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Case Report

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A Rare Case Report of Acrania

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Abstract
Acrania is a rare congenital anomaly and characterized by partial or complete absence of the calvarium. Although acrania associated with anencephaly is a well-recognized entity but isolated acrania is a rare anomaly. Ultrasound allows early diagnosis of this anomaly. The fetus was found to have a completely formed brain, base of the skull and facial structures but lacking a cranium. Authors present a rare case of acrania. [Journal of National Institute of Neurosciences Bangladesh, July 2023;9(2):157-160]

Keywords: Acrania; brain; cranium

Introduction

Acrania is a rare congenital anomaly characterized by partial or complete absence of the cranium and by existence of abnormal development of the brain tissue[1]. The term of acrania and anencephaly are often confused since every case of anencephaly has calvarian bone defects[2]. Approximately 40 cases have been reported in the English literature since the first description by Mannes et al[1]. The pathogenesis of acrania is unknown, but it is suggested that acrania is a congenital anomaly resulting from failure of the mesenchyma to migrate under the ectoderm overlying the brain tissue over the cerebral hemispheres[3].

The acrania - exencephaly – anencephaly sequence together with spina bifida are the two most common neural tube defects worldwide with a prevalence of 1.86 per 1,000 live births[4]. The acrania is not actually an isolated neural tube alteration, it belongs to a sequence called acrania exencephaly anencephaly, since the lack of bones that make up the cranial vault will cause a protrusion of the cerebral parenchyma (exencephaly) and with the sudden movements of the fetus along with the chemical irritation of the amniotic fluid to the unprotected brain structure causes degeneration and destruction of it causing in the absence of brain mass (anencephaly)[3,5]. Due to this pattern of progression, anencephaly is considered relatively more common than exencephaly[6].

It is described as a post-neurulation defect that, after the closure of the cranial neuropore, there is an alteration in the migration of the membranous portion of the neurocranium[2]. Among the risk factors for this defect, low intake of foods rich in folates[7] is important. Access to food enriched with folates, which are public norm in some countries, are the protective factor to avoid malformations[8], in the same way the genetic origin has been demonstrated as a risk factor for acrania, as is the case of the polymorphism of the methylenetetrahydrofolate reductase found in 2.0% of Afro-descendants and more than 35% in Chinese and Mexican[9-10]. We present a case of acrania in Dhaka, Bangladesh.

Case Presentation

A 17 years old non-diabetic, non-hypertensive, non-alcoholic primigravida mother with normal obstetric examination gave birth a female baby with acrania in full term. There is no family history of congenital anomalies. There was no history of intake of teratogenic drugs and other relevant past illness. During ultrasonographic scan a single live fetus with cephalic presentation was found. Ultrasonologist did not mention any congenital anomaly. Live fetus born through normal vaginal delivery without any

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complication, having well-formed brain without skull bone covering (acrania). Brain tissue cover with thin membrane. Facial feature was normal. Nasal bone, lips were seen normally formed. Baby cried at birth and did not need initial resuscitation. Baby had well-coordinated body movements. Baby's respiration was regular and there were no heart murmurs suggestive of cardiovascular involvement. New born baby was brought to our hospital after 2 days of delivery on 25.11.2020 from Norsingdi district of bangladesh. We found new born baby without cranium, baby was otherwise normal. Dressing with Sofratulle and gauze was done. Death of neonate was occurring at age of 23 days at home.

