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Case History

A neonate with Omphalocele-Extrophy-Imperforate anus-Spinal defects (OEIS) complex

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Introduction

Omphalocele-Extrophy-Imperforate anus-Spinal defects Complex **(**OEIS complex) consists of multiple midline defects including, cloacal exstrophy, imperforate anus, omphalocele and spinal defects[1]. It occurs as a result of defective closure of anterior abdominal wall and mal-development of cloaca and urogenital septum due to defect in blastogenesis. It is estimated to affect 1: 200,000 to 400,000 pregnancies[1].Even though it has been reported in siblings[2], its occurrence is believed to be sporadic[3]. The prognosis is variable[3].

Case Report

We report a neonate with OEIS complex with additional defects. The baby was born at term to healthy non consanguineous parents as the second baby. At birth, she was found to have minor omphalocoele, cloacal exstrophy and high lying imperforate anus (figure 1). There were no dysmorphic features. Interestingly, the defects had not been detected in antenatal scans.

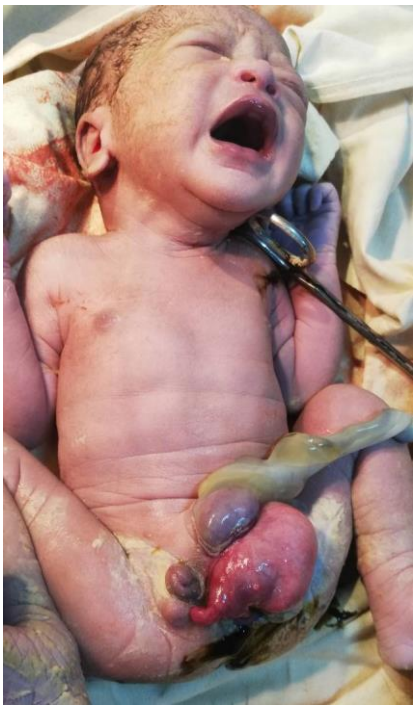

Figure 1

In addition, there were rib deformities (figure 2) and ultrasound abdomen revealed single right side kidney. Renal functions were normal. Karyotype was 46, XX. Furthermore, baby had ostium secundum ASD. Ultrasound brain was normal. She underwent the first stage of repair around 6 weeks of age, but needs to undergo reconstructive surgery in multiple stages in the future.

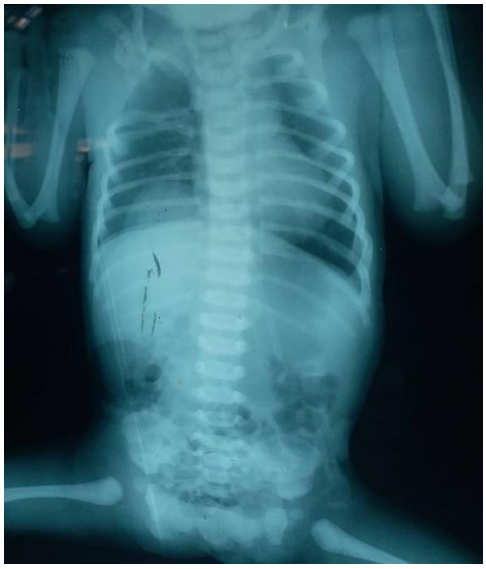

Figure 2

Discussion:

Abnormalities of almost every body system have been described in OEIS complex[4]. Gastrointestinal involvement is invariable[4].Similarly, in almost all babies, Bladder exstrophy is recognized. Renal anomalies include, ureteropelvic junction obstruction, renal agenesis, rudimentary kidney and pelvic kidney[5]. Among the reported skeletal anomalies, majority had pubis diastasis. Other spinal abnormalities include extra vertebrae, absent vertebrae, hemivertebrae and tethered cords. Limb anomalies, which are likely to be secondary to spinal defects are reported as well[6].

Correction is occurs in stages, which involves closure of omphalocele, separation of ileocecal connection from the exstrophied bladder plate and reapproximating the end colostomy with preservation of all bowels. Even though survival has been improved greatly due to advancement of surgical techniques, prognosis is variable which depends on the magnitude of the associated anomalies[3]. However, long term complications like bladder and bowel incontinence can result in reduced quality of life[3].

Antenatal diagnosis is challenging, but possible as early as 16 weeks of gestation. OEIS complex must be suspected when there is a combination of, abdominal wall defect, spinal defect and a non-visualized bladder in the antenatal ultrasound[4].

Conclusion

OEIS is a complex disorder with variable prognosis. Multidisciplinary approach is required to deliver optimum patient care.

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