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Case Report

Rare case of congenital anomaly in two different gestational ages: a case series

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ABSTRACT

Acrania is a rare congenital anomaly involving faulty development of membranous flat bones of neurocalvarium. Commonest differential diagnosis is anencephaly. The first case was diagnosed in a second gravida at 13 weeks and terminated by vaginal misoprostol instillation. The second case was diagnosed only by scan at 32 weeks in an unbooked second gravida with previous LSCS. She underwent emergency LSCS for failed induction and a fresh stillborn baby of 1.9 kg was born.

Keywords: Acrania, Anencephaly

INTRODUCTION

Acrania is a rare, lethal congenital anomaly affecting the membranous flat bones of the neurocalvarium with complete but abnormal development of brain tissue.^{1,2} Also known as exencephaly, this condition is frequently associated with anencephaly though not always and isolated acrania is a rare entity.³

CASE REPORT

The first case is a 26 year old G2P1L1 at 13 weeks diagnosed with acrania by prenatal scan and referred to our institute for management. She was otherwise uncomplicated without any family history and an uneventful prior normal delivery at term. Ultrasound was done for confirmation and pregnancy was terminated by 400µg vaginal misoprostol.

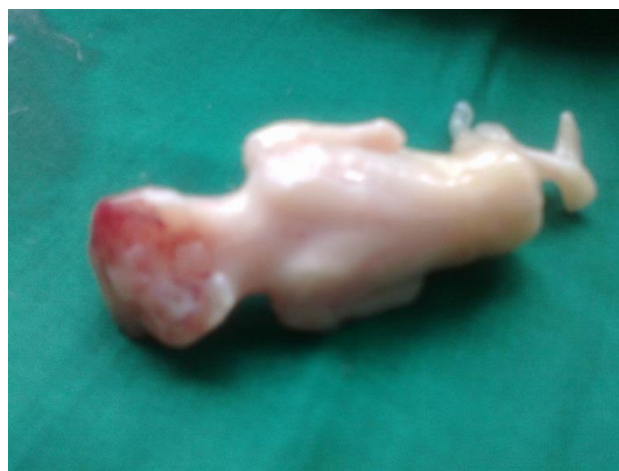


Figure 1: Acrania at 13 weeks.



Figure 2: Acrania at 32 weeks.

The second case is a 28 year old G2P1L1 with previous LSCS at 32 weeks of gestation booked outside. On her first visit to our institute she was clinically suspected to be polyhydramnios, ultrasound was done and acrania of the foetus diagnosed. She did not have any other associated medical complications, no significant family history and her prior pregnancy was uneventful. Induction of labour was attempted with Foley's catheter followed by cerviprime gel which remained unsuccessful, extra amniotic saline infusion also failed. Emergency LSCS was done and a 1.9 kg fresh stillborn female baby delivered with no other obvious congenital anomalies.

DISCUSSION

Incidence of isolated acrania is unknown though it is 1:10,000 when associated with anencephaly. It begins at 4th week of gestation when the anterior neuropore closes. Aetiology may be attributed to embryological fault or disruption by amniotic bands. It results from failure of migration of mesenchymal tissue under the calvarial ectoderm leading to absence of calvarial bones, muscles and dura mater whereas cerebellum, brainstem and cranial nerves are normal. The cerebellar tissue fails to differentiate into 2 hemispheres.⁴

Diagnosis can be made by prenatal ultrasound by 12 weeks by the absence of high echogenic calvarial rim surrounding the brain tissue.⁵ However, to differentiate

this condition from anencephaly ultrasound at 15-16 weeks is required.⁴ Differential diagnosis is with anencephaly (absence of major portion of brain tissue), meroacrania (absence of all calvarial bones except occipital bone), encephalocele (part of brain is intracranial) and mineralisation defects like hypophosphatosis and osteogenesis imperfect.⁶

This congenital anomaly is not compatible with life and hence when diagnosed antenatally subjects the obstetrician to the difficult task of counselling the expecting parents for termination of pregnancy. No evidence regarding its recurrence is available and neither does it increase the chance of open neural tube defect in the future.³

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Ethical approval: Not required

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