Journal of Oral Biology and Craniofacial Research 11 (2021) 13–16

Contents lists available at ScienceDirect

Journal of Oral Biology and Craniofacial Research

journal homepage: www.elsevier.com/locate/jobcr

Massive posterior cranial vault erosion and its reconstruction: A peculiar presentation of "mega cisterna magna"

Adity Bansal [a,*], Gosla Srinivas Reddy [a], Ashi Chug [a], Sriram Chandra Damaraju [b]

[a] Department of Dentistry (Cranio-maxillofacial Surgery), AIIMS, Rishikesh, Uttarakhand, 249203, India
[b] GSR Institute of Craniofacial Surgery, Vinaynagar Colony, Saidabad, Hyderabad, Telangana, 500059, India

ARTICLE INFO

Keywords:
Dandy-walker complex
Cranioplasty
Posterior cranial fossa malformation
Ventriculo-peritoneal shunting

ABSTRACT

Dandy-Walker (DW) complex is a rare central nervous system malformation, commonly associated with complex non-neurological conditions, defined by four variants with characteristic anatomic features, still inadequately known for its etiological aspects. "Mega Cisterna Magna" (MCM) is a type of DW complex which is represented by an enlarged posterior cranial fossa. Though reduction cranioplasty has been reported in enlarged posterior cranial fossa malformations, however we report a peculiar case of MCM with massive posterior cranial bone erosion, presenting completely asymptomatic at an age of 8 years, without any associated co-morbidities till date, nor with any evident occipital mass at birth. Survival of the child till this age is an exception in itself, but this can probably be explained by the presence of the giant defect of the posterior cranial bone, which must have acted like a vent and prevented the elevation of the ICP. Hence no symptoms were observed till date. Ventriculo-peritoneal (VP) shunting was done to drain the accumulated fluid, followed by massive cranioplasty, which was challenging and was performed with autologous cranial and fibular bone grafts, along with alloplastic titanium mesh, and thus achieving marked aesthetic improvement with satisfactory bone healing at a 3-year follow-up.

1. Introduction

Dandy-Walker (DW) complex is a rare multi-entity neurological malformation, defined by four variants with characteristic anatomic features, still inadequately known for its etiological aspects.[1] The extended period of embryonic development of the cerebellum makes it vulnerable to wide spectrum of disruptions and malformations.[2] "Mega Cisterna Magna" (MCM) is a type of DW complex which is represented by an enlarged posterior cranial fossa.[3] Though reduction cranioplasty has been reported in enlarged posterior cranial fossa malformations,[4,5] however PubMed literature search for (("posterior cranial vault resorption") AND ("dandy walker complex" OR "mega cisterna magna" OR "dandy walker syndrome" OR "posterior fossa malformation")) yielded zero results. Hence we report a peculiar case of MCM with massive posterior cranial bone erosion, presenting completely asymptomatic at an age of 8 years, without any associated co-morbidities till date, nor with any evident occipital mass at birth. Ventriculo-peritoneal (VP) shunting was done to drain the accumulated fluid, followed by massive cranioplasty, which was challenging and was performed with autologous cranial and fibular bone grafts, along with alloplastic titanium mesh, and thus achieving marked aesthetic improvement with satisfactory bone healing at a 3-year follow-up.

1.1. Case report

An 8 year old female patient presented with an enlarged posterior cranium (Fig. 1a). She was born at full term, by normal delivery with no apparent occipital mass. It was first noticed at 1 year of age, but the parents did not get any specific assessment done at that time. No symptoms were reported till date, except for progressive posterior cranial enlargement in the following years. No similar condition was reported in siblings or first degree relatives.

Clinical evaluation revealed a massive posterior cranium with bulging occiput. Occipito-frontal circumference (OFC) was measured to be 70 cm (normal for the age: 55 cm). The skin over the cranium was normal and non tender. The bony breach could be palpated with respect to the occipital region. General examination observed no abnormality.

