SAJS
Case Report

Congenital hepatophrenic fusion: septum transversum-liver primordium anomaly

L. M. NTLHE, M.B. CH.B., F.C.S. (S.A.)
T. D. MOKOTEDI, M.B. CH.B., M.MED. (SURG.)
Department of Surgery, University of Limpopo (Medunsa Campus), Pretoria

Congenital fusion of the liver and diaphragm has not been reported in the literature. Surgery of the liver, e.g. in the case of trauma or transplantation, could be challenging in this situation.

A patient was admitted with blunt abdominal trauma and bowel perforation. Hepatic injury, especially grades III, IV and V, could have posed severe difficulties because mobilisation of the liver could have entailed extensive additional haemorrhage.[1-4] A midline thoraco-abdominal incision may be required in this situation for adequate access.

Case report

A 31-year-old man was admitted following an assault. He had sustained extensive bruises and blunt abdominal trauma with peritonitis. Investigations revealed a leucoyte count of 15 944/mm^3 and normal chest and abdominal radiographs. He was resuscitated with intravenous fluids and parenteral antibiotics in preparation for surgery.

At laparotomy the patient was found to have gross fibrinous peritonitis as a result of a mid-small bowel perforation. In addition, he had a completely fused right liver lobe and diaphragm (Fig. 1).

The bowel injury was repaired, the patient's postoperative course was uneventful, and he was discharged after 9 days.

Ultrasonographic screening during follow-up showed a normal diaphragm.