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Case Report

A case report of body stalk anomaly at 15 weeks and management in a patient with a previous cesarean

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Abstract

Body stalk anomaly is a fatal disfiguring abdominal wall defect resulting from abnormalities in the development of the cephalic, caudal, and lateral embryonic body folds[1]. The inability to fold in the correct axes leads to these body wall deformities. Daskalakis et al. reported an incidence of 14/106,727 fetuses[2]. There are few cases of this complex in the United States and little is known about the cause of this abnormality. Proposed mechanisms include early amnion rupture, vascular disruption of the early embryo, or an abnormality in the germinal disk (X). Given that body stalk anomaly is a fatal anomaly, early diagnosis is critical to the counseling and management of affected patients. Herein, we present a case of body stalk syndrome and discuss management in a patient with a history of a prior cesarean delivery who declined termination.

Keywords: Body stalk anomaly, body stalk syndrome, abdominal wall defects.

INTRODUCTION

CASE: A pregnant patient presented as referral to Maternal Fetal Medicine (MFM) for evaluation of a 'right ovarian cyst seen' on patient's anatomy ultrasound and an abnormal quad screen. MFM ultrasound revealed multiple body wall anomalies concerning for the diagnosis of body stalk syndrome.

CONCLUSION: Several abdominal wall defects could present as body stalk anomaly. Despite this, body stalk anomaly is considered invariably fatal and so the safest mode of delivery should be chosen for patient. Prompt diagnosis is also essential for the multidisciplinary approach needed in the care of these patients which should include Palliative care.

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CASE

A 27 year old multigravida at 18 weeks 2 days gestation (GA) was referred by her primary OBGYN to the Maternal Fetal medicine clinic due to concerns for a 'right ovarian cyst' and an abnormal quad screen (AFP: 3.03 MOM). She was referred for a detailed ultrasound and her fetus was found to have multiple fetal anomalies. Visualised intracranial structures were noted to be normal, however the abdominal wall was in close association with the anterior placenta and no umbilical cord was seen. The fetus was also noted to have a large cyst loosely attached at the sacrum, one lower extremity with clubbed foot, short long bones and absent ribs. Other abdominal organs including stomach, liver and spleen were also noted to be in abnormal locations of the body within the thoracic cavity. Amniotic fluid volume was adequate (Fig 1).

At this time, a diagnosis of body stalk anomaly was made. Given this, she was counseled on the poor prog-

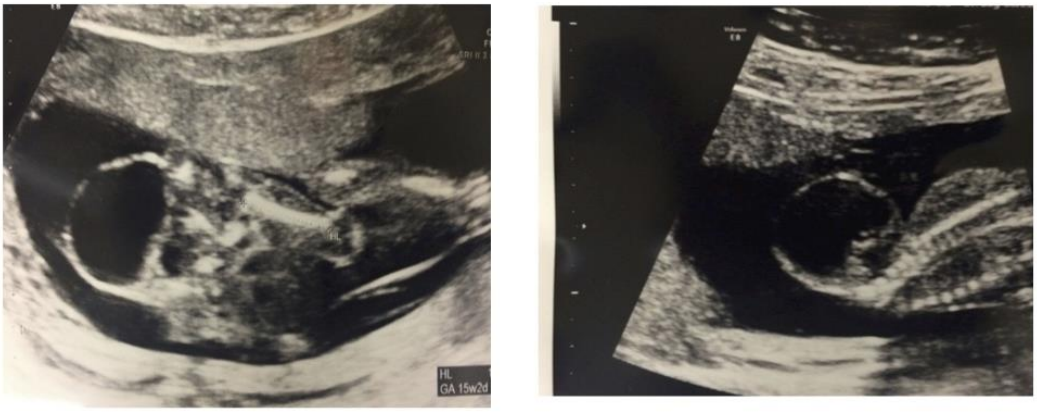

Fig. 1. Ultrasound images of fetus at 18 weeks gestation.

nosis of this diagnosis, but she declined a termination and decided to follow up with her OBGYN provider.

At 25 weeks and 5 days GA, she was transferred to our hospital after presenting with complaints of 'leaking of fluid' to her physician. She was found to have ruptured membranes and given a diagnosis of preterm premature rupture of membranes. Fetal ultrasound was repeated at that time showing similar findings outlined above. Given the fatal fetal anomaly she was counseled towards induction, but elected for expectant management and was admitted to the Antepartum service. The following day, after further discussion with her partner, she decided to proceed with induction.