There were no signs of increased intra-cranial pressure (ICP), with no motor deficits or spastic paresis, and normal developmental milestones. No intellectual disability or hypotonia was discerned. Ophthalmic abnormalities like strabismus or nystagmus were ruled out. Cranial nerve functions were within normal limits, with no speech difficulty. Other

* Corresponding author.
E-mail addresses: aditybansal@rediffmail.com (A. Bansal), ashichug@gmail.com (A. Chug), Damaraju.sriram@gmail.com (S.C. Damaraju).

https://doi.org/10.1016/j.jobcr.2020.11.010
Received 21 September 2020; Received in revised form 6 November 2020; Accepted 8 November 2020
Available online 12 November 2020

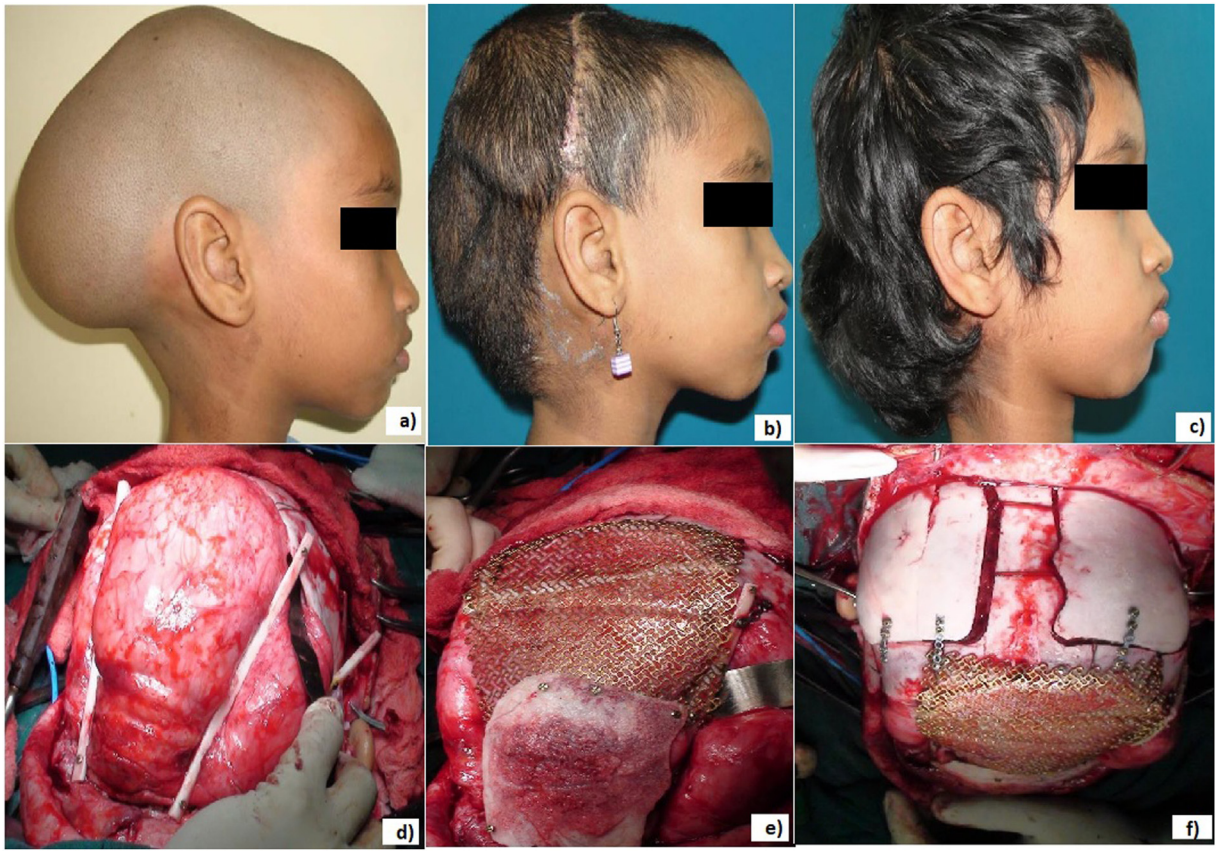

Fig. 1. a) Side profile of the patient revealing massively enlarged posterior cranium; b) and c) satisfactory and marked improved can be observed aesthetically at one week and 3 year follow up respectively; d) Fibular bone grafts used as bony struts on bilateral lateral sides of the bony defect; e) Titanium mesh and inner parietal cortical plate used for reconstruction of the bony defect; f) outer parietal cortical plate placed back at the parietal harvest site.

focal neurological signs like seizures were absent. No psychological, dermatological, cardio-vascular, gastro-intestinal, endocrinological, musculoskeletal, or uro-genital abnormality was detected. Chromosomal abnormalities were ruled out.

