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Case Report

Antenatal diagnosis of omphalocele and spinal dysraphism: a possible OEIS complex

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ABSTRACT

OEIS complex, which comprises of omphalocele, exstrophy bladder, imperforate anus and spinal dysraphism, is a rare disorder. The prognosis depends upon the severity of the structural defects. Survival depends on the extension of the cloacal exstrophy and the neural tube defect. The antenatal diagnosis in early pregnancy is very tough, and a high level of suspicion should be kept for the possibility of its existence, even only if omphalocele and spinal dysraphism are seen on a prenatal sonography. Authors present the case of a 33-year-old, G1P0 female, who on her prenatal routine sonography scan, was found to have a fetus with omphalocele and spinal dysraphism. Despite the repeated attempts to follow her up, she refused and hence the possibility of the presence of other associated malformations could not be ascertained. Considering the difficulty of antenatal diagnosis of OEIS complex and its associated mortality, the objective of this case report is to discuss the vital sonographic findings of OEIS, along with its etiology and prognosis, which can aid in its early detection and subsequently help parents to make a decision regarding the continuation of pregnancy.

Keywords: Antenatal, OEIS complex, Omphalocele, Spinal dysraphism, Ultrasonography

INTRODUCTION

The OEIS complex is a rare disorder comprising a combination of defects-omphalocele, exstrophy bladder/cloaca, imperforate anus and spinal defects. Incidence is 1 in 200000-400000 live births with only a few reported cases of prenatal diagnosis.[1] The acronym was first used by Carey et al and apart from the findings indicated in the acronym, there have been descriptions of a variety of abnormalities, mainly in the lower region of the body and including defects of genitalia.[2]

CASE REPORT

A 33-year-old G1P0 female comes in for a routine prenatal ultrasound scan. On ultrasonography, abdominal contents were seen herniating into the amniotic cavity; Liver and bowel loops were seen within it and these were limited by a membrane (Figure 1).

Umbilical Cord was seen inserting into the midline of herniated contents (Figure 2). Widening of spinal canal at the lumbar level (Spinal dysraphism) was noted (Figure 3) along with kyphoscoliosis. The size of the defect appeared to be large. Great vessels from the base of heart were seen entering into the herniated abdominal contents (Figure 4).

Lower limbs and genitalia could not be discerned because of the obscuration by large omphalocele. Only one femur could be seen. The patient refused to come for a follow up and hence, the possibility of more malformations

could not be ascertained and a definite diagnosis of OEIS complex could not be made.

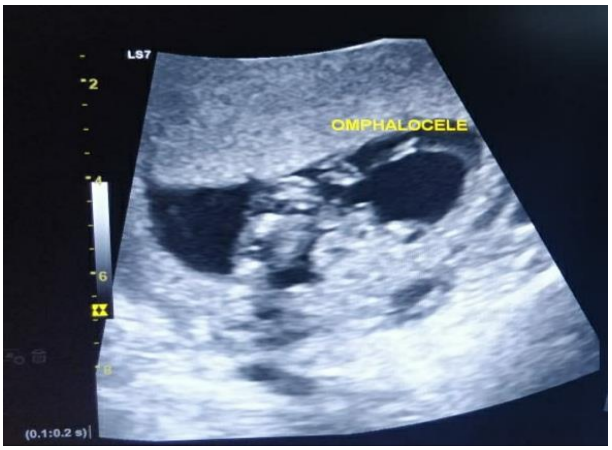

Figure1: On ultrasonography, abdominal contents were seen herniating into the amniotic cavity; Liver and bowel loops were seen within it and were these limited by a membrane.

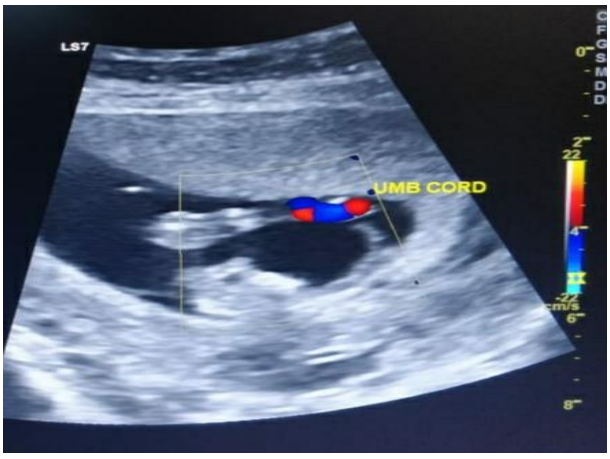

Figure 2: Umbilical cord was seen inserting into the midline of herniated contents.

