to minimize upcoming complications.[6-9]

We visually observed the neural tissue after the dural sac was identified as decompressed. Three scenarios may be encountered at this point. The lesion might be a simple meningocele, allowing the redundant dura to be amputated, leaving enough cuff at the base for a watertight dural closure. The second scenario is that the dura sac contains a gliotic or malformed brain, as often encountered. This extracranial mass of tissue should be amputated at its base after it is bluntly dissected off the dura, maintaining hemostasis with bipolar. The third scenario is that the sac content may be viable neural tissue, necessitating reduction into the cranial space, which might be facilitated by enlarging the skull defect using rongeurs. Minimizing blood entry into the ventricles is important. In this patient, we found viable neural tissue.

We modified occipital bone expansion to provide room for the viable neural tissue by multiple trimming perpendicular to the bone defect circularly with craniotome, with periosteum preservation. The opening was done blossomed-flower with the neural tissue in the center. As the extra space was created, we narrowed the trimmed bone as close as possible. The periosteal was used for closure as covering above the bone. This modification provided extra space for the viable neural tissue and avoided subsequent increases in intracranial pressure. Skin closure was done non-tensioned to prevent unnecessary skin traction, possibly leading to surgical site dehiscence. This was an important mark to be made considering the skin fragility in this patient due to being in an infant age group and the pathology itself.

Post-operative care is another challenge that must be managed. We closely monitor the patient at the NICU. The patient was put in an incubator with ventilator support in spontaneous breathing mode. Using a modified donut pillow, the patient was also positioned carefully to avoid any pressure on the operated side. Careful observation was done for CSF leakage

and healthy perfusion of the skin edges. Encephaloceles involving the brainstem should also be monitored for brainstem dysfunction, including apnea, feeding problems, and aspiration. Postoperative hydrocephalus may occur in 30 to 60% of cases, requiring a CSF shunt. Patients with significant brain abnormalities may have severe developmental delays. Patients with atretic encephaloceles may be completely normal, with the lesion being an incidental finding.[6-8] For developmental delay, swallowing dysfunction, and cortical vision deficiency, a rehabilitation procedure was initiated as in the report of the previous study.[5] Trunk equilibrium monitoring and pelvic postural modification were carried out for trunk ataxia related to the cerebellar lesion using the principle of neurodevelopmental therapy, and functional electrical stimulation of the lower cervicothoracic paravertebral muscles was repeatedly implemented.[5] The relaxation process of visual perception has been used to treat cortical visual disability. Previous studies also presented the definition based on Hoyt's classification for the first time.[10,11] In each eye, a spotlight was mirrored in a dark and silent space, and preparation for light detection and contrast perception using a white/blackboard and pen-light proceeded.[12-14] Important enhancement was reported in children handled with this technique.

The limitation of this case report is its reliance on a single case, which hinders the generalizability of the findings to a broader population. To overcome this limitation, future studies should consider conducting larger-scale research involving diverse cohorts to enhance the validity of conclusions and provide more robust evidence regarding the effectiveness and safety of specific interventions or treatments. In addition, these case reports describe a single case, making it challenging to generalize the findings to a larger population or make broad conclusions about the effectiveness or safety of a particular intervention or treatment.

CONCLUSION

CM III is rare and has poor outcomes, usually followed by neurological deficits and respiratory problems. Careful pre-operative preparation and meticulous post-operative care are needed to ensure better outcomes and avoid potential complications. Occipital bone reconstruction with a blossomed-flower-like technique is a viable option to create less invasive and adequate room for viable neural tissue. Long-term follow-up is mandatory to treat possible side effects such as hydrocephalus and to manage developmental delay.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest regarding this manuscript.

ETHICS CONSIDERATION

It is not applicable as no patient identity was disclosed.

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AUTHOR CONTRIBUTIONS

Yusuf Baktir contributed to the **study's** conceptualization, data collection, writing, and editing. Muhammad Arifin Parenrengi and Wihasto Suryaningtyas contributed to reviewing, editing, and finalizing the manuscript of the study.

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