Computed tomography (CT) revealed massive erosion of posterior compartment of brain, measuring 15 cm x 12 cm x 5 cm (Fig. 2a–c). Magnetic resonance imaging (MRI) showed patent aqueduct, with normal cerebellar vermis and fourth ventricle, with evident hydrocephalus, which led to the diagnosis of MCM.

Surgical management was planned in a single stage. VP shunt was placed initially to drain the accumulated fluid, which led to the over-drainage of the fluid due to the absence of bony skull to prevent the skin from flopping inside, thus necessitating the need for the bony reconstruction. The posterior cranial bony defect was then exposed through standard bi-coronal approach under general anesthesia. Bilateral avascular fibular bone grafts were harvested, along with full thickness bilateral parietal bone grafts. Fibular bone grafts were used as bony struts on bilateral lateral sides of the bony defect, extending vertically from parietal to occipital bone (Fig. 1d). Parietal bone grafts were then split into inner and outer cortical plate grafts. The inner parietal cortical grafts (as bony strips) and titanium mesh was used to reconstruct the occipital defect (Fig. 1e), while the outer cortical parietal plate was placed back to contour the harvested parietal region (Fig. 1f). Stabilization of the grafts was followed by their fixation with 2 mm titanium mini-plates and 2 mm titanium screws. Closure was then done in layers.

The child had an uneventful recovery in the post operative period (Fig. 1b). She was kept in intensive care unit. Patient was fully alert the following day with no neurological complications. Age-appropriate mental and motor development was observed on neuro-psychological assessment. Child is stable and asymptomatic at a 3 year follow up and achieved marked improvement aesthetically (Fig. 1c). Follow up CT at the age of 11 years revealed satisfactory bony healing (Figure d–f).

The work has been carried out in accordance with the Code of the Ethics of the World Medical Association. Written and informed consent was obtained from the patient and the parents for the treatment and for the publication of the case and the images.

2. Discussion

Dandy-Walker (DW) complex is a rare central nervous system malformation, commonly associated with complex non-neurological conditions.[3] Such disfigurements are formed at 7–10 weeks of gestation during embryogenesis.[6] It comprises of four different types, with variable functional, clinical and radiological manifestations, thus requiring adequate diagnosis and management. “DW malformation” is defined by agenesis or hypoplasia of vermis, cystic enlargement of the fourth ventricle and a large posterior cranial fossa. “DW variant” has normal posterior fossa with vermis hypoplasia and fourth ventricle dilatation. “Mega cisterna magna” (MCM) has only enlarged posterior cranial fossa; while last type of DW complex is defined as “posterior fossa arachnoid cyst”.[3]

Its etiology is considered heterogenous, with Stambolliu et al. reporting association with chromosomal abnormality or a syndrome in 33% of the DW cases,[1] however meta-analysis by D’Antonio et al. detected no chromosomal abnormality in fetus tested prenatally for mega cysterna magna.[7] Other co-morbidities associated are cardio-vascular like patent ductus arteriosus (PDA), ophthalmic like cataract, otologic like hearing impairment, malignancies like nephroblastoma, mental illness, mild or severe intellectual disability.[1]