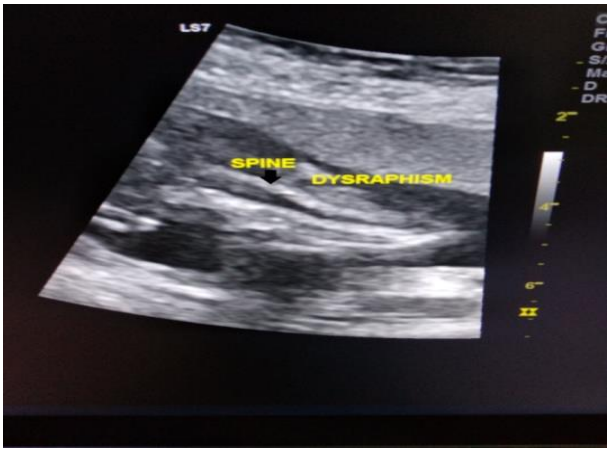

Figure 3: Widening of spinal canal at the lumbar level (Spinal dysraphism) was noted.

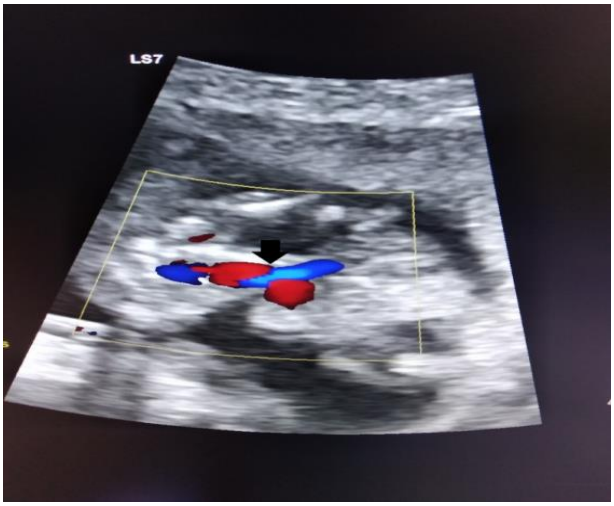

Figure 4: Great vessels from the base of heart were seen entering into the herniated abdominal contents.

DISCUSSION

OEIS complex represents the most severe end of a spectrum of birth defects, the exstrophy-epispadias sequence, which, in order of increasing severity, included phallic separation with epispadias, pubic diastasis, exstrophy of the bladder, cloacal exstrophyand OEIS complex.[3]

It results from defective blastogenesis in 4th week of gestation, resulting in improper closure of anterior abdominal wall and defective development of cloaca and urogenital septum.[4]

Common skeletal defects in spine include hemivertebrae, absent or hypoplastic sacrum. Neurological abnormalities may include tethered cord, lipomielocistocele, myelomeningocele, Arnold chiari, hydrocephalus, among others.[5] Kyphoscoliosis may also be seen (as noted in our case).[6]

Limb anomalies may be secondary to the defects of the spinal cord. Arthrogryposis, talipes, syndactyly, and thumb hypoplasia have all been documented in patients with this condition.[7]

The etiology of OEIS complex is thought to be multifactorial. It may be related to teratogen exposure in the early stage of pregnancy. Although most cases occur sporadically, a case with family history and recurrences in siblings has been reported. OEIS has also been identified in the fetus of a twin gestation.[7]

OEIS complex has been reported associated with trisomy 21, trisomy 18, triple X syndrome, turner mosaicism and 1p36 deletion. However, given the relatively high prevalence of these chromosomal disorders in the general population, it is unlikely that OEIS complex is associated with these chromosomal aberrations.[8]

In the 8 mm stage, the primitive streak mesoderm invades the cloacal membrane to form the lower abdominal wall. During the 8mm-16mm stage, the urorectal septum grows to divide a single cloacal chamber into the anterior urogenital system and the posterior alimentary system.

In cloacal exstrophy, the persistent cloacal membrane produces a wedge effect that impedes the migration of the mesoderm streak to form the anterior abdominal wall. The persistent cloacal membrane is thought to be unstable and will rupture.

If it ruptures before the division of the cloacal chamber by the urogenital septum, cloacal exstrophy occurs. If it ruptures after the formation of urogenital septum, the result will be exstrophy of the bladder. The coexistence of cloacal exstrophy and spinal dysraphism may be explained by a single defect in the embryonic tail in early pregnancy.[7]

The prognosis of infants with OEIS complex is variable, depending on the severity of structural defects. Survival will depend on the extension of the cloacal exstrophy and the neural tube defect. In less severe forms, good outcome with corrective surgery is possible.[9]

CONCLUSION

Prenatal diagnosis of OEIS complex is difficult and mainly relies on sonographic findings. The major findings include an absent bladder, persistent cloaca, omphalocele, and myelomeningocele. Diagnosis of OEIS complex in early pregnancy is extremely difficult. The findings may only be omphalocele and spinal defects (as seen in our case). The coexistence of these 2 defects should prompt the consideration of this disorder.

Cloacal exstrophy is lethal due to obstruction of the urinary tract. Hence, early prenatal diagnosis of OEIS complex is required to give parents the option to terminate the pregnancy. In cases which parents decide to continue the pregnancy, serial scans to see the progression of ventriculomegaly and an eventual C section to avoid dystocia and trauma should be done.

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