IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 14, Issue 6 Ver. VIII (Jun. 2015), PP 59-61
www.iosrjournals.org

Giant Omphalocele with OEIS Complex - A Case Report

Nirmalya Saha[1], Chirom Pritam Kumar Singh[2], Moirangthem Matum Singh[3], Ruma Sarkar.[4]

[1]Assistant Professor, Department of Anatomy, Tripura Medical College & Dr. B. R. A. M. Teaching Hospital, Tripura, India. [2]Associate Professor, Department of Obstretrics & Gynaecology, Regional Institute of Medical Sciences, Imphal, Manipur, India. [3]Professor, Department of Anatomy, Regional Institute of Medical Sciences, Imphal, Manipur, India. [4]Senior Resident, Department of Radiodiagnosis, Jawharlal Nehru Institute of Medical Sciences, Imphal, Manipur, India.

Abstract: *An omphalocele represents an embryological defect of the umbilical ring and medial segments of the two lateral abdominal folds during fetal growth and can be divided into two groups depending on the size of the hernial defect. A major or giant omphalocoele is classified as a 5 cm or larger defect. The incidence is nearly 2.5 cases per 10,000 live births. The hernial sac may contain small and large bowel, stomach, liver, spleen, urinary bladder, gonads. A combination of defects including omphalocele, exostrophy of cloaca, imperforate anus, spinal defects is known as OEIS complex, affecting 1 in 2,00,000 to 4,00,000 pregnancies. During routine dissection in the Department of Anatomy, RIMS, Imphal, a 26 weeks gestational aged foetus of giant omphalocele with OEIS complex in was observed, with short umbilical cord, adherent to membrane was found with right sided scoliosis, imperforated anus with no external genitalias. On removal of the membrane all the abdominal viscera were malpositioned, malformed, malrotated and were found outside the abdominal cavity including the heart and both the lungs. Serial ultrasonography can be used to accurately diagnose abdominal wall defect in utero for monitoring fetal growth.*

Key Words: *Abdominal Wall Defect, Omphalocele, Umbilical Cord.*

I. Introduction

An omphalocele is an embryological defect of the umbilical ring and medial segments of the two lateral abdominal folds during foetal growth.[1] It represents when portions of the midgut fails to return to the abdominal cavity from its physiological umbilical herniation during 6^{th} to 10^{th} weeks. The incidence is nearly 2.5 cases per 10,000 live births.[2] An omphalocele can be divided into two groups depending on the size of the hernial defect. A major or giant omphalocoele is classified as a 5 cm or larger defect and a minor omphalocele occurs when the defect is 4 cm or less.[3] The hernial sac may contain small and large bowel, stomach, liver, spleen, urinary bladder, gonads and the defect is covered by amnion as umbilical cord is covered by its reflection.[2] OEIS complex is a combination of defects comprising of omphalocele, exostrophy of cloaca, imperforate anus, and spinal defect.[4] Our case belongs to giant omphalocele with OEIS complex.

II. Materials & Method

The fetus was collected from Department of Obstetrics & Gynaecology in RIMS Hospital during the routine dissection of the foetuses, in the Department of Anatomy, RIMS, Imphal. Due permission from concerned authorities and persons and Institutional Ethical Clearance were taken. Immediately after collection, gestational age (GA) was determined by Crown-Rump length (CRL) and maternal history. Weight of the fetus and length of umbilical cord (in centimeters) were measured. The fetus was immersed and fixed in 10% formalin for two weeks and then was dissected by removing the amnion or covering. The herniated intra abdominal organs were observed. Vertical and highest side to side dimensions of the herniated sac (in centimeters) had been measured to categorize the omphalocele either in minor or major groups. A major or giant omphalocele is classified as a 5 cm or larger defect. The thoracic cage with the organs was observed by opening through a midline incision extending below the chin to the upper limit of the herniated sac and extending towards both right and left mid axillary lines.

III. Observation

The foetus was of 26 weeks of gestational age (GA) with a weight of 1150 grams. The hernia sac was covered by a thin membrane extending on the abdominal region (fig 1), covering the organs. Short umbilical cord (14.5 cm) with one umbilical artery and one umbilical vein (fig 2), was firmly adherent to membrane on the right side towards foetal side but towards the maternal side the umbilical cord was centrally placed on placenta.

