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A Rare Case of Omphalocele (Exomphalos) of Intestine and Liver

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Abstract: Omphalocele is one of the most common congenital abdominal wall defects. It is normally associated with other anomalies and isolated cases of omphalocele are rare. It is frequently detected prenatally due to routine maternal serum screening and fetal ultrasound. We report a case of congenital anomalies in a female newborn infant, born to the mother from a hill tribe. The baby presented exomphalos of intestine and liver with mild hydrocephalus. Its right lower limb was normal and the left lower limb was very short.
Keywords: congenital anomalies, exomphalos, abdominal wall defects.

INTRODUCTION

Omphalocele (exomphalos) is one of the most common congenital abdominal wall defects along with gastroschisis. It is reported to occur in 2.5/10,000 births and is associated with a high rate of mortality (25%) and severe malformations, such as cardiac anomalies (50%) and neural tube defects (40%)[1]. Its isolated cases are not frequently reported. It is frequently detected prenatally by routine maternal serum screening and fetal ultrasound [2]. We report a rare isolated case of exomphalos in a newborn female.

CASE REPORT

At Department of Obstetrics and Gynecology, Mookambika Institute of Medical Sciences, Kulasekaram, Tamil Nadu, India, a female baby was born with a rare anterior abdominal wall defect. The case was diagnosed prenatally. At the time of birth, the baby presented exomphalos of intestine and liver with mild hydrocephalus. Its right lower limb was normal and the left lower limb was very short. The baby was born to a mother who is from hill tribe [Fig-1]. The herniated mass contained major part of the intestine and whole of the liver.

Fig-1: Picture of the newborn showing the exomphalos small intestine and liver.

DISCUSSION

Omphalocele is considered as a common developmental anomaly of the abdominal wall. However, generally it is associated with other congenital abnormalities of heart, head or other regions. Isolated omphalocele as in this case is generally

regarded as an infrequent malformation with a very little risk of recurrence[3]. In some cases, when omphaloceles occur in a syndromal context, they are strongly correlated with various types of chromosomal anomalies [3].

It has been reported that only 60% of children with omphalocele survive until the end of the first year of age[4]. Exomphalos is very commonly reported to be associated with omphalomesenteric duct and urachal anomalies and a major group of these congenital disorders and are often associated with umbilical hernia[5]. A case of hernia of the umbilical cord with the whole of the liver herniating through the small defect into the hernial sac as the only content of the hernia has also been reported.[6] Shakya et al. have reported a case of omphalocele with dextrocardia[7]. A study conducted, to evaluate that in long run, whether the liver and other solid organs reach their normal position, shape, and size, in patients born with a giant omphalocele and underwent treatment for it, showed that the liver was in abnormal position in all patients and in majority of them, an incisional hernia was also observed. It was also observed that in some patients the spleen and the kidneys did not migrate to their normal position [8]. With all these reports, it is evident that omphalocele is a major and complicated congenital anomaly.

Prenatal diagnosis of omphalocele is vital as it may influence timing, mode and location of delivery. Prognosis for omphalocele depends mainly on the number and severity of associated anomalies. The surgical management consists of closure of the abdominal wall defect, ensuring minimal risk of injury to the abdominal viscera either through direct trauma or due to increased intra-abdominal pressure. Other options of management of an omphalocele include primary closure or a variety of staged approaches [2].

CONCLUSION

Isolated occurrence of omphalocele of liver and intestine is rare. Its prenatal diagnosis is vital in effective management of the case with minimal risk.

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