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Case Report

Anatomy Section

True Cyclopia–Very Rare Anomaly

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ABSTRACT

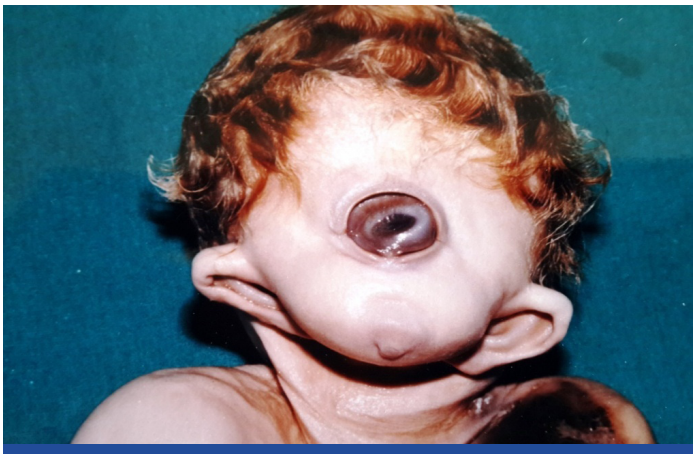
During our routine postmortem examination, we found a female infant with "True Cyclopia." The two entire eye balls had fused completely to form one midline organ. The infant was a female who weighed 2910 g. The gestation period was 35 weeks. Infant lived for a short period of less than 20 minutes. The birth was a product of a 4th pregnancy.

Keywords: Cephalic presentation, Gestation, Hexcaprone, Intrauterine contraceptive device, Micrognathia, Osseous defects, Respiratory distress, Proboscis

CASE REPORT

The mother was a 36-year-old woman. Mother's all previous pregnancies and deliveries were normal. An intra uterine contraceptive device (IUCD) was inserted. A course of hexcaprone (tranexamic acid) for five days and ampicillin 500 mg were given for two weeks, to prevent haemorrhage and infection, after introduction of an IUCD. Three weeks before the baby's birth, the mother was exposed to X–Rays for the diagnosis of some other complaint. Infant was born spontaneously, with a cephalic presentation.

Infant moved her hands and legs moderately and died after 20 minutes of birth, due to respiratory distress. The appearance of the body and limbs was normal. The face had a median orbit and a single large eye ball with a pair of eyelids. There was no mouth, nose and proboscis. Ears were present at a lower level, in the ventral aspect. Micrognathia was present. Multiple, ancillary, facial and osseous defects were noticed. Mouth was replaced by a plug of soft tissue [Table/Fig-1].



[Table/Fig-1]: Our case report photograph

DISCUSSION

Stated that Cyclopia referred to a rare foetal malformation which was characterized by a single, palpebral fissure and a single midline orbit. This orbit could contain either a single globe or two separate globes. In true Cyclopia, organo-genetic development of the two separate eyes is suppressed [1]. Exposure to drugs or to other teratogenitic environmental factors is regarded as the basis of this anomaly. These factors may be ionic radiations, contraceptives, viraemia plus corticosreroids and salicylates, Rubella vaccine, antibiotics and amidopyrine (aminopyrine) [2,3]. Genetic errors with chromosomal abnormalities such as trisomy-D, monosomy – G, mosaicism, translocations which affect chromosome 3 and group C chromosomes and deletion of short arm of chromosome 10, have also been recorded in cyclopia. The above factors must act before the first manifestation of the bilateral nature. The two chief mechanisms are defective inheritance and an unfavorable environment. In a human embryo, this occurs at 1.8 mm stage or during third week of gestation. Defects which occur in the prochordal mesoderm, which are caused by mechanical, genetic or environmental teratogens, can lead to arrest or malformation of facial bones, thus leading to micrognathia.

"Cyclopia", which is a very rare anomaly of organo-genesis of the eye, results from arrest of development of the anterior end of the neural plate. Thus, cyclopia is always associated with abnormalities of the brain. This anomaly was classified by, De Myer et al., [4]. Torczynski et al., reported data which was based on the analysis of a series of 35 cases of synophthalmia / cyclopia. Seventeen of 35 cases were females, nine were males and for the remaining nine (9), the sex was not stated [1]. Cohen and Gorlin reported a case and considered the genetic aspects of holoprosencephaly [5]. In addition to their case, with "Parenteral Consanguinity," they mentioned two other cases which were reported by Klopfstock in 1921 and Grebe in 1954. Francois suggested that "Cyclopia is a lethal Autosomal Recessive Trait [6]." Gorlin et al., suggested that some cases of holoprosencephly may be caused by an autosomal recessive gene in individuals with normal karyotypes and they suggested that the responsible gene was located on the short arm of chromosome 18 [7]. Karseras and Laurence concluded that normal chromosomal studies did not necessarily exclude an underlying chromosomal disorder, since this may not have been discovered by earlier methods and could still escape detection by current methods [8].

CONCLUSION

Making a pre natal diagnosis of Cyclopia by ultrasound examination can help in preventing complications which are associated with Cyclopia, through careful usage of antibiotics, exposure to ionic radiations, contraceptives, corticosteroids, salicylates, rubella vaccine and amidopyrine.

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