Spontaneous Bowel Perforation after Exchange Transfusion

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Introduction

INTESTINAL PERFORATION in the newborn can follow mechanical obstruction (atresia, volvulus, Hirschsprung's disease, etc.), high acidity (peptic ulceration in a Meckel's diverticulum) or may be spontaneous (necrotising enterocolitis). The last mentioned, necrotising enterocolitis, is not uncommon, as evident by the numerous reports on it. Perforation of the bowel is the last stage in its pathological course. With the accumulation of clinical data from several neonatal units, predisposing and/or associated factors are being increasingly recognised. The relationship between spontaneous bowel perforation and exchange transfusion, however, is a relatively new experience. In this report, a case is described to illustrate this association, and to emphasise the importance of passage of blood per rectum as an early diagnostic indicator.

Case Report

A 3.59 kg male infant was born normally at term to a healthy secundigravida in December, 1972. The mother's blood group was O, Rh negative, and the baby's group, A, Rh positive. A previous pregnancy had resulted in a rhesus-positive infant without any clinical or haematological evidence of isoimmunisation. No anti-D globulin had been given to the mother because examination of postpartum maternal blood had not revealed any foetal red blood cells. During the present pregnancy, tests for Rh antibodies were negative on two occasions.

The infant's clinical state at birth was excellent, the one-minute Apgar score being 9. Cord blood results were: Direct Coomb's test positive; Hb 14.7 gm% and bilirubin 4 mg%. Rising serum bilirubin necessitated two exchange transfusions, 10 and 24 hours after birth, via an umbilical vein catheter. 14 hours after the second transfusion, at the age of 38 hours, the child developed tachypnoea and grunting. He passed fresh blood and mucus per rectum and was reluctant to feed. Slight abdominal distension was also noticed. Bowel sounds were normal and a chest X-ray showed no abnormalities. Abdominal X-rays showed dilatation of the small bowel and gas in the bowel wall (pneumatosis intestinalis). There was no air-fluid level to indicate obstruction, nor was there any free gas in the peritoneum. A diagnosis of necrotising enterocolitis was made.

Treatment and progress. The child was managed conservatively by intravenous drip, gastric suction and parenteral penicillin and kanamycin. He, however, continued to pass blood per rectum, developed further abdominal distension and gradually developed oedema of the lower chest wall, anterior abdominal wall, scrotum, penis and buttocks. Gastric aspirate became bilious and bowel sounds scanty, but repeated abdominal X-rays failed to show free gas in the peritoneum or air-fluid levels. The intramural gas was no longer seen in the follow-up films. Conservative management was continued because there was no evidence of intestinal obstruction. On the sixth day of life, the child's condition deteriorated rapidly. Gastric aspirate was initially brownish but soon became frankly haemorrhagic. Bowel sounds were not heard and there were clinical signs of a left pleural effusion which was confirmed radiologically. There was haematological evidence of disseminated intravascular coagulation — pro-

thrombin 39%, platelets, 59,000 per ul. and fibrinogen 195 mg%. Intravenous hepairn was therefore commenced. The patient, however, succumbed after a respiratory arrest on the following day.

Autopsy. The whole ileum and much of the colon were infarcted and friable and there was a small perforation in one of the infarcted loops of ileum. Histologically, there was autolysis of the mucosal layer with haemorrhagic and neutrophilic infiltrates. There was generalised peritonitis, the umbilical vein was partially thrombosed and infected, and a peripheral hepatic artery was thrombosed, but the inferior vena cava, its major tributaries and the portal veins were free from thrombosis.

Discussion

Neonatal hyperbilirubinaemia is a common medical emergency in paediatric units, and exchange transfusion through the umbilical vein is an established method of treatment. Complications of exchange transfusion such as infection, portal thrombosis and cardiovascular disturbances are well-known. On the other hand, spontaneous bowel perforation (necrotising enterocolitis) has only been recognised as a complication of exchange transfusion since 1965[5]. Exchange transfusion was suggested as an aetiological factor after it was found to be the one important common denominator in mature infants (as compared to premature newborns) who developed spontaneous bowel perforation.[10]

The clinical presentation is typical, and the diagnosis easy if one is aware of the entity. Patients are usually well at birth. Shortly after the exchange transfusion, the early signs appear, heralded by passage of blood and mucus per rectum, refusal to feed and slight abdominal distension with gastric retention of bilious material; there may be grunting, tachypnoea and lethargy. Later, the abdominal distension becomes more marked and bilious vomiting may occur. Oedema of the anterior abdominal wall follows, due to umbilical sepsis and cellutitis, inferior vena caval obstruction or portal pyaemia and thrombosis. Pallor, poor peripheral circulation and hypothermia are late signs. Bowel sounds become inaudible when peritonitis sets in. A tympanitic abdomen with loss of liver dullness indicates intestinal perforation. The clue to the diagnosis is the appearance of fresh blood per rectum after the exchange transfusion.

