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Case Report

A Rare Case of Alobar Holoprosencephaly With Cyclopia in a Male Neonate

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ABSTRACT

Cyclopia is a rare congenital anomaly characterized by a single eye in the middle of the forehead, among other features. This anomaly is usually not compatible with life. It is a serious median facio-cerebral developmental deformity in which the baby is stillborn or dies soon after birth. We present the case of a woman aged 38 years (primigravida) referred to us at 33 weeks of pregnancy and delivered a male macerated stillborn weighing 2.8 kg. The baby had a single central eye, low-set ears, and a proboscis above the eye. The patient was said to have been on a weight-loss supplement, which was discontinued after the confirmation of pregnancy by ultrasound at 6 weeks of gestation. This case report underscores the need for more public enlightenment on the adverse effects of drugs on pregnancy, preconception clinic, regular antenatal visit, and mandatory anomaly scan as part of our routine investigations for at-risk patients, as this will help in early diagnosis and treatment of congenital anomalies.

Key words: Cyclopia, neonate, alobar holoprosencephaly, male, rare, congenital

INTRODUCTION

Alobar holoprosencephaly with cyclopia is a rare and lethal congenital human malformation. [1] The term "cyclopia" was derived from the word cyclops, the single-eyed giants of Greek and later Roman mythology. [2] Cyclopia is a rare congenital anomaly characterized by a single eye in the center of the forehead, among other features. [1–4] The prevalence of holoprosencephaly is about 1 in 11,000 to 20,000 live births, 1 in 250 in embryos. The ratio of female to male newborns with holoprosencephaly at birth is 2:1. The cause of this sex discrepancy remains unknown. [1,5,6] Approximately 1.05 in 100,000 births, including stillbirths, are identified as cyclopean. [2–4] Three forms of holoprosencephaly exist, which are: alobar, semilobar, and lobar varieties. [1,5] Alobar holoprosencephaly is the most severe form of HPE, resulting from incomplete cleavage of the prosencephalon into right and left hemispheres. This occurs during organogenesis between the 18th and the 28th day of gestation. [4] The two lateral ventricles appear as a single ventricle; thalamic fusion is observed, no interhemispheric fissure, optic tracts, or olfactory processes were seen, and the corpus callosum does not exist. Facial development may also be affected as there may be cyclopia, proboscis, ethmocephaly, scaphocephaly, and median cleft lip and palate. [1] There are three types of eye deformities seen in cyclopia: one eye (monophthalmia), two fused eyeballs (synophthalmia), or complete absence of eyeballs (anophthalmia). The fetus in this case report had monophthalmia (one eye). Occasionally, extra cranial characteristics that can present along with Cyclopia are Omphalocele, renal dysplasia, polydactyly, and ventricular septum defect. [3,4]

The semilobar holoprosencephaly and the lobar holoprosencephaly are the intermediate and the mildest forms of HPE, respectively. [1] Alobar holoprosencephaly with cyclopia is incompatible with life, and when neonates are born with signs of life, they often don't survive beyond 10 hours after birth. [5–7] As at the time of this report, literature search revealed that only three cases had been reported in Nigeria, one from the University of Port Harcourt Teaching Hospital, Port-Harcourt, and two from the University of Benin Teaching Hospital, Benin City, Nigeria. [5,6] Herein, we present a unique case of alobar holoprosencephaly with cyclopia, which was incidentally diagnosed when the patient presented with an ultrasound suggestive of polyhydramnios with an active baby at 33 weeks of gestation.

We represent this rare case to create awareness among people about the adverse effects of the injudicious use of drugs during pregnancy, early diagnosis of fetal anomalies if present, and regular antenatal visits.

CASE PRESENTATION

A 38-year-old woman, $G_1P_0+^0$ (primigravida) she was unsure of her last menstrual period, but an early ultrasound scan done at 6 weeks of gestation put her expected date of delivery at February 7, 2024. The estimated gestational age was 33 weeks at presentation. She was referred to our facility on account of an ultrasound diagnosis of polyhydramnios with symptoms of abdominal distention, abdominal pain, and frequency of urination, all of less than 2 weeks duration. There was a history of ingestion of a weight-reducing supplement (name not known), which she discontinued after the confirmation of pregnancy by ultrasound. There was no history of infectious disease, febrile illnesses, or exposure to environmental agents (radiation or chemicals) during pregnancy. She was not a known diabetic, and there was no history of second-degree consanguinity. An anomaly scan done at presentation revealed a single active fetus with good cardiac activity. The thalamus is heart-shaped and fused in the midline. The falx is absent with a single large ventricle. There is marked polyhydramnios (**Figure 1**). No other obvious gross fetal abnormality was seen. A diagnosis of Alobar holoprosencephaly with marked hydrocephalus was made. She was counseled on the poor prognostic outcome of this condition, and she opted for termination of the pregnancy. She had cervical ripening with a Foley's catheter and induction of labor with oxytocin and delivered of macerated stillborn male neonate weighing 2.8 kg with facial congenital anomalies. The abnormalities noted were (**Figure 2**): single central eye, low-set ear, proboscis about 3 cm above the single eye. The parents declined consent for an autopsy.

