Congenital volvulus without associated malrotation

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Abstract

Introduction

Congenital volvulus is a rare condition with a high morbidity and mortality. It may present as a surgical emergency at birth with a distended, discoloured abdomen. This paper reports a case of congenital volvulus without associated malrotation.

Case Report

A preterm newborn was born with abdominal distension and required resuscitation. An emergency transfer to a surgical unit was organised but she rapidly deteriorated. Post-mortem examination showed a congenital volvulus without an associated malrotation.

Discussion

In recent years, antenatal diagnosis has become possible by ultrasound scanning of the bowel and its vessels. However, this is technically difficult and only possible during the third trimester scans. The birth can then be planned in a unit equipped to provide the necessary early surgical assessment. There is an increased incidence of spontaneous preterm delivery and a majority of pregnancies do not receive late scans; hence, the diagnosis is commonly made postnatally. Early consideration of congenital volvulus in the differential diagnosis is vital to allow timely surgical intervention.

Conclusion

This case stresses the importance of planned delivery if an antenatal diagnosis has been made. However, in those infants without an antenatal diagnosis, who are born with similar clinical findings, transfer for a surgical opinion is time-critical but may not always be possible.

Introdution

Congenital volvulus is a rare condition with a high mortality and morbidity. Antenatal diagnosis is becoming possible on the third trimester scan, thus allowing for a planned delivery in an appropriate tertiary surgical centre. However, if an antenatal diagnosis has not been made or if premature labour occurs, considering this diagnosis and planning for an urgent surgical opinion may not be without difficulties. This case highlights the presenting features and discusses the current literature surrounding this rare diagnosis.

Case Report

A preterm neonate was born with a distended, discoloured abdomen, and had recurrent pulmonary haemorrhages. Despite intensive care management, she rapidly deteriorated and post-mortem confirmed the diagnosis of a congenital volvulus without associated malrotation. While in this case it was not possible to stabilize her for transfer, this may not be the case in other newborns.

A 38-year-old Brazilian woman presented in the early stages of preterm labour with intact membranes. There was no family history to note and her relationship was non-consanguineous. Antenatal serology was unremarkable and her routine antenatal scans at 20 and 26 weeks had shown appropriate growth with no obvious abnormalities. Cardiotocograph monitoring showed a pathological trace and a decision was made for emergency caesarean section. A female infant was delivered at 32 + 3 weeks gestation, with an estimated weight of 2 kg.

At delivery, the infant cried and had a normal heart rate. However, her abdomen was distended and she required intubation to support her respiration. Despite intubation and supplementary oxygen, she remained cyanosed and was pale. A nasogastric tube was inserted and 2 ml of yellow serous fluid was removed. At 20 minutes of age, she had a large pulmonary haemorrhage and a capillary blood gas revealed a severe mixed acidosis with a pH of 6.7. Despite optimizing ventilation, the administration of surfactant, packed cells, fresh frozen plasma, cryoprecipitate and bicarbonate, her acidosis and cyanosis persisted. Non-invasive measurement suggested she was hypertensive with a mean blood pressure of 74 mmHg, but arterial access was not possible to verify this. Echocardiogram showed a structurally normal heart with poor contractility, so inotropic support with dobutamine was commenced.

On examination, she did not appear to be dysmorphic but was pale, poorly perfused and had on-going pulmonary haemorrhage. Her abdomen was distended with a blue discolouration of the skin. She passed normal meconium and a small volume of blood stained urine. Abdominal radiography showed a radio-opaque, featureless abdomen. At 2 hours of life, her haemoglobin was falling despite repeated transfusions and she remained severely acidotic with poor oxygenation. She was considered clinically too unstable to transfer as she was rapidly deteriorating despite maximum intensive care. After

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Competing interests: none declared. Conflict of interests: none declared.

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extensive discussions between her family, the local and tertiary neonatal teams as well as the neonatal transport team, a consensus decision to withdraw life-sustaining treatment was made. The care was redirected towards comfort care and she died in her father’s arms.

A coroner’s post-mortem revealed a small bowel intrauterine volvulus without associated malrotation or perforation. The small intestine twisted in a clockwise fashion around the mesentery (which was haemorrhagic, oedematous, and thickened). There was necrotic bowel which was 42 cm long, extending from 29 cm from the duodeno-jejunal flexure to 36 cm from the ileocaecal valve. It was distended, dark red, friable, and contained approximately 200 ml of dark, bloody mucus. No other abnormalities or malformations were noted.

Discussion
Intestinal volvulus is a condition where the intestine, which is suspended along the free margin of the mesentery, twists around the axis of the superior mesenteric artery. This leads to mesenteric obstruction of the affected section of the bowel and may result in necrosis. In most cases, volvulus is associated with intestinal malrotation. However, in some cases, volvulus occurs without malrotation and it is speculated that this may be resultant upon a defect in the mesentery. A few cases of intestinal volvulus with or without malrotation of the intestines in foetal life have been described. Congenital volvulus usually manifests at birth as a lifethreatening condition with high associated morbidity and mortality. The diagnosis is suggested by a tense dark discoloured abdomen, indicative of subcutaneous intraperitoneal haemorrhage (Cullen’s sign), with bloody diarrhoea.

Case reports suggest that this diagnosis should be considered with the combination of polyhydramnios, ascites, a static abdominal mass with dilated intestinal loops, foetal distress with decreasing foetal movements, and reduced variability of the cardiotocograph (non-specific findings). It has been postulated that the reduction in heart rate variability may be due to parasympathetic over activity from the volvulus or foetal pain. Congenital volvulus is commonly associated with preterm delivery, possibly due to foetal stress and activation of adrenal and hypothalamic hormones.

Volvulus is associated with abnormalities such as gastrochisis, exomphalos, intestinal atresia, annular pancreas, and meconium ileus/peritonitis. Congenital volvulus is complicated in approximately one quarter of cases by intestinal atresia, which is thought to be due to previous antenatal episodes of volvulus resulting in intestinal necrosis and subsequent fibrosis. Milder cases have been seen with the volvulus being present for a longer duration and the development of collateral circulations. The prognosis of patients who survive initial surgery depends on the remaining length of bowel post-resection, the level of the obstruction, the presence of bowel perforation, and any other associated abnormalities. However, delay in obtaining surgical assessment increases morbidity and mortality.

Congentinal volvulus is sometimes diagnosed on antenatal ultrasonography by direct visualisation of the bowel loop and vessels (known as a whirlpool-like configuration or snail sign). Doppler measurements of the mesenteric vessels can be used to predict postnatal prognosis and aid in delivery management. Prenatal diagnosis enables planned delivery in a surgical centre, surgical assessment +/- surgery soon after birth and counselling of families. However, since the sonographic abnormalities are not usually found until at least 27 weeks gestation, this limits diagnostic potential to the small number of babies who have later scans.

Conclusion
In this case, prenatal diagnosis was not made. Due to the infant’s abdominal pathology, transfer for urgent surgical review had been arranged. However, it was impossible to stabilize her condition to allow transfer and her intensive care was re-directed. This case stresses the importance of planned delivery, if an antenatal diagnosis has been made. However, in those infants without an antenatal diagnosis, who are born with similar clinical findings, transfer for a surgical opinion is time-critical, but may not always be possible.

Consent
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

References
6. Yoo SJ, Park KW, Cho SY, Sim JS, Hhan KS. Definitive diagnosis of intestinal volvulus...