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Case Report

Intrapartum Challenges in a Term Fetus with Anencephaly: Case Report

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Abstract

Introduction: Anencephaly is a lethal congenital defect due to the failure of closure of the cranial end of the embryologic neural tube. Termination of pregnancy is offered for all prenatally diagnosed cases, but some parents choose to continue with pregnancy with full obstetric intervention.

Case Report: I describe the case of a 36 year old woman who had five pregnancies with anencephalic babies terminated before 5 months and in current pregnancy pre-natal ultrasonography at 12 weeks revealed an intrauterine anencephalic live gestation. She refused pregnancy termination and developed Gestational diabetes and polyhydramnios in the course of pregnancy and delivery was complicated by intrapartum shoulder dystocia, obstructed labour and delivered a full term anencephalic female baby who was still born.

Conclusion: Anencephaly is a severe neural tube defect resulting in postdated pregnancy and prolonged labour. This can result in shoulder dystocia which may have adverse consequences for mother. Overall, the incidence of obstetric complication is less however, there is a tendency towards delivery via repeated caesarean section in women with a previous uterine scar and anencephaly. Management of pregnancies reaching term with anencephaly is not based on any standard protocols and thus there are dilamatous situations that need consensus for patient safety. This case raises important practical questions: Management strategies of recurrent pregnancies with anencephaly includes best supportive care during and after birth and counseling the expectant parents.

Keywords: *Ultrasonography; Anencephaly; Shoulder Dystocia; Obstructed Labour; Gestational Diabetes; Polyhydramnios; Recurrent Anencephaly*

Introduction

Anencephaly is a severe congenital neural tube defect that is incompatible with life. The prevalence remains at 2.1/100,000 live births 1st-year mortality rate for this neural tube defect is of 100%. This deformity is predominance in females and Caucasians with a multifactorial polygenic etiology. Prenatal diagnosis can be made reliably by testing maternal serum α-fetoprotein levels and ultrasonographic imaging of fetal brain [1]. Most of the parents decide to terminate the pregnancy on diagnosis, a few families choose to continue even after counseling about poor prognosis and survival rates mainly due to religious beliefs [2].

It is an important aspect to address all the potential possibilities of maternal outcomes with parents, especially in case if pregnancy termination is not, or no longer an option. The decision about pregnancy termination is associated with parents' religious beliefs and restricted legal termination rules, in addition to other factors such as late detection of the anomaly and delayed fetal anatomy scans. Commonly, the major reason for continuation of pregnancy is the request of parents due to religious beliefs.

Shoulder dystocia is an obstetric emergency. The rate of shoulder dystocia is about 1.4% of all deliveries and 0.7% for vaginal births. Typically defined as a delivery in which additional maneuvers are required to deliver the fetus after normal gentle downward traction has failed, Shoulder dystocia can be an obstetrician's nightmare, usually occurring unexpectedly. There can be serious consequences for the mother and a fatal outcome for the fetus. It happens when the fetal shoulders impact at the pelvic inlet following delivery of the head [3].

Case Report

Mrs. H, a 36 year old Pakistani housewife, married to her first cousin, Gravida 7 Para 1Live 1 Abortion 5, attended AQWCH at 35 weeks of gestation for check up in the clinic. She a history of previous five pregnancies terminated due to anencephaly, three pregnancies at around 20 weeks and rest at 15 - 16 weeks. Last pregnancy ended as a cesarean section in 2018 for breech presentation, for a girl child weighing 3.7 kg. She was diabetic in that pregnancy and was taking metformin and insulin.

She never had any gynecological problems in the past with regular menstrual cycles prior to conceiving. She underwent evacuation of retained products of conception following medical management of previous 3 pregnancies complicated with anencephalic babies and had a cesarean section in her last pregnancy. Interestingly, she has history of pregnancies with anencephaly in her family. She has not taken pre-pregnancy folic acid.

Patient was booked in a private clinic since 12 weeks on regular follow up and presented to our facility at 35 weeks. Her last menstrual period (LMP) was on 6th of November 2019 and her expected date of delivery (EDD) Naegele rule) was 24 September 2020 (as per 12 weeks scan). It was a planned pregnancy. Scan done at 12 week at private hospital showed Acrania for which patient refused termination of pregnancy. Anomaly scan done at private confirmed the findings. She had glucose tolerance test (GTT) at 24 weeks, GDM was diagnosed and was taking Tab metformin 500 mg TID from 32 weeks. Sugars were well controlled with medication. Her Blood group O negative was similar to her husband's blood group.

All routine baseline investigations at booking were done and were normal.

Patient had her last scan at 36 weeks estimated fetal weight was 2.5 kg and DVP was 9 cm (mild polyhydramnios). At 38 weeks her sugars were well controlled, vaginal examination indicated a favorable cervix (3 cm dilated, 2 cm long, medium consistency). In view of her current pregnancy being one with anencephalic fetus with previous caesarian section patient opted to wait for spontaneous on set of labour till term, wanting to avoid repeat cesarean section at all costs. She did not want to affect her future obstetric career due to 2 cesarean section on herself and preferred spontaneous and vaginal mode of delivery the most.

Patient was admitted at 40 weeks with spontaneous onset of labour pains, with stable vitals and general condition. Obstetric examination revealed a uterus corresponding to 36 weeks pregnancy with no scar tenderness and moderate amplitude contractions, occurring 3 in 10 minutes. She had a favorable bishop score (5 cm dilated and 1 cm long, soft cervix) and soon had SROM progressing to 2nd stage of labor in 3 hours, which persisted for more than 1.5 hours Episiotomy was given and vacuum was applied over the forehead on maternal request to avoid cesarean section, as maternal effort was poor then. Shoulder dystocia was encountered and each maneuver was tried unsuccessfully. Even after continuous traction, shoulders were not reachable for rotational maneuver and posterior arm delivery.

