Case Report

Oeis Complex: a rare Case Report

Sowmya M[1], Shwetha S[2]

[1]Dr Sowmya M, Consultant, Obstetric & Gynecology, [2]Dr Shwetha Shashidhar, Consultant, Obstetric & Gynecology, Both are affiliated with Sri Shivarathri Rajendra Hospital, Chamrajnagar, Karnataka, India.

Address for correspondence: Dr Sowmya M, Email: sowmyam2006@gmail.com

Abstract: We describe a case of OEIS complex [Omphalocele-Exstrophy of the Bladder-anal imperforation-spina bifida] a rare congenital malformation complex. It results from improper closure of ventral abdominal wall due to failure of convergence of cephalo-caudal and lateral foldings of embryo during development, with asociated defects in development of cloaca and urorectal septum. A second gravida, with previous normal obstetric history of 35 wks of gestation spontaneously delivered a live pre term baby with gross malformations of a large omphalocele containing kidney,liver and intestines, bladder exstrophy, anal imperforation and spina bifida. Patients with OEIS complex require the care of a multidisciplinary team with multiple surgeries with its own potential complications. Hence, early antenatal diagnosis and evaluation provides prognostic information, treatment options, decisions concerning the management and the plan for specific needs at birth.

Keywords: Omphalocele, Exstrophy of cloaca, Imperforate anus

Introduction

Carey et al reported a series of cases with an abnormality of body wall development and proposed the term OEIS complex [1].

OEIS complex (Omphalocele, Exstrophy of the cloaca, Imperforate anus, and Spine abnormalities) is a rare congenital syndrome with a reported incidence of 1 in 2-400000 live births [2].

There is no obvious etiology while, sporadic nature being the commonest in most of the reported cases. Besides the clinically recognised classic malformations, it can be variably associated with spina bifida, genital abnormalities, renal abnormalities, symphysis pubis diastasis, and limb abnormalities [3].

Here we report a Case of OEIS complex delivered in our institution along with the review of literature.

Case Report

29 year old second gravida of 35 wks of gestation presented to the OPD with labour pain. She was a manual labourer, with non consanguinous marriage. Her past obstetric history was normal and uneventful.

The mother had only one antenatal checkup in this pregnancy with no obstetric ultrasound. She spontaneously delivered a live pre term male baby of 2kg with multiple malformations-

Omphalocele containing liver, spleen and coils of intestine covered by thin membrane;

Exstrophy of the bladder- both uretric orifices were visible;

Small bifid phallus with underdeveloped scrotal folds, testis was not palpable in scrotum;

Anal imperforation (Fig 1)

Spina Bifida (Fig 2).

The attendants were counseled for detailed examination, staged surgical procedure and genetic evaluation; the attendants did not consent and was discharged against medical advice.

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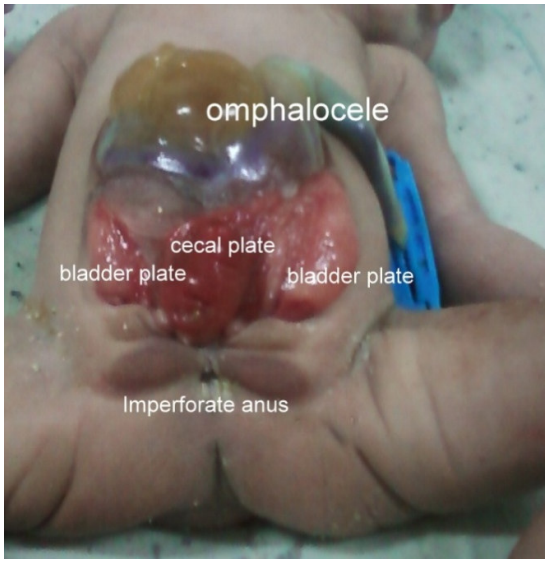

Fig 1: Omphalocele with lateral bladder plates, Cecal plate and imperforate anus

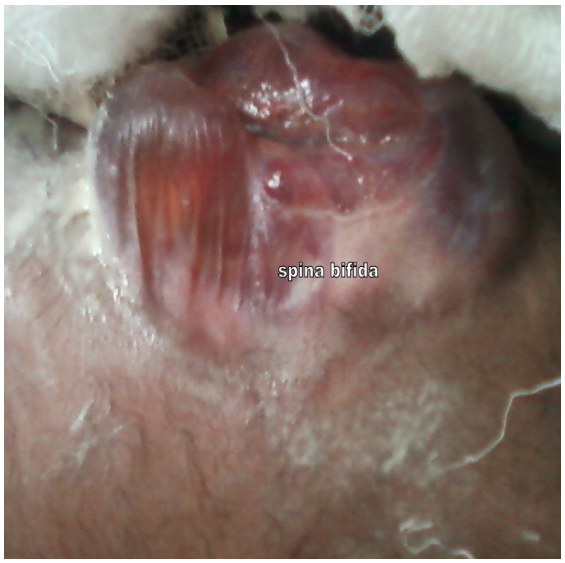

Fig 2: Spina Bifida

Discussion

OEIS complex represents a spectrum of congenital malformations ranging in severity from epispadias to bladder exstrophy to cloacal exstrophy [1]. Along with the classical features of OEIS complex, a strong association with spina bifida and intersex was also reported [4]. Cloacal exstrophy is considered the most severe and rare ventral abdominal wall defect among the four components of the OEIS complex. It is due to defects of caudal mesodermal migration at around 4 weeks of gestation [5]. Simultaneous failure of convergence of cranial, caudal and two lateral body folds results in the defective ventral abdominal wall. Rupture of cloacal membrane before urorectal septum descent results in exposure of exstrophied cloacal membrane. This cloacal exstrophy prevents the development of proctodeum manifesting as anorectal anomalies [6]. This caudal dysgenesis also affects the sacral vertebral development [7]. Spine malformations can occur more cranially than the normal lumbosacral level [4].

Cloacal exstrophy has also been reported in patients with chromosomal deletion [8], trisomy 18[1], twinning and in vitro fertilization[9]. Uteroplacental vascular insufficiency has a role in pathogenesis of clocal exstrophy [10].

Kundal et al reported a classical case of cloacal exstrophy consisting of an infra-umbilical omphalocele superiorly, lateral hemi-bladder plates, ureteric openings inferiorly near trigone and central caecal plate. Ileum was prolapsed from the central caecal plate manifesting as elephant trunk deformity[11]. Significant overlap between OEIS and limb body wall complex is also reported [12].

Diagnosis can be made by Ultrasound as early as 16 week of gestation [13]. Low-set umbilicus, ventral wall defect, spinal defect, wide pubic ramus and a non-visualized bladder with or without limb defects, are characteristic of OEIS complex. Lower abdominal mass, genital abnormalities can also be diagnostic, in addition to the absent bladder filling [14,15].

After initial stabilization of the baby, exposed organs and mucosal surface should be kept moist and non adherent dressing or sterile plastic wrap should be covered.

If the gender is not made out, Karyotyping can be done. Management is usually done by multispecialty team in a tertiary centre.

Conclusion

There is increase in survival rate in OEIS complex cases due to improved neonatal care and advanced surgical treatment and thus the quality of life.

However, early antenatal diagnosis of this condition will help in reducing the incidence of highly lethal condition.

Further prenatal diagnostic studies can be done to see for associated anomalies which helps in prognosis prediction. This case is reported due to its rarity and also for parental counselling about future conceptions.

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