DISCUSSION

In this case report, we presented a rare case of alobar holoprosencephaly with cyclopia in a male neonate delivered at 33 weeks of gestation at the Federal Medical Centre, Keffi. Cyclopia is the severest facial expression of the holoprosencephaly syndrome. [1] Cardinal facial features of cyclopia may include a median single eye or a partially divided eye in a single orbit, an absent nose or a non-functioning nose in the form of a proboscis above the eye. [1,3,4] As seen in the case presented. Other facial features

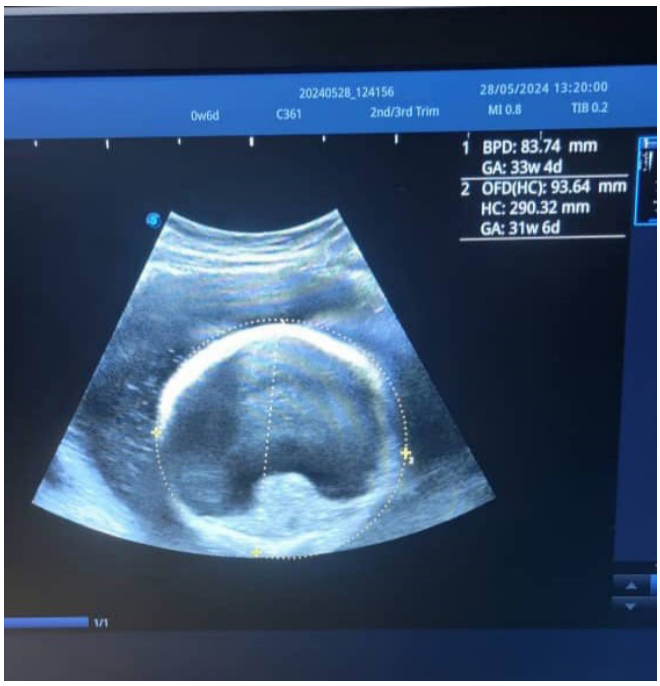

Figure 1: Ultrasound scan showing a heart-shaped thalamus fused in the midline, absent falx with a single large ventricle.

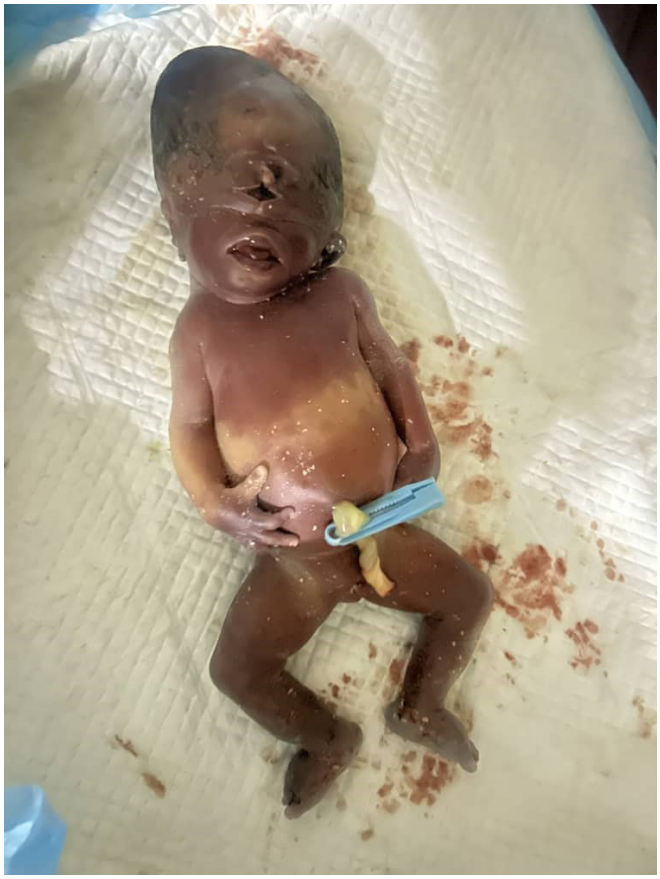

Figure 2: Image of cyclopia male neonate.

are absent philtrum, otocephaly, and astomia or microstomia. No other abnormality was seen in the neonate. Cyclopia can be diagnosed using ultrasonography while the fetus is

growing inside the uterus. [4,5] The patient presented with a scan that reported polyhydramnios. An anomaly scan done at presentation revealed features in keeping with the diagnosis. Holoprosencephaly has been noted to present in more than 16% of cases, with fetal hydrocephalus seen in this case report. The exact cause of this rare malformation is unknown. Still, researchers have reported multiple risk factors, including long-term use of aspirin, statins, methotrexate, poor glycemic control (hyperglycemia), high alcohol consumption, and environmental agents. [3–6] The likely incriminating risk factors in our case are advanced maternal age and the use of a reducing supplement. Although some researchers had argued that there is no correlation between maternal age and holoprosencephaly. [5]

Cyclopic neonates that survive pregnancy are either born stillborn or die shortly after birth. [6] However, a case of a cyclopic baby had been reported to have lived for up to ten years. [7,8] Cyclopia had been observed to be more common in female neonates than males. All three cases reported in Nigeria were females. But in our case report, it was seen in a male neonate.

CONCLUSIONS

Pregnancy is a physiological state that comes with joy. However, this can become distressing when the fetus is diagnosed to have conditions that are not compatible with life. In developing countries, where there is poor utilization of antenatal care services, there is a need for more public enlightenment on the adverse effects of drugs on pregnancy, regular antenatal visits, and mandatory anomaly scan as part of our routine investigations for at-risk patients, as this will help in early diagnosis and treatment of congenital anomalies.

PATIENT CONSENT

Written informed consent was obtained from the parents for publication of this case report.

AUTHORS' CONTRIBUTION

All authors have significantly contributed to the work, whether by following the case at the bedside, conducting literature searches, drafting, revising, or critically reviewing the article. They have given their final approval of the version to be published, have agreed with the journal to which the article has been submitted, and agree to be accountable for all aspects of the work.

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CONFLICT OF INTEREST

None.

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