Patient was shifted to OT and under General anesthesia and again all maneuvers were tried and after 90 minutes of traction delivered a female still born fetus weighing 3360 gms. Patient sustained a 3A perineal tear which was sutured in layers, after appropriate visualization. Multiple superficial tears in the vagina sutured and pack was kept and urinary catheter was inserted. Intravenous antibiotics and analgesics were given.

Patient developed postpartum urine retention after removing urinary catheter in 24 hours after delivery. USG KUB was done-mild renal fullness noted, and urologist was involved. After keeping catheter for 24 hours patient passed urine freely and residual urine was less than 50 ml on ultrasound.

Patient was discharged on day 4 after delivery with oral antibiotics and she came for 6 weeks postpartum checkup without any bowel or bladder complaints.

Discussion

Anencephaly is a severe defect of development of the neuraxis, in which the developing forebrain and variable amounts of the brainstem are exposed in utero and fail to develop or are destroyed [4]. Around postovulatory day 25, failure of the rostral neuropore to close causes anencephaly to occur [5], whereas spina bifida results from failure of the caudal neuropore to close around postovulatory day 27.

Recurrence risk for neural tube defects (NTDs) (spina bifida or anencephaly) is approximately 2 to 4 percent with one affected sibling [6,7] and approximately 10% with two affected siblings [8]. May factors like genetic factors, environmental influences, or a combination of both are responsible for the familial clustering [9].

Ultrasonography is the mode of prenatal diagnosis showing absence of brain and calvaria superior to the orbits on coronal views of the fetal head. Another feature that may develop in upto 50 percent cases is polyhydramnios, which becomes evident particularly in the second and third trimester due to decreased fetal swallowing.

Risk factors for NTDs include dietary deficiency of folic acid, administration of valproate or folic acid antagonists such as trimethoprim, carbamazepine, phenytoin, and phenobarbital, and genetic polymorphisms in genes encoding folate-dependent enzymes. Maternal diabetes mellitus with poor glycemic control during the first trimester, hyperthermia, and some genetic syndromes could be some other contributors to NTD risk [10].

Prevention is the key aspect in management of anencephaly, due to paucity of many neurosurgical options. Nearly all live born infants die shortly after birth. Multiple reports published prove a link between folic acid deficiency and the development of neural tube defects (NTDs) including anencephaly. Measures like pre-pregnancy diet supplementation with folic acid extending into the 1st month of pregnancy have been recommended to decrease both the frequency and severity of the condition. Fortification of foods with folic acid has also been recommended. Early detection of this anomaly, by the implementation of the program of the prenatal diagnosis is another secondary line of prevention. The knowledge helps diagnose and treat neural tube defects [11].

Shoulder dystocia is an obstetric emergency because it may result in life-threatening infant injuries, as well as less serious maternal injuries. This needs additional obstetric maneuvers beyond gentle guidance to enable delivery of the fetal shoulders, avoiding trauma to the mother maximally.

Shoulder dystocia in anencephalic pregnancies is an expected complication that can be attributed to diminished head size, which cannot dilate the cervix enough to deliver the fetal trunk and shoulders. While early detection with termination should be the preferred algorithm, ongoing anencephalic pregnancies are associated with higher probability of cesarean operations for non-viable babies. With prolongation of pregnancy being a possibility in anencephalic pregnancies reaching advanced gestational ages due to fetal hypothalamic-pituitary axis dysfunction, women with previous cesarean sections must be made to understand the risks of repeat cesarean [12].

In the above case, the women she had a consanguineous marriage which might be a reason for recurrent Anencephaly Patient chose to continue pregnancy despite knowing it is anencephalic fetus. Near term, she hoped for a vaginal delivery. In all efforts to avoid re-

peat cesarean section for an anencephalic baby shoulder dystocia and obstructed labor occurred. While it was managed diligently with minimal maternal morbidity, the above case is a good example to highlight the importance of accurate weight estimation of fetus close to delivery, which itself is challenging in a case of anencephaly due to difficulty in measuring BPD and HC. Thus, experts with customized charts using abdominal circumference, which is the only reliable measurement can be used to get the best estimated weight. Even though she had a prolonged labor with third degree perineal tear, this patient was still happy at avoiding a repeat cesarean section even though she preferred to continue this pregnancy, which proves that maternal satisfaction is more when they are involved in decision making. In support from literature is the article by Cope., *et al*. that continuation of pregnancy in lethal fetal defects may have psychological benefit to women compared to termination [13].

Conclusion

Couples expecting an anencephalic fetus must be given a realistic picture when counselling about the management options including all challenges of continuing the pregnancy before they make their decision. High incidence of cesarean deliveries, shoulder dystocia, polyhydramnios and associated complications including obstetrical hemorrhage risk must be clearly explained with minimal benefit considering a non life expectant baby. Any delay in delivery of a anencephalic fetus during labour must be treated with caution and instrumental delivery is best avoided. In pregnant patients with previous scared uterus, repeat cesarean section should be preferred anticipating macrosomia, possible prolonged labor and risk of scar rupture, as this only increases maternal morbidity. Involvement of patient in decision making is prime-most in such cases and plans should be agreed mutually in the best interest of both physical and mental health of the couple, particularly when there is a recurrence possible.

Conflict of Interest

Nil.

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