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Autograft cranioplasty for correction of severe frontal skull defect due to hypocalvaria: A case report and review in management

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ABSTRACT

Objective: Here, the authors describe their institutional experience managing patient with severe frontal skull defect due to hypocalvaria with brain herniation.

Material and methods: The study was conducted in Neurosurgery Department, "Dr. Soetomo" Academic General Hospital, Surabaya, Indonesia. Neonates has a congenital anomaly, with skull defect in frontal skull duo to hypocalvaria. Defect closure with autograft cranioplasty with skull bone was performed in surgery.

Outcome: The surgical was performed with autograft cranioplasty using the temporal bone on the left region. We perform decompression of the left temporal area and insertion of a ventriculo peritoneal shunt at the right keen point to reduce intracranial pressure. Surgical treatment of patient with severe skull defect should be done.

Conclusion: Autograft with skull cranioplasty are biocompatible, which are easy harvested and with less infection and reaction risks.

Conflict of interest:
I undersign, certificate that I do not have any financial or personal relationships that might bias the content of this work.

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INTRODUCTION

Hypocalvaria is a rare congenital anomaly in neonates. It is characterized by the partial absence of flat bones of the skull. Hypocalvaria is a post neurulation defect, although some argue that it occurs because of defective primary neurulation resulting in non-closure of anterior neuropore [1]. The bones of the membranous neurocranium are present, but are hypoplastic, This post neurulation defect has been attributed to an abnormal migration of mesenchymal tissue at the beginning of the fourth week of embryonal development [1,2]. This congenital anomaly is usually associated with variable spectrum of cerebral malformations [3]. The Congenital defects of scalp bones are rarely reported and surgical treatment for correcting the skull defect is rarely discussed in literature till date. Highlights the importance of initial management and supportive care in patients with calvaria, in this case report we perform skull reconstruction by bone grafting and cranioplasty.

CASE PRESENTATION

A male new-born baby was referred to our hospital, because of multiple congenital anomaly such bilateral anopthalmia, labiognatopalatoschizis, microtia and unusual lesion on frontal region with skull defect. He was born at 41 weeks of gestation (unexplained preterm onset of labor) with a birth weight of 2600 g and an uncomplicated antenatal course. The baby crying at birth and didn't need resuscitation. There was no history of consumption of drugs during the antenatal period. He was born in first gravid mother. In this baby, a skull defect was found in the frontal region with a size of 9 cm x 8.3 cm x 4cm, his skull was palpable over the frontal region, translumination is negative and no discharge.

Surgical approach was performed while the patient was under the influence of general anesthesia. The surgical procedure was performed when the patient is 7 days old. The utilization of an MRI procedure serves to meticulously identify any irregularities and intricacies within the brain's structures, without exerting any influence or modification upon the subsequent procedures and interventions undertaken for the benefit of this

patient. Prior to cerebrospinal fluid diversion and reducing intracranial pressure, the patient was positioned supine with a planned coronal incision, and a ventriculoperitoneal shunt was performed before autograft cranioplasty. Ventriculoperitoneal shunt was surgically implanted at the precise anatomical location known as Keen's point on the right side, following a trajectory that precisely aligns with the right lateral ventricle. Incision was made, and the periosteum was incised with a rasparatorium to locate the frontal calvarial defect's border. Then, 9 x 7 centimeters of autograft bone were harvested from the left calvarial bone. The reparative procedure for calvarial herniation entails the meticulous execution of left temporal bone decompression. Following the meticulous decompression of the left temporal and parietal bones, accompanied by the precise insertion of the ventricular shunt, the herniated brain in the frontal region was skillfully and effectively reduced, culminating in the successful execution of the cranioplasty reconstruction procedure. We reconstruct the bone graft to close the frontal defect and then secure it with silk 2.0 thread. After autograft cranioplasty, the defect in the frontal region of the skull can be adequately reconstructed, and then the skin is sutured layer by layer.

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ventricular shunt, the herniated brain in the frontal region was skillfully and effectively reduced, culminating in the successful execution of the cranioplasty reconstruction procedure. We reconstruct the bone graft to close the frontal defect and then secure it with silk 2.0 thread. After autograft cranioplasty, the defect in the frontal region of the skull can be adequately reconstructed, and then the skin is sutured layer by layer.

DISCUSSION

Cranial vault defects and congenital defects of scalp bones are rarely reported. Hypocalvaria is an extremely rare condition with incidence is rarely and has been sparingly reported in literature with only a handful of case reports. Its causation is poorly understood and no concrete etiology has been described. In some literature hypocalvaria associated with Intrauterine Exposure to an *Angiotensin Receptor Blocker* (ARB) (4,5). The cause of hypocalvaria is still largely unknown, but it is believed to be a result of a developmental defect during embryonic growth. This suggests that it is caused by a disrupted movement of the mesenchyme (which forms muscle and bone) alongside a normally positioned embryonic ectoderm (which develops into skin and scalp) in the 4th week of gestation when the cranial end of the neural tube closes. Consequently, the brain is left covered only with an intact layer of skin with absence of calvarium (6). Hypocalvaria is typically not linked to any chromosomal abnormalities, and most cases occur randomly. In cases related to amniotic band, early amnion rupture is the triggering event. The amnion can sometimes coil tightly, causing the fetal head to become trapped. This leads to acalvaria due to the incorrect migration of the membranous neurocranium. For other cases of exposure to *angiotensin converting enzyme* (ACE) inhibitors or ARBs during the second and third trimesters of pregnancy, complications such as oligohydramnios, renal failure, neonatal hypotension, bony deformity and skull ossification defects have been reported (4,5,7). Hypocalvaria is different with Fronto-ethmoidal (anterior) encephalocele. Anterior encephalocel is a congenital malformation where intracranial contents protrude through a defect in the skull at the junction of the frontal and ethmoidal bones. One commonly accepted theory suggests that the development of fronto-ethmoidal encephalocele may be linked to a disruption in the closure of the rostral neuropore towards the end of the third week of intrauterine life. This disruption causes long-lasting

adhesions between the neuroectoderm and surface ectoderm where the nasal fields eventually close. (8).

Congenital defects of the scalp and skull present a challenge for care providers because of a combination of their rarity and the magnitude of potential morbidity. Recent advancements in autogenous and alloplastic cranioplasty and scalp reconstruction techniques argue for a comprehensive consideration of this problem (9).

Cranioplasty involves surgically repairing a defect in the calvaria. Several materials have been utilized for the purpose of repairing cranial defects. An ideal material for cranioplasty should possess qualities such as being radiolucent, resistant to infections, non-conductive to heat or cold, and resistant to biomechanical processes. Autograft bone grafts are highly favored due to their ability to minimize the introduction of foreign materials into the body. Additionally, the bone flap can be easily accepted by the host and seamlessly integrated back into the skull. It is more favorable in pediatric patients, as the child's original skull material will naturally reintegrate as they grow older (10,11). In addition the patient shall undergo periodic clinical evaluations on a monthly basis, followed by a cerebral computed tomography (CT) MRI Examination at the 6-month mark, and subsequently at the 1-year milestone post the primary intervention to show an adequate bone ossification.

Another method for skull reconstruction involves using autogenous calvaria and employing an expansion technique. This procedure involves making multiple interdigitating osteotomies to expand the cranium. In infants and neonates, the calvaria is carefully opened to fully utilize the available donor bone, as it can be challenging to separate. With this technique, smaller cranial defects are generated, increasing the likelihood of spontaneous re-ossification (12,13).

CONCLUSION

We had presented a management patient with congenital skull bone defect. When surgical intervention as warranted, autograft bone is the ideal reconstruction material.

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