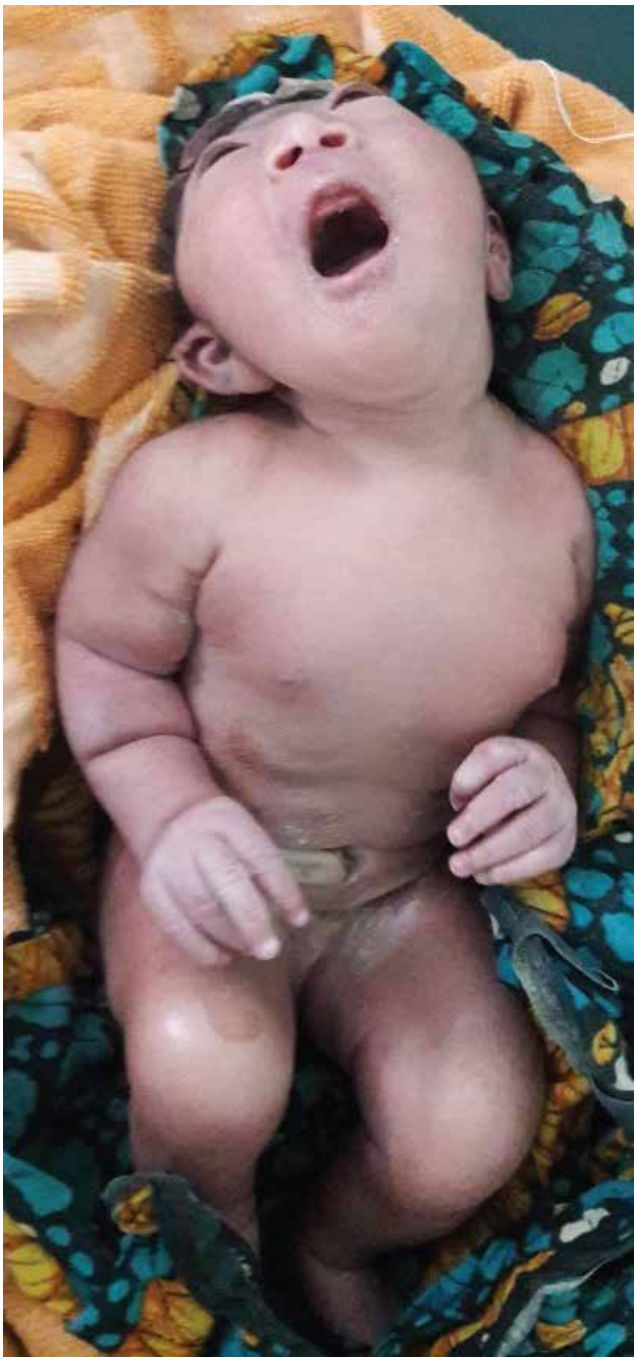

Figure I: baby with acrania

Figure II a,b,c,d : Baby with acrania

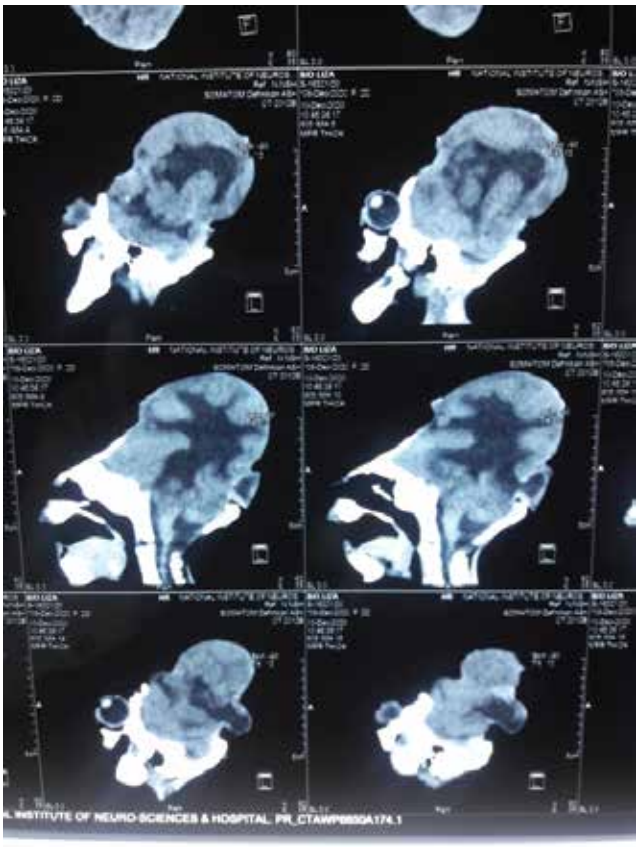

Figure III: CT scan of baby with acrania

Discussion

Acrania is a congenital abnormality characterized by the complete or partial absence of skull bones surrounding the fetal brain with complete, but abnormal development of brain tissue[11]. Although acrania associated with anencephaly is a well-recognized entity with an incidence of about 10:10,000 births, isolated acrania is a rare anomaly. The fetal cranium is not fully calcified before 10 to 11 weeks; fetal acrania can be diagnosed from 11 weeks onward[12]. At 11 to 14 weeks' gestation, the majority of cranial ossification is in the lateral aspects of the frontal bones and lower parietal bones, and no vault ossification is visible in the midline on a perfect midsagittal image. Hence, misdiagnosis may occur if only midsagittal views of the fetus are obtained. It is important to look specifically for frontal bone ossification in the axial and coronal planes[13-14]. For prenatal diagnosis, ultrasound remains the primary modality[15].

Neural tube defects have a prevalence of 1.86 per 1000 live births[5], among the two most common pathologies are spina bifida and acrania. The latter is not actually an isolated neural tube alteration, it belongs to a sequence called acrania exencephaly anencephaly, since the lack of bones that make up the cranial vault will cause a protrusion of the cerebral parenchyma (exencephaly) and with sudden movements of the fetus and the chemical irritation of the amniotic fluid to the unprotected brain structure causes degeneration and destruction of the brain and causing its absence (anencephaly)[3]. There is a confusion between what is acrania and acalvaria, the first is the absence of the scalp and partial or complete cranial vault that inevitably causes anencephaly, while the second is the total or partial absence of the skull bones but with an intact scalp and it will not cause anencephaly[16]. Therefore, acrania that is incompatible with life, acalvaria could have a survival expectancy.

