

Answers to Medical Quiz

A1. There is a large abdominal wall defect with covering layer at the umbilical area.

A2. Cardiac defect, chromosomal anomalies and Beckwith-Wiedemann syndrome.

A3. Omphalocele.

DISCUSSION

Omphalocele or exomphalos is characterized by protrusion of some abdominal contents, such as small or large intestine, stomach and even liver through a defect in the abdominal wall¹.

This defect is covered by a membrane which protects the exteriorized abdominal organs. The umbilical cord of the fetus is inserted at the top of the membrane. The covering membrane consists of peritoneum on the inner surface, amnion on the outer surface and Wharton's jelly between the layers¹.

In that condition, the abdominal content does not return to the abdomen but remains attached to umbilical cord. It is probably due to a failure of the abdominal wall in-folding leading to some of the organs herniating through the abdominal wall^{2,3}.

The incidence of abdominal wall defects including gastroschisis (one of the abdominal wall defects) and omphalocele ranges from 0.4 to 3 per 10,000 births and 1.5 to 3 per 10,000 births respectively. Most cases of abdominal wall defects are sporadic, but familial and possibly genetically determined cases of gastroschisis and omphalocele have been documented⁴.

Patients with omphalocele have a very high incidence of associated anomalies, up to 50%-70%. Chromosomal anomalies, notably trisomy 13, 14, 15, 18, and 21 are present in up to 30% of cases. Cardiac defects are also common in 30% to 50% of cases. Multiple anomalies are frequent and may be clustered in syndromic patterns. One important pattern is the Beckwith-Wiedemann syndrome^{5,6}.

Abdominal wall defects are often diagnosed by prenatal ultrasound performed for routine screening and confirmed by clinical examination post-delivery⁶.

The initial management of newborns who have abdominal wall defects starts with resuscitation. After stabilization of the baby, screen for associated anomalies should be initiated as soon as possible before the definite surgical repair⁶.

Surgical repair of omphalocele consists of reduction of the herniated viscera into the abdominal cavity and closure of the fascia and skin to create a solid abdominal wall. This surgical repair can be performed as primary closure when the abdominal wall defect is small, and can be repaired as staged closure when the abdominal wall defect is large⁷.

The prognosis of patients with omphalocele depends largely on the associated anomalies and medical conditions⁷.

CONCLUSION

Omphalocele is a rare abdominal wall defect with variant sizes leading to herniation of abdominal viscera. Diagnosis of omphalocele is by ultrasound and physical examination. The management of omphalocele is surgical repair as primary closure of small defects and staged repair for large defects. The outcome of patients with omphalocele depends on the associated anomalies.

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Competing Interest: None.

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