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Gastroschisis

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CASE REPORT

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ABSTRACT

We report a case of gastroschisis which was not diagnosed antenatally and was delivered through lower segment caesarean section due to non-reassuring cardiotocograph and small for gestational age fetus in a 21-year old mother. It was associated with oligohydramnios and partial extension of wrist joint in the neonate. After delivery, baby was referred to tertiary care for specialized care by paediatric surgeon and neonatologist where he had silo reduction and surgical repair. Postnatally, the baby is in healthy condition till now.

Key words: *Gastroschisis. Wrist joint deformity. Oligohydramnios.*

INTRODUCTION

Gastroschisis is herniation of abdominal contents through a cleft in the anterior abdominal wall, particularly the intestine and stomach. Viscera are not covered by peritoneum or amnion, and the bowel may be damaged by exposure to amniotic fluid. Loane *et al*. reported increase in prevalence of gastroschisis in Europe.[1] Vu *et al*. showed that prevalence has been increased to 2.6 per 10,000 births in California from 1987 – 2003.[2] Although survival rate is extensively variable, gastroschisis still accounts for a significant fraction of stillbirths. The study from Europe reported 6% fetal death and 20% terminated pregnancies over a period of 22-year.[1]

In Pakistan, no study has been conducted to determine the prevalence of gastroschisis or any possible associated factors. Hereby, we report an antenatally undiagnosed case of gastroschisis despite several detailed prenatal ultrasounds. Oligohydramnios, partial extension of wrist joints and reduced fetal heart variability were also reported which are rare for the condition.

CASE REPORT

A 21-year old female, second gravida (first-trimester complete abortion) visited to The Aga Khan Hospital for Women, Karimabad, for booking at 11th week of gestation. She paid 9 regular antenatal visits during her pregnancy. Family history was insignificant except that the father was diabetic. Anomaly scan performed in 24th week of gestation reported normal anatomy. Group B streptococcal infection was picked-up at 34th week of gestation. Growth scan was performed in 36th week, reporting the fetus with parameters corresponding to 30th week with no congenital abnormality.

Fetus was diagnosed to be small for gestational age (SGA) along with oligohydramnios. She was admitted to the department for Induction of Labour (IOL) due to small for gestational age case and good Bishop score. Labour was induced by administration of Prostaglandin PGE2 tablets, and later on membrane was artificially ruptured for augmentation of labour. GBS prophylaxis injection Benzyl penicillin 3 g stat and 1.5 g six hourly was given during labour. The fetal heart record showed reduced variability and second grade meconium was passed. History of the mother taken just before the delivery reported vomiting, frequent urination, dyspnea, constipation, discharge and headache. Blood pressure was low (100/70 mm of Hg). The blood group of mother was AB+. Her routine antenatal work-up was normal. Emergency lower segment caesarean section was performed in secondary care hospital under spinal anaesthesia.

An alive male baby weighing 2100 g was delivered with good APGAR score. Although not diagnosed by any of the antenatal ultrasound reports, Gastroschisis was found with protrusion of part of small intestine, so the baby was immediately sent to the related tertiary care hospital after detailed counselling of couple regarding the unusual finding and closure of anterior wall defect was done over there. Interestingly, the wrist joints were only partially extensible. After 3 months, mother brought the baby with repaired abdominal wall and baby was doing well.

DISCUSSION

Gastroschisis is a developmental anomaly with defect in anterior abdominal wall allowing the protrusion of intestine, stomach and, at times, even the urinary bladder.[3] Studies from the United States report a varying prevalence rate in different regions, but all the studies

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suggest an increase in the prevalence. A longitudinal study in Utah reported an increase of prevalence from 0.36 to 3.92 cases per 10,000 births over a period of 31 years.[4] The data from California Birth Defects Monitoring Program reported the prevalence to be 2.6 cases per 10,000 births.[2] It was also reported that mothers of age group 12-15 years had a 4.2 times greater birth prevalence.[2]

Gastroschisis is diagnosed through antenatal sonography in the second trimester. Studies have proven antenatal sonography to be the most accurate and sensitive diagnostic tool for gastroschisis.[5] When diagnosed antenatally, mothers are kept in tertiary care departments with immediate facilities of operating the condition. Although this case was a 36th week delivery and was subjected to several assessments during the antenatal visits, still it remained undiagnosed and the mother was kept in secondary care and the newborn was then transferred to the tertiary care department. Hence, correct diagnosis through ultrasound cannot be assumed in every case.

Studies have also reported incidence to be more common in newborns of younger mothers.[1] The possible cause can be the undersized uterus unprepared to support the fetus. In Pakistan, especially the ratio of teenage mothers is quite high yet there are no studies to determine the incidence of gastroschisis.

There was a reduced fetal heart-rate variability in this case. Although reported earlier, this is a rare combination with gastroschisis.[6] Ingamells *et al*. reported 7 cases diagnosed gastroschisis with reduced fetal heart-rate variability and each fetus died on birth.[6]

Similarly, the partial extension of wrist joint is a very rare association, although a few cases have reported wrist deformities in association with gastroschisis.[7] In this case, three rare conditions (antenatal non-diagnosed, reduced fetal heart-rate variability and deformity of wrist joint) have been combined and which makes it the first report of its kind.

The development of gastroschisis has long been debated. Five hypotheses have been proposed which include failure of mesoderm formation in body wall, para-umbilical rupture of amnion with succeeding herniation of intestine, abnormal degeneration of right umbilical vein, disintegration of the right vitelline artery and abnormal folding of the ventral body wall.[8] Various studies have been conducted on animal models to determine the factors involved in the folding of ventral body wall.[9,10] Ogi *et al*. reported mutations of Msx1 and Msx2 genes might lead to abdominal wall defects.[9] However, a detailed and confirmed explanation of the disorder still awaits further researches.

The cause in this case could not be determined. Also notable are the positive Rubella and Streptococcus-B in mother. There are no previous reports that determine any association between these infections and gastroschisis.

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