It is described as a post-neurulation defect that, after the closure of the cranial neuropore, there is an alteration in the migration of the membranous portion of the neurocranium[2]. The fetal neurocranium has two sections, the chondrocranium that forms the base bones and the membranous flat bones that form the cranial vault, acrania is an alteration in said formation and occurs at 4 weeks of gestation when there is a defect in the closure cranial neuropore[17]. The diagnosis is made in the 12-week ultrasound where the bones of the cranial vault are not visualized, which are characterized by being a hyperechoic ring surrounding the brain parenchyma[17].

Since the 1960s, the relationship of folate deficiency with neural tube defects was studied, it was determined that folate was essential for the transfer of a carbon unit for the transformation of homocysteine to methionine, DNA methylation and others. Cellular reactions are an essential part for rapid tissue growth and cell replication[18-20], it is for this reason that many countries are obliged to supply folic acid to all pregnant women. Acrania has a high mortality rate of almost 100.0% cases, due to incompatibility with life, it is decided to terminate the pregnancy.

Conclusion

Acrania is a rare congenital anomaly. Most cases are incompatible with life. Antenatal identification allows the clinician to make appropriate and timely management decisions.

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Conflict of interest
Other than technical and logistic support from the scientific partner the investigators did not have any conflict of interest in any means

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Contribution to authors
Arman DM, Mukherjee SK, Islam J conceived and designed the study, analyzed the data, interpreted the results, and wrote up the draft manuscript. Arman DM, Mukherjee SK involved in the manuscript review and editing. All authors read and approved the final manuscript.

Data Availability
Any inquiries regarding supporting data availability of this study should be directed to the corresponding author and are available from the corresponding author on reasonable request.

Ethics Approval and Consent to Participate
Ethical approval for the study was obtained from the Institutional Review Board. As this was a prospective study the written informed consent was obtained from all study participants. All methods were performed in accordance with the relevant guidelines and regulations.

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