Usually macrocephaly is a consequence of hydrocephaly in DW

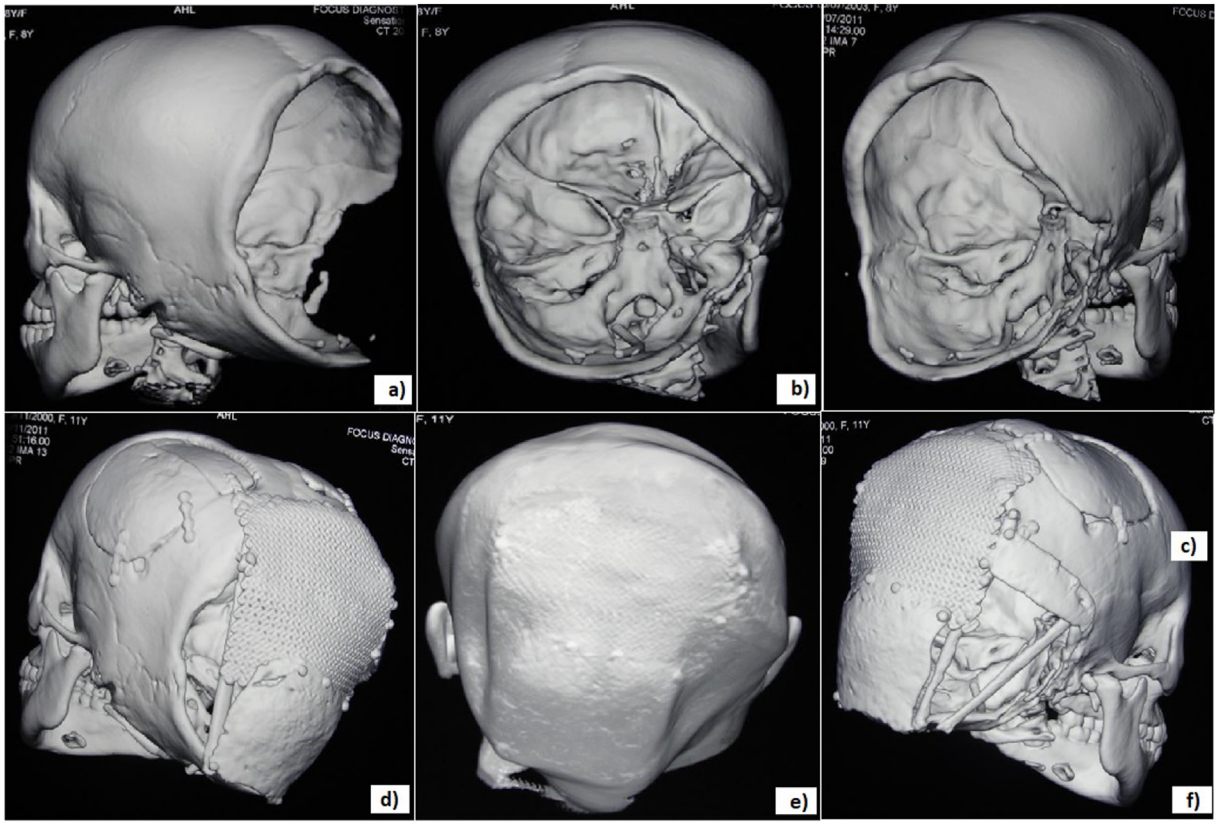

Fig. 2. a-c) 3-Dimensional computed tomography sections depicting massive erosion of the posterior cranium; d-f) -Dimensional computed tomography sections at a three year follow up showing satisfactory bone healing.

complex, but can also result due to posterior fossa enlargement due to cyst formation.[4] Occpital meningocele (OCM) has also been seen associated with such malformations.[8] Survival of the child till this age is an exception in itself, but this can probably be explained by the presence of the giant defect of the posterior cranial bone, which must have acted like a vent and prevented the elevation of the ICP. Hence no symptoms were observed till date. In this case, VP shunt was placed initially to facilitate fluid drainage, hence reducing the bulged-out occiput. Since the resorption of cranial vault was massive, hence cranioplasty was considered mandatory to provide cranium it's near to ideal shape. Such reconstruction technique requires skill and understanding of the identifiable surface landmarks along with venous anatomy to minimize vascular injuries.[9] After adequate planning to cover as wide-a-defect as possible, bilateral fibular grafts were harvested to act as lateral bony struts, which then acted as a scaffold for further reconstruction. Outer and inner table of bilateral parietal bone grafts were then harvested, and were used individually so that maximum benefit can be attained for the coverage of the defect. Titanium mesh was used to complete the cranial vault concealment. Knowledge of imaging patterns at various stages of embryogenesis, and precise parental counseling is strongly suggested to detect such spectrum of malformations at the earliest. Development of 3-D sonography has also gained importance in last few decades.[10]

3. Conclusion

The surgery in such giant DW defects is challenging and aims at improving the quality of life and aesthetics of the child. Though cranioplasty has been reported in various DW complex cases, but reconstruction in such a massive erosion of posterior cranium in MCM is rarely presented, especially with the asymptomatic patient presentation.

Funding source

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Declaration of competing interest

There is no conflict of interest to declare from any of the authors.

Acknowledgements

None.

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