Umbilical cord ulceration and jejunal atresia

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The association between umbilical cord ulceration and congenital intestinal atresia is being increasingly reported and carries a high mortality. We report on a case of jejunal atresia associated with massive fetal haemorrhage from an umbilical cord ulcer. Fetal distress noted on continuous fetal monitoring allowed for delivery by emergency caesarean section followed by appropriate neonatal resuscitation and intact survival. This and other reported cases highlight the need for identification of high-risk fetuses with congenital intestinal atresia. Close fetal monitoring during labour is imperative in order to improve outcomes.

Congenital intestinal atresia is not an uncommon anomaly, occurring in approximately 1 in 3 448 live births. Intestinal atresias have been known to be associated with a number of other congenital anomalies, including chromosomal abnormalities, cardiac defects and other intestinal anomalies. In addition, there have been an increasing number of reports of congenital intestinal atresia associated with fetal haemorrhage. This association signifies a high mortality and requires close fetal monitoring and appropriate neonatal resuscitation to improve outcomes. We report on a recent case of jejunal atresia associated with massive fetal haemorrhage in an academic hospital in Johannesburg, South Africa.

Case report

A 21-year-old woman (para 0, gravida 1) booked for antenatal care at a Johannesburg clinic at 28 weeks’ gestation. Her blood group was Rhesus positive and both HIV and syphilis serology were negative. A scan for fetal anomalies was not performed.

At 32 weeks’ gestation the woman presented to Charlotte Maxeke Johannesburg Academic Hospital with signs of preterm labour. A fetal ultrasound scan showed an amniotic fluid index of 19 (not suggestive of polyhydramnios). Continuous fetal monitoring showed prolonged decelerations and decreased variability in keeping with fetal distress. An emergency caesarean section was performed and a live female infant with a birth weight of 1 360 g was delivered. Apgar scores were 5, 6 and 7 at 1, 5 and 10 minutes, respectively. The infant required assisted ventilation at birth and started breathing spontaneously by 10 minutes. Aggressive fluid resuscitation was required after delivery and the infant was noted to be extremely pale. She was transfused O negative blood as an emergency. Formal blood results from birth showed a haemoglobin concentration of 5.5 g/dl, a white cell count of 42.4×10⁹/l (27.6×10⁹/l once corrected for normoblasts), and a platelet count of 178×10⁹/l. Close examination of the umbilical cord revealed a 1 cm linear ulcer approximately 1 - 2 cm from the fetal insertion of the umbilical cord, which was actively bleeding during resuscitation. An umbilical venous catheter was inserted and the affected area of the cord resected. Unfortunately the placenta and umbilical cord were not sent for histological examination.

The infant responded well to resuscitation. She was treated for hyaline membrane disease with surfactant administration and nasal continuous positive airway pressure, from which she weaned by day 2 of life. On day 2 of life she was noted to have bile-stained gastric aspirates. A plain abdominal radiograph revealed a ‘triple bubble’ sign in keeping with jejunal atresia. Laparotomy and surgical correction were performed on day 5 of life. Her recovery was uneventful.

Discussion

The association of umbilical cord ulceration with intestinal atresia was first reported in 3 infants by Bendon et al. in 1991. Since then, a further 17 cases have been reported (Table I). In addition, thinning of Wharton’s jelly in the absence of macroscopic ulceration of the umbilical cord has been reported.

The pathogenesis of this process is, however, poorly understood. Bendon et al. suggested three possible causes. Firstly, umbilical cord ulceration and intestinal atresia may share an ischaemic aetiology. This is consistent with the ischaemic aetiology of intestinal atresia first described by Louw and Barnard. Secondly, the two pathologies may result from primary epithelial dysgenesis similar to the association of epidermolyis bullosa with intestinal atresia. Thirdly, regurgitation of bile acids and gastric contents may lead to ulceration of the umbilical cord. The latter theory is supported by the finding of elevated bile acid concentrations in the amniotic fluid of affected fetuses and the absence of cases of cord ulceration in infants with atresias proximal to the ampulla of Vater.

Despite the hypotheses described above, much remains to be studied and understood about the pathophysiology of cord ulceration in the context of intestinal atresia.

The combination of umbilical cord ulceration and intestinal atresia is associated with a high mortality rate. Massive fetal haemorrhage appears to occur only with the increase in intra-uterine pressure that results from the onset of labour. It is therefore important to diagnose intestinal atresia antenatally, and ideally identify high-risk fetuses before the onset of labour. Strategies to identify fetuses at risk of cord ulceration are largely experimental, but could potentially include ultrasonographic quantification of Wharton’s jelly and measurement of the total bile acid concentration in the amniotic fluid. Both these strategies may help to identify the fetus at high risk, but neither is able to definitively diagnose umbilical cord ulceration in utero. For this reason close fetal monitoring during labour is essential. The ideal management strategy remains delivery of the affected fetus before the onset of massive and possibly fatal haemorrhage.

In conclusion, the association of umbilical cord ulceration with intestinal atresia is being increasingly reported and is associated with high morbidity and mortality. Further research into the pathophysiology of cord ulceration and definitive diagnostic strategies is needed, as appropriate and timely management can significantly improve outcomes.
References


