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CASE REPORT

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A rare case of cyclops with hydrocephaly

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Introduction

Cyclopia is an abnormality characterized by a single median eye located in the area normally occupied by the premaxilla (incisive), and nasal bones and cartilages. Infants with cyclopia or sirenomelia are born at an approximate rate of 1 in 100,000 births [1].

Case report

Mrs. SM, a 22 year old woman married for 1½ years, presented at the antenatal outpatient clinic, on 16th August, 2002. Her LMP was on 24th February, 2002. Sonography done on 9th August, 2002 showed fetal early hydrocephalus of the non-communicating type (Figure 1).

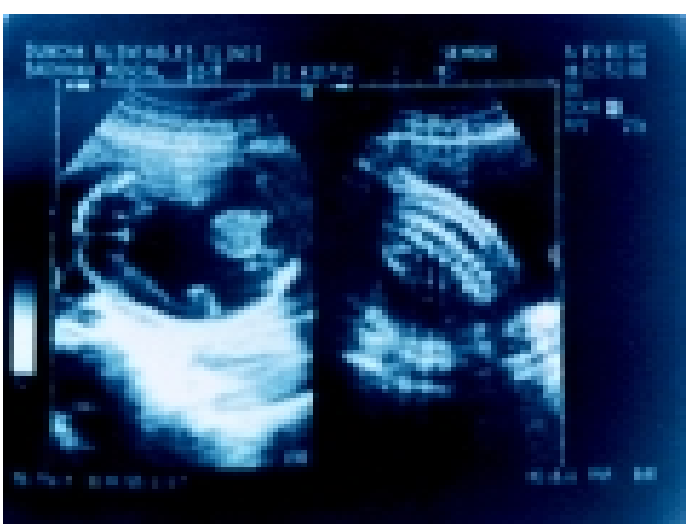

Figure 1. Sonography showing hydrocephalus

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She was G_3P_0 with a history of two spontaneous first trimester abortions. There was no significant past medical, surgical or family history in both the partners. She wished to continue the pregnancy and pediatric and neurosurgical opinion was taken regarding prognosis of the fetus. They advised continuation of pregnancy and surgical management after delivery. After preliminary obstetric examination revealed other obstetric parameters to be within normal limits, she was advised routine antenatal medications and told to come for regular check-ups. All routine investigations were within normal limits. On 27th September, 2002 the maternal serum α fetoprotein level was 69.0 IU/mL. Routine antenatal examination was done and antenatal medication continued.

On 8th October, 2002 at 9.55 am, she was admitted as an emergency case with complaints of pain in abdomen, and vaginal leaking and loss of fetal movement since the last 12 hours.

On examination her vital signs were stable. Abdominal examination revealed a uterus of 34 weeks size. Contractions were normal and fetal heart sound could not be located. The fetus was presenting with breech and the head seemed to be larger than the expected size. On vaginal examination, the cervix was 6 cm dilated and 80% effaced. The presentation was breech at 'O' station. The membranes were absent with vaginal leaking.

She was shifted to the labor ward where a decision for assisted breech delivery with craniotomy of the aftercoming head (if necessary) was taken. Within 2 hours the cervix was fully dilated and effaced. There was good uterine activity. After delivery of the shoulders, craniotomy of the aftercoming head was done and a fresh stillborn female baby was delivered at 12.30 pm. The placenta expelled spontaneously and there was a large retroplacental clot.

The baby was a fresh still born female, weighing 2 kg with a single central eye and a half formed nose above the eye on the collapsed hydrocephalic head (Figure 2). No other apparent anomaly was seen. Karyotyping of the fetus was advised but the woman could not afford it.

Figure 2. The cyclops

Discussion

The possible etiologies of this disorder include autosomal dominant or recessive and monogenic inheritance, infection (toxoplasmosis), and drug (hydantoin) toxicity and maternal conditions such as gestational diabetes have been also implicated [2]. There is also some evidence for a defect in the cholesterol biosynthesis [2]. This apparent diverse origin of cyclopia can be clarified if in future, cyclopic specimen are carefully investigated. The evaluation should include a careful gross and microscopic examination of all organs, including the eye and chromosome banding studies of at least two cyctopic tissues [2].

References

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