

LETTER TO EDITOR (VIEWERS CHOICE)

## A RARE CASE OF OMPHALOCELE, EXSTROPHY OF BLADDER, IMPERFORATE ANUS AND SPINAL DEFECT COMPLEX WITH GENITAL ANOMALIES IN A TERM NEONATE

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### KEYWORDS

Cloacal exstrophy, exstrophy of the bladder, omphalocele

### ARTICLE HISTORY

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A 22-year-old pregnant female presented at 39.1 weeks of gestation in third stage of labour with hand prolapse to our emergency department. She immediately underwent emergency lower segment caesarean section (LSCS). There was no significant antenatal history of exposure to teratogenic drugs or radiation, infection, diabetes mellitus or hypertension. There was no history of treatment for infertility. No antenatal registration or antenatal scan was done.

**Figure 1.** Exstrophy of bladder, omphalocele along with imperforate anus



**Figure 2.** Absent Genitalia



She delivered a live baby weighing 2.8 kg with head circumference of 34.5 cm, who had multiple congenital abnormalities. There was no history of consanguinity or similar malformations in previous deliveries. There was presence of omphalocele, exstrophy of bladder, imperforate anus, and genital anomalies (Figure 1 and 2) along with bilateral congenital talipes equinovarus (CTEV) and lumbosacral meningocele.

X-ray thoracolumbar spine lateral view showed lumbosacral meningocele. Diagnosis of OEIS complex was made. Ultrasound (USG) of the abdomen and pelvis confirmed OEIS but the gender could not be evaluated due to obscure genital organs. Chromosomal analysis could not be done as the parents financial status did not permit for any future work up and intervention. The neonate died on day 5 of life.

OEIS complex is a severe form of exstrophy-epispadias complex which results from improper closure of ventral abdominal wall due to failure of cephalocaudal and lateral folding with associated defects of cloaca and urorectal septum.<sup>1</sup> The term OEIS complex was proposed to describe findings by Carey et al based on a retrospective search of medical records of 175 infants and has been described in the literature and is regarded as an entity.<sup>2</sup> The incidence is one in 200,000 to 400,000 pregnancies.<sup>3</sup> OEIS complex

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results from developmental defects affecting the mesenchyme, which is required for the development of infraumbilical mesoderm, the urorectal septum and lumbosacral somites. Therefore, any abnormality in mesodermal migration leads to premature rupture of cloacal membrane leading to omphalocele and exstrophy of cloaca.<sup>3</sup> Abnormalities of urorectal septum led to persistent cloaca and imperforate anus.<sup>3</sup> Genital anomalies are due to the failure of fusion of genital tubercles and separation of pubic rami.<sup>3</sup> Association of OEIS with deletion of chromosome 1p36 has been reported and also with maternal exposure to diazepam, cigarette smoking, and maternal obesity.<sup>4</sup> We could not elicit such a history in our patient. Additional malformations reported include rib anomalies, abnormal ears, hydrocephaly, microcephaly, encephalocele, cardiac defects, trachea-oesophageal fistula, and oesophageal atresia.<sup>5</sup> Microcephaly with head circumference less than 10<sup>th</sup> percentile was noted in our patient. An early antenatal diagnosis and urgent surgical correction is crucial in the management. The mother of our patient was not registered for antenatal care at any hospital; neither any obstetrics scan was done throughout the pregnancy. She did not receive periconceptional folic acid supplementation which is recommended by World Health Organization (WHO) to prevent neural tube defects in fetus.

### **Compliance with Ethical Standards**

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Conflict of Interest: None

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