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OEIS SYNDROME IN DIZYGOTIC TWINS: A RARE CASE REPORT AND REVIEW OF LITERATURE

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A B S T R A C T

We are reporting a case of diamniotic dichorionic discordant twins pregnancy with one of the twin having OIES syndrome and second twin normal. Most of the cases of OIES are reported with monoamniotic and monochorionic pregnancy. Moreover ours was the case complicated by medical factors like impending eclamsia, uncontrolled gestational diabetes and subclinical hypothyroidism, that may help to further know the etiology of OIES better.

INTRODUCTION

OIES Syndrome is a rare disorder comprising of omphalocele, imperforate anus, cloacal exstrophy and spinal defects seen in 1 in 2-4 lakh pregnancies. It is the most severe form of the exstrophy- epispadias spectrum of birth defects.

Literature on the topic remains limited. Few cases and case series have been reported and the aetiology is still unknown.

Case Summary

35 yr old primigravida at 28 weeks conceived by IVF (dichorionic diamniotic discordant twins) presented at our centre with impending eclampsia (BP 220/140 mm Hg, urine albumen 3+) with gestational diabetes mellitus (HbA1C- 7.5) and hypothyroidism (TSH-3.92 mIU/ml) not on treatment for any of these. One fetus had multiple anomalies (meningomyelocele, hydronephrosis, omphalocele, dilated renal pelvis) and the other fetus was morphologically normal on ultrasound scan. She was admitted and managed with IV labetalol and magesium sulphate, and relevant investigations were carried out. An emergency caesarean section was arranged for impending eclampsia and fetal distress in the normal twin.

First twin was a male baby weighing 1.2 kg, grossly normal, resuscitated and transferred to NICU for prematurity and discharged after 35 days. The anomalous twin was only 300 grams and had multiple anomalies (figure 1) namely omphalocele, single umbilical artery, imperforate anus, bladder exstrophy and clubfoot. Paediatric examination revealed no other gross anomaly suggestive of any aneupoidy. Autopsy and karyotype was advised but refused by patient.

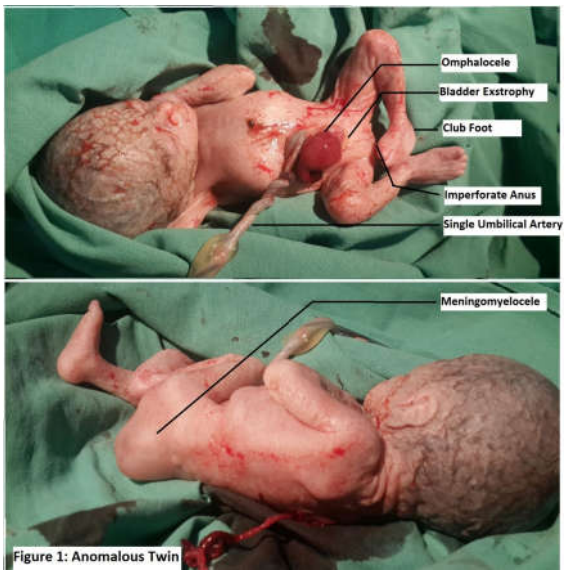

Figure 1: Anomalous Twin

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DISCUSSION

Fetal OEIS occurs in 1:250,000 live births (1). The occurrence of exstrophy of the bladder is more common (1:30 000 to 1:40 000) than exostrophy of cloaca (1:200.000 to 250,000) (6). It is more common in males and more commonly seen in monozygotic twins. Lee *et al* found 18 pairs of twins with OEIS syndrome of which 13 pairs were monozygotic, 1 dizygotic and 4 were unknown zygosity **(**2). Ours is a case of dizygotic twins. Omphalocele is reported more commonly than OEIS as at times all criteria of the syndrome are not met. OEIS complex results from improper closure of anterior abdominal wall due to failure of convergence of cephalocaudal wall and lateral folds of embryo during 4th week of development and; defects in development of cloaca and urogenital septum, non reduction of physiological hernia and non fixation of gut during 8-12 weeks (figure 2)(3).

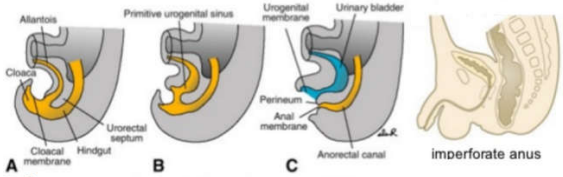

Figure 2: | Langman's Medical Embryology, 10th ed. 2006.

There is mesodermal in growth between ectodermal and endodermal layers of bilamellar cloacal membrane which results in formation of lower abdominal muscles and pelvic bone. This in growth separates bladder anteriorly and rectum posteriorly. The stage of development when the membrane rupture occurs determines whether there will be exstrophy of bladder, cloaca or epispadias. Etiologies and associations are summarized in the table 1.

Table 1 Etiology and Associations (5)

Genetic Basis	Developmental Problems	Acquired
Trisomy 18	Defective blastogenesis:	Sporadic familial-commonest
Single gene defects- HLBX9 mutation	i) failure of cloacal septation- common cloaca	Teratogens- diazepam, diphenyl hydantoin
Monozygotic Twins	ii) incomplete vertebral fusion- open neural tube defects	Uncontrolled diabetes during embryogenesis period
	iii) breakdown of cloacal membrane- exstrophy, omphalocele	
	iv) abnormal urorectal septum- fused urogenital septum and rectum	

Prenatal diagnosis requires identification of a) midline infraumbilical defect with inferior abdominal wall mass or cystic abdominal wall structure or omphalocele. b) absence of bladder between 2 umbilical arteries. c) lumbosacral meningomyelocele (almost always seen) or other spinal defects. d) Inferior limb anomalies. e) Single abdominal artery (frequently associated). f) Wide pubic arch. g) Genital anomalies making sex determination difficult. h) Anal atresia. i) omphalocele(4). Apart from these, there may be associated cardiac and renal anomalies or increased nuchal translucency. Austin *et al* gave criteria for prenatal diagnosis of OEIS. Major criteria were: 1. Non visualisation of fetal bladder, 2. infraumbilical abdominal wall defect, 3. omphalocele, 4. myelomeningocele; and minor criteria were i. lower limb malformation, ii. Renal anomalies, iii. ascites, iv. Wide pubic arch, v. narrow thorax, vi. hydrocephalus, vii. single umbilical artery. Out of these, 3 major (2, 3, 4) and 3 minor (i,ii,vii) criteria were present in our case. Most cases of OEIS are diagnosed prenatally or on autopsy.

Prognosis depends on severity of structural defects. Cloacal exstrophy is lethal due to urinary tract obstruction and associated renal anomalies, hence, early prenatal diagnosis is important for appropriate parental counselling and termination can be offered. However, if these pregnancies are continued, caesarean delivery is preferred to avoid dystocia and trauma. Neonate with OEIS requires care of paediatric surgeon, neonatologist, urologist, neurosurgeon and if available, anatomic pathologist. An early scan in the next pregnancy is the most valuable advice given to such patients.

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