Her surgical history was significant for a low transverse cesarean section at 37weeks with delivery of a 4lbs 7oz infant noted to have bilateral club feet. The patient was counseled for trial of labor after cesarean (TOLAC) to minimize maternal morbidity given the lethal fetal anomaly versus repeat cesarean section. The patient agreed to proceed with a TOLAC after understanding of risks and alternatives. The induction was started with cervical ripening with Misoprostol 800 mcg intravaginally for 24 hours then transitioned to mechanical cervical ripening with foley bulb and pitocin after delivery was not achieved. Twenty two hours later, she delivered a non-viable male vaginally with Apgars 0 / 0 and weight 875g. The fetus was noted to have abnormal facies, 10 fingers and 10 toes seen (Figure 2). Lower limbs were twisted, with one club foot seen. There was an anterior abdominal wall deformity with visualization of liver, spleen, and intestines connecting to placenta, no clear umbilical cord was identified.

DISCUSSION

Abdominal wall defects occur sporadically during organogenesis. Included in this category are 3 main abdominal wall defects: omphalocele, gastroschisis and body stalk syndrome. Omphalocele and gastroschisis should be included in the differential diagnoses for body stalk syndrome.

Body stalk anomaly is the least prevalent with a report of 0.32 per 10,000 births seen in large epidemiological studies[3]. The first report of body stalk anomaly was published in 1986 by Lockwood et al[1]. The severity of the defect varies based on the degree of abnormal folding occurring in each of these axes. The diagnosis of body stalk syndrome is characterized by the development of the abdominal organs outside of the abdominal cavity in a sac of amnioperitoneum[4].The pathogenesis of this development is not well understood but there are a few hypotheses to explain this defect. One of these is the disruption of vascular flow due to an inciting event to the mother, but then negatively influences normal embryonic blood supply, thereby interrupting normal morphogenesis[5]. A case report of body stalk syndrome associated with cocaine abuse supports this hypothesis[6]. The second hypothesis, early amnion rupture theory proposes that the anomalies in body stalk syndrome result from multiple amniotic bands interrupting embryogenesis. Finally, the Streeter theory of embryonic dysgenesis suggests improper histogenesis resulting in disturbances in embryonic folding[7,9].

On prenatal imaging, the fetus can be noted to have massive abdominoschisis, severe kyphoscoliosis, and a relatively short umbilical cord[4]. There is usually no need for invasive testing as all cases are karyotypically normal. These cases are usually considered sporadic and so recurrence is usually not seen. The typical features of body stalk anomaly can be detected by ultrasound by the end of the first trimester, which is important in patient management, given the poor fetal prognosis. Consequently, body stalk anomaly should be distinguished from other fetal abdominal wall defects.

It is essential to discuss the optimal mode of delivery for these patients. A review published by Kuller et al.discussed that cesarean delivery may improve neonatal outcome for fetuses with isolated meningomyelocele,

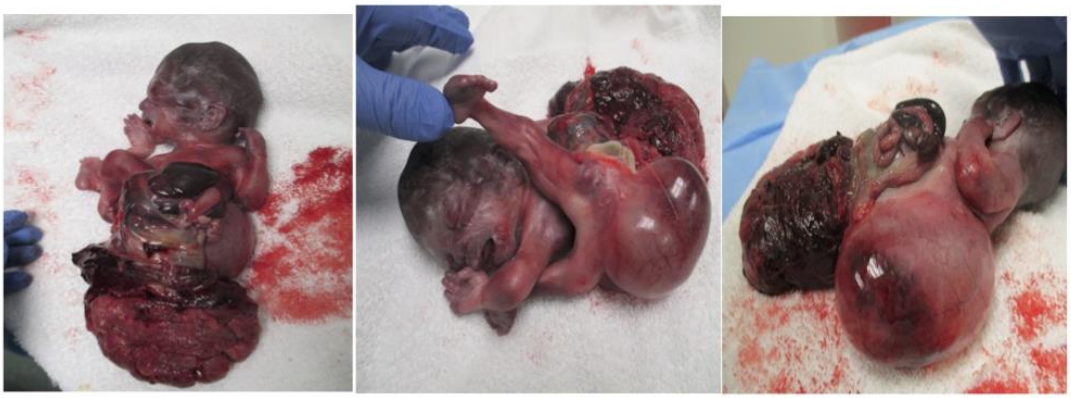

Fig. 2. Images of fetus born 25 weeks 5 days gestation.

hydrocephalus with concomitant macrocephaly, anterior wall defects with extracorporeal liver, sacrococcygeal teratomas, hydrops, and alloimmune thrombocytopenia with low platelet count at term[8]. However, given that body stalk anomaly is considered invariably fatal, the mode of delivery with the least maternal morbidity should be considered for patients even if this involves induction of labor with a history of cesarean delivery. In order to provide the best care for patients, a multidisciplinary team that includes Neonatal Palliative care and chaplain services (per patient's request) is essential.

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