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A Rare Case of Cyclops Deformity from India

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Abstract

Background: *Cyclopia is a disorder characterized by failure of the embryonic prosencephalon to properly divide orbits of the eye into two cavities. Chromosomal aberrations or toxins can cause problems in the forebrain-dividing process. Its incidence is 1 in 13,000 live births. The nose is either missing or replaced with a non-functional form of proboscis. Most such cases are naturally aborted.*

Case: *A 25 year old primigravida reported with date–size disparity (dates: 23+1, first trimester USG: 21+5, size: 16 weeks) and subsequent USG revealed a dead fetus. Patient gave history of low grade fever in the past. She was induced and expelled a dead fetus of 300 gms which showed features of cyclopia with a proboscis above the single eye. Chromosomal analyses, however could not be done as the fetus was partially macerated.*

To conclude, cyclopia is a rare disorder, incompatible with life, with a complex and multifactorial etiology.

Keywords: *Cyclopia, Holoprosencephaly, Midline forebrain anomaly*

Introduction

Cyclops is a rare congenital abnormality; a severe form of holoprosencephaly resulting in children being born with just one eye. There is failure of the cerebral hemisphere to separate during fetal development. The incidence is 1 in 13,000 live births [1].

Case Report

A 25 year old primigravida presented with ultrasound diagnosed intrauterine death at 23 weeks+1 day (LMP 29/10/2013). On history, she revealed one episode of low grade fever for 4 days, 1 week prior to admission, for which no treatment was taken. There was no other significant history.

Systemic examination was normal. On local examination, uterus was 16 weeks size, the cervical os closed and uneffaced.

Laboratory features also did not reveal any abnormality.Hb was 10.7 gm%, TLC 7,500 /cumm, Differential count normal, MCV 94.8 fl, MCH 33.6 pg, MCHC 33.5 gm%. Urine routine microscopy revealed no abnormality. Random blood sugar was 105 mg %. Thyroid, liver and renal function tests were normal.HIV, HBSAg and VDRL was non-reactive. Coagulation profile normal. Patient had 2 ultrasounds both showing live pregnancy of8 weeks +6 days and 9 weeks+3 days respectively with no comment on gross abnormality in the fetus. A third ultrasound prior to induction showed an anomalous dead fetus of 15 weeks+6days with no details. The patient was induced, delivering afetus of 300 gms showing a single eye orbit with a proboscis above and no nose. Placenta was normal. Chromosomal analysis however could not be done due to partial maceration.

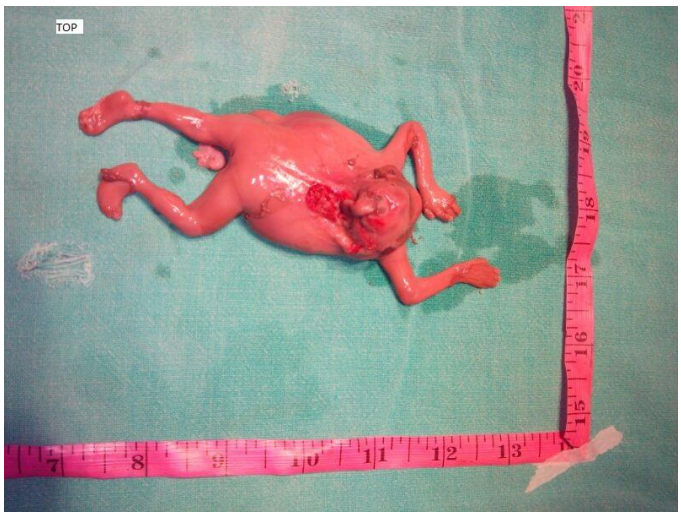

Figure 1: Fetus with linear scale showing a grotesque face, single eye orbit, no nose and a proboscis above the orbit. Hands and feet appear normal.

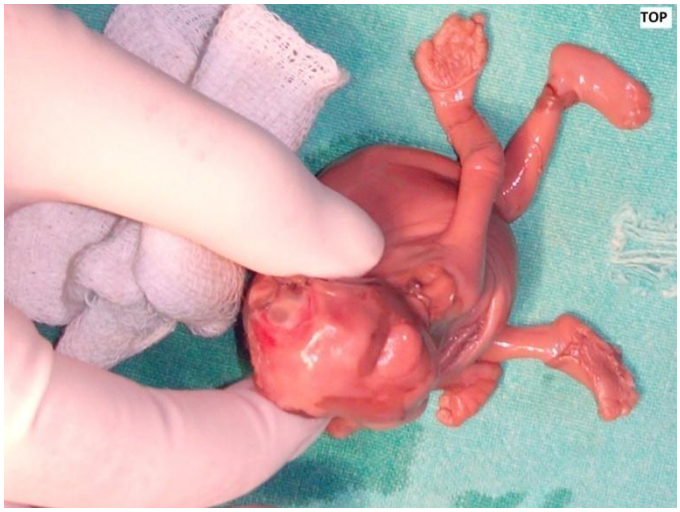

Figure 2: Fetus with close up view of deformed face with mouth, absent nose and single eye orbit

Discussion

During human neural development, the neural plate appears by day 16 and forms a tube by day 23. The rostrum closes first by days 24, 25, followed by the caudal end by day 28.Holoprosencephalymay arise if there is failure of the forebrain to divide by day 33 and its alobarform is the most severe. It can be diagnosed ultrasono graphically by 7 to 9 weeks by non appearance of the butterfly sign and a single large ventricle. Normally, humanoid features appear by 8th week. In cyclopia however, there is a single eye orbit and an absent nose, sometimes replaced by a proboscis above the orbit.

The etiology of cyclopia is multifactorial but one of the direct causes may be ingestion of a plant called Veratrumcalifornicum mistaken for one taken for relief of nausea during pregnancy, yielding an alkaloid cyclopamine (anti-cancer agent) known to cause cyclopia in sheep [1,3]. The mechanism implicated is inhibition of the Hedgehog Signalling Pathway, important for embryonic forebrain cleavage[2].Abnormality of theZic2, SIX 3and TGIF gene have also been implicated [1,2]. Other causes

include diabetes, TORCH infections, ingestion of retinoic acid [2]

Conclusion

Cyclopia is a rare condition, having an incidence of 1:2500 that end in miscarriages [1]. In its severe form it is incompatible with life. Very few cases of full term deliveries have been reported in literature and all end in death.

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