

Body Stalk anomaly

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Abstract: Body stalk anomaly is a rare developmental anomaly characterized by an abdominal wall defect, severe kyphoscoliosis and a rudimentary umbilical cord along with other organs' anomaly. It is diagnosed by ultrasonography, mostly in the second trimester. No treatment option is there for this universally fatal condition. We here present a case of term, live born female baby with body stalk anomaly.

Keywords: Body stalk anomaly, etio-pathogenesis, treatment.

INTRODUCTION

Body stalk anomaly, also known as limb-body wall complex is a lethal malformation of the thorax and abdomen, where intra-thoracic and abdominal organs lie outside the body cavity. The organs are usually contained within a sac of amnio-peritoneal membrane attached directly to the placenta [1]. The umbilical cord may be totally absent or extremely shortened. It is usually associated with limb defects and severe kyphoscoliosis. This anomaly might also occur in conjunction with neural tube defects, genitourinary malformations, intestinal atresia, and craniofacial defects, among others [2].

CASE REPORT

A 26years old, unbooked second gravida mother was referred to our hospital for non progression of labour. Her antenatal period was uneventful. There was history of consanguinity but no family history of malformation was there. No history of teratogenic drug intake during pregnancy. Antenatal USG was not done. A live born female baby was born by emergency cesarean section. Birth weight was 2.5 kg. On examination (Fig A & B), the baby was normal facies with low set ear. Head circumference was 34 cm. But there was evisceration of all abdominal contents including liver (liver was bi-lobed but right and left lobe was almost similar in size), spleen, pancreas, intestine, a spherical right kidney and normal ureter through a gap in left side of the body. The contents were covered by a peritoneal membrane. There was ectopicacordis also with herniation of hypoplastic lungs (a tri-lobed right lung and one lobed left lung). Sternum was normal. Both the hands had post axial polydactyly. Lower limbs had pre-axial polydactyly with little toe loosely attached to the feet. Severe kyphoscoliosis was there but no evidence of neural tube defect was seen.

Umbilical cord was 3 cm in length with one artery and one vein. Due to its short length, placenta was attached directly to the peritoneal membrane covering the eviscerated organs. On separation placenta weighed 490gms. Baby died within 2 minutes of birth. No other investigations or autopsy were done as family member did not give consent for that.

DISCUSSION

Body stalk anomaly is a severe defect of the abdominal wall in which there is evisceration of abdominal organs and in more severe cases of thoracic organs as well. It is sporadic and rare; prevalence ranges from 0.4 to 3.2 per 100,000 live births and stillbirths [3]. There is no sex predilection and the risk of recurrence in future pregnancy is negligible. Abuse of cocaine, cigarette, alcohol or marijuhana may increase the incidence of this anomaly [4, 5].

According to Colpaert *et al.*; body stalk anomaly may be classified into two types: type I, complicated by craniofacial defects, and type II, complicated by ventral wall defects without craniofacial defects.[6] Three major theories have been proposed to explain its' origin. The first one is an early rupture of the amnion before there is an obliteration of the coelom with passage of lower half of the body into the coelomic cavity [7]. Van Allen *et al.*; proposed that a vascular compromise in 4-6 wks of gestational age leads to failure of closure of abdominal wall as well as persistence of extra-embryonic coelomic cavity [8]. Most accepted theory proposed by Steeter *et al.*; suggests an abnormal folding of the trilaminar disc in its cephalic, caudal, and lateral directions that might lead to the persistence of the coelomic cavity [9].

Prenatal diagnosis of body stalk anomaly is usually done in second trimester of pregnancy with ultrasound examination and elevated alpha-fetoprotein in maternal serum [10]. In ultrasound, internal organs are demonstrated in the extra embryonic coelom and a short or absent umbilical cord is seen [11]. Other pathologies that affect the abdominal wall such as omphalocele, gastroschisis, exstrophy bladder, Cantrell pentalogy, amniotic band syndrome and the OEIS complex (omphalocele, exstrophy of cloaca,

imperforate anus, and spinal defects) should be excluded to reach the final diagnosis [12].

There are no successful therapeutic interventions available to fetuses or newborns [13]. As it is a lethal anomaly, to avoid the complication of the mother during pregnancy or childbirth, abortion is recommended when the diagnosis is fully established [14].



Fig A & B: showing female baby with evisceration of thoracic and abdominal contents through defect in the body wall along with limb abnormality. Placenta is attached directly with herniated organs

CONCLUSION

It is important to distinguish a body stalk anomaly from other types of anterior abdominal wall defects prenatally because it is a fatal syndrome and early diagnosis and termination of pregnancy is required.

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