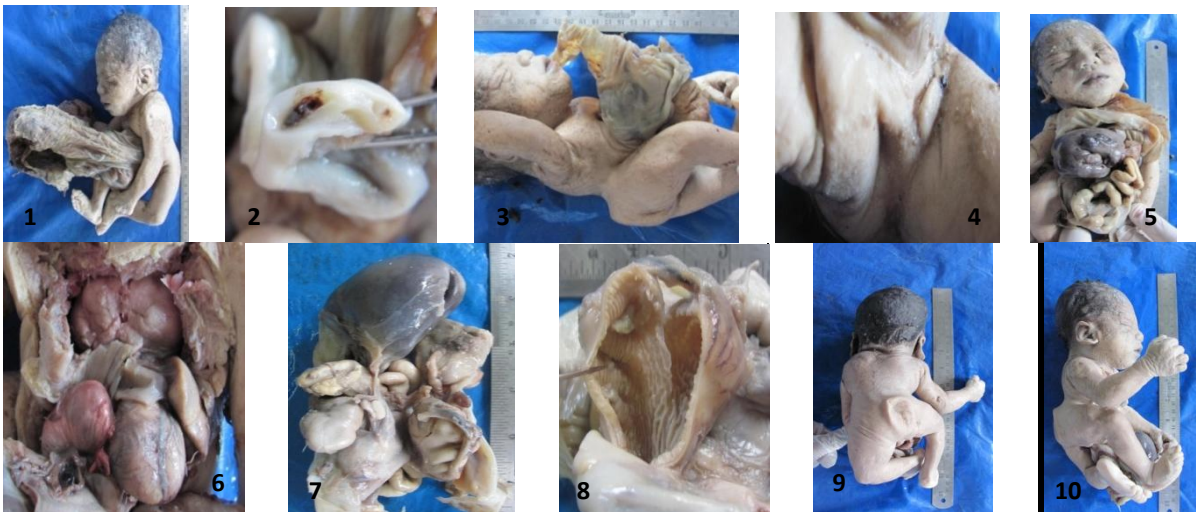

Figure (fig): 1. The hernia sac, covered by a thin membrane. 2. The umbilical cord with one umbilical artery and one umbilical vein. 3. Right sided scoliosis. 4. Imperforate anus with no external genitalia. 5. Anterior abdominal wall with defect. 6. Small abdominal cavity with visceras outside the abdominal cavity including the heart and both the lungs. 7. The malformed intra abdominal and pelvic organs. 8. Bladder and cloacal exostrophy. 9. A fluid filled sac was found on the right lumbo-sacral region. 10. Short lower limb with deformity in pelvic girdle and inverted right foot.

Right sided scoliosis (fig 3), imperforate anus with no external genitalia (fig 4) were observed. On removal of the membrane, whole of the anterior abdominal wall was with the defect (fig 5). The vertical dimension of the herniated sac was 7.5 cm and highest side to side dimension was 6 cm. The abdominal cavity was small and all the visceras were outside the abdominal cavity including the heart and both the lungs (fig 6). Malrotated small intestines were found anteriorly with malrotated and malpositioned dilated large intestine, observed posterior to the liver and small intestine. The malformed liver, malpositioned stomach, both malrotated kidneys with ureters, hydronephrotic left kidney (fig 7), bladder and cloacal exostrophy (fig 8) with undifferentiated gonads (fig 7) were observed.

Short lower limb with deformity in pelvic girdle and inverted right foot were observed. A fluid filled sac was found on the right lumbo-sacral region, which on dissection found to have vertebral arch defect (fig 9, 10). Short neck with no facial abnormality was detected (fig 5). Small thoracic cavity with whitish translucent membranous diaphragm was observed. Thoracic cavity was filled up with the thymus gland only. Heart and both the lungs were not observed in thoracic cavity, rather found inside the hernia sac below the diaphragm. Atrial and ventricular walls & cavities were normal. Both the lungs were hypoplastic and found by the side of the heart. The right lung was triangular in shape, reddish in colour and left lung was whitish in colour. Both the lungs were solid in consistency (fig 6).

IV. Discussion

An omphalocele results from the failure of lateral body folds to migrate normally toward the midline and consists of the midline defect in the anterior abdominal wall through which abdominal viscera, covered by a thin membrane herniated into the base of the umbilical cord and produces a defect at the umbilical ring.[5] Omphalocele can be subdivided according to the site of defect into epigastric (classic omphalocele) with cephalic fold defect, central type with a lateral wall defect greater than 4 cm and the hypogastric/caudal type with caudal fold defect.[6] In our case we found that the omphalocele was extended from epigastric region to the hypogastric region.