Radiologically, five signs have been described — (1) intestinal distension, (2) intramural gas or pneumatosis intestinalis, (3) intrahepatic portal vein gas, (4) pneumoperitoneum (erect or lateral decubitus film), and (5) toxic dilatation of the colon.[1] The abdominal X-rays in the present case initially showed intestinal distension and intraumral gas, but follow-up films showed just non-specific intestinal dilatation. Although a perforation was detected at autopsy, pneumoperitoneum was not demonstrated radiologically. In reviewing 311 cases of necrotising enterocolitis, Pochaczevsky and Kassner[10] found bowel perforation in 43%. Yet, abdominal X-rays had shown pneumoperitoneum in only 19% of the cases. Intramural gas is a constant diagnostic feature although it has been reported as an isolated finding in severe diarrhoea with carbohydrate intolerance.[3]

Management. Conservative management — intravenous fluids and gastric suction, penicillin and kanamycin or gentamycin, and correction of acidosis — is still the initial line of approach. Surgery — laparotomy and ileostomy, colostomy or end-to-end anastomosis — is indicated when there is definite evidence of bowel perforation or when the clinical course is downhill.[1] In our patient, there was no clinical or radiological evidence of perforation and the deterioration in general condition was so fast that the stress of surgery was considered too great to be tolerated. As it was, the gut was so friable at autopsy that it crumbled in the pathologist's hands. Fairly good results have been obtained with surgery when the general condition of the child is better.

Patients with bowel perforation after exchange transfusion seem to fare better than those with perforation not associated with transfusions.[4,5,10] The survival rate for perforation associated with transfusion alone has been reported at 75%.[10] That for spontaneous perforation of mixed aetiology has been quoted to be as low as 15%.[7] One reason for these contrasting results is that the group with the mixed aetiological factors includes a large number of premature infants who do not tolerate surgery well. The group with perforation following transfusions consists mainly of infants who weigh 2.25 kg or more, and they natrally tolerate the stresses of surgery better.

Pathology. Necrosis of the bowel wall is the essential finding, with haemorrhagic and neutrophilic infiltrates. Perforation is found in the majority of cases of so-called "necrotizing enterocolitis" at operation or autopsy, although the clinical and radiological picture may not suggest it, as in the present case.

Aetiology. Many causes have been postulated but none proved. The association with exchange transfusion has been well documented. Several aetiologic mechanisms have been suggested, all implicating the portal venous system. Mechanical

disturbance of the portal circulation occurs in every exchange transfusion although the exchange is supposed to take place with inferior vena caval blood. An umbilical vein catheter inserted to the recommended legnth of 5 to 7 cm has its tip well short of the junction of the umbilical and portal veins.[4] Blood exchanged involves blood from the inferior vena cava via the nearby ductus venosus and blood from the portal circulation. (Fig. 1). Great

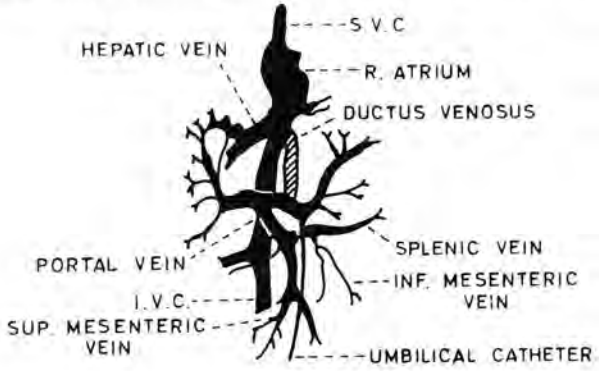

Fig. 1
The portal venous system, showing the relationship of the umbilical vein catheter and the ductus venosus.

pressure variations can therefore occur in the portal circulation during the procedure. Furthermore, the ductus venosus closes within a few days after birth,[6] and the umbilical vein catheter finds its way to the desired position in the inferior vena cava in only 20% of the procedures. In the other 80%, it is in a branch of the portal vein.[9] The sudden pressure variations can cause a vascular accident such as dislodging a small thrombus and causing retrograde embolism in the mesenteric vessels.[4] Reflex venospasm has been suggested to lead to infarction, but alone, it needs at least 18 hours to cause macroscopic infarction.[2] Temporary ischaemia has been postulated to predispose the bowel mucosa to enzymic autodigestion leading to ulceration and necrosis.[1] It has also been suggested that the pressure variations damage the portal microcirculation primarily,[10] causing haemorrhage and thrombosis in the capillaries which have an extremely small pressure difference between rupture and collapse.[8] Necrosis and perforation then follow.

That a vascular cause is responsible is supported by the constant finding of blood per rectum. This implies that a haemorrhagic infarct has occurred during the procedure. The haemorrhagic infiltrates found on histology also support this belief.

Summary

Necrotising enterocolitis complicating exchange transfusion in the newborn is a relatively new experience. Awareness of the entity and the recognition of the early sign, namely passage of blood per rectum, enables one to make a confident diagnosis, and this can be supported by the findings in the abdominal X-rays. Surgical correction is the treatment of choice in the presence of perforation. The procedure of exchange transfusion has been aetiologically implicated through the great pressure variations it causes in the portal circulation, and the damage is believed to occur in the portal microcirculation.

Preventive measures will include the use of a slow even pressure during the exchange transfusion to minimise the pressure variation, and the use of fresh catheters for every exchange transfusion to prevent thrombi formed at the catheter tip in an indwelling catheter from being dislodged and forced into the portal circulation. It is important that exchange transfusion be recognised as a skilled procedure, to be undertaken with all the proper precautions, as it can have fatal consequences.

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