Several authors have reported that, the presence of viscera other than bowel in an abdominal wall defect is more likely with omphalocele. The presence of liver outside the foetal abdomen is a strong evidence for an omphalocele. Omphalocele containing liver and spleen are more likely associated with the abnormal karyotype with subsequent poor fetal outcome while containing only bowel loops are less likely associated.[7] Ectopia cordis is frequently associated with omphalocele, and it results from abnormal formation of the cephalic body fold as well as the lateral body folds.[5,8]

The OEIS complex is rare and affects 1 in 2,00,000 to 4,00,000 pregnancies and is of unknown cause.[9] Features of OEIS and associated malformation may have a genetic basis as recurrence found in siblings.[10] The occurrence of exstrophy of the bladder appears to be more frequent at 1:30000 to 40000 births[11] than exostrophy of the cloaca at 1:200000 to 250000.[12]

In giant omphalocele, the abdominal cavity is usually small and underdeveloped due to the absence of intestinal viscera in the abdominal cavity to stimulate growth.[13, 14] Adequate intra abdominal pressure may be necessary for the thoracic cage to develop, and with displacement of the liver and other abdominal viscera in case of omphalocele, the intra abdominal pressure decreases, potentially altering development of the fetal thoracic cage.[15] Pulmonary hypoplasia may be a primary defect or may result from restricted lung expansion due to a small, narrow thoracic cage.[16] Abnormalities in thoracic and abdominal muscle development may also contribute to the small, narrow thorax. Underdeveloped abdominal musculature related to the defect may cause scoliosis and a secondary thoracic deformity. Low intra abdominal pressure secondary to the displaced abdominal viscera may modify diaphragm mobility and function, resulting in pulmonary hypoplasia and thoracic deformity.[15]

V. Conclusion

The survival rate of foetuses with abdominal wall defect is less. Ultrasonography can be used to accurately diagnose abdominal wall defect in utero. In case of pregnancy continuation, serial ultrasonography is recommended to detect any alteration in fetal growth.

References

[1] D'Andrea F, Brongo S, Grella E, Grella R, Nicoletti G. Hepatic omphalocele in an adult: a case report. Scand J Plast Reconstr Surg Hand Surg 2004;38:236-9.
[2] Sadler TW. Langman's medical embryology. 11th ed. New Delhi: Wolters Kluwer / Lippincott Williams & Wilkins; 2010.
[3] van Eijck FC, Hoogeveen YL, van Weel C, Rieu PN, Winjen RM. Minor and giant omphalocele: long term outcomes and quality of life. J Pediatr Surg 2009;44:1355-9.
[4] Carey JC, Greenbaum B, Hall BD. The OEIS complex (omphalocele, exstrophy, imperforate anus, spinal defects). Birth Defects 1978;14:253-63.
[5] Klein MD, Heatzler JH. Congenital defects of the abdominal wall. Surg Gynecol Obstet 1981;152:805-8.
[6] Grosfeld JL, Weber TR. Congenital abdominal wall defects: gastroschisis and omphalocele. Curr Probl Surg 1982;19:157-213.
[7] Hasan Y, Malkawi MD Hussein S, Qublam MD Ahmad. Omphalocele containing bowel, liver and spleen: a case report. JRMS June 2005;12(1):35-7.
[8] Carpenter MW, Curci MR, Dibbins AW, Haddow JE. Perinatal management of ventral wall defects. Obstet Gynecol 1984;64:646-51.
[9] Hurwitz RS, Manzoni GAM, Ransley PG, Stephens FD. Cloacal exstrophy: a report of 34 cases. J Urol 1987;138:1060-4.
[10] Smith NM, Chambers HM, Furness ME, Haan EA. The OEIS complex (omphalocele-exstrophy-imperforate anus-spinal defects): recurrence in sibs. J Med Gener 1982;29:730-2.
[11] Higgins CC. Exstrophy of the bladder: report of 158 cases. Am Surg 1962;28:99-102.
[12] Redman JF, Seibert JJ, Page BC. Cloacal exstrophy in identical twins. Urology 1981;17:73-4.
[13] Dillon PW, Cilley RE. Newborn surgical emergencies:Gastrointestinal anomalies, abdominal wall defects. Pediatric Clinics of North America 1993;15(3):245-51.
[14] Dykes EH.Prenatal diagnosis and management of abdominal wall defects. Seminars in Pediatric Surgery 1996;5(2):90-4.
[15] Biard J, et al. Prenatally diagnosed giant omphaloceles: Short- and long-term outcomes. Prenatal Diagnosis 2004;24(6):434-9.
[16] Hershenson MB, et al. Respiratory insufficiency in newborns with abdominal wall defects. Journal of Pediatric Surgery 1985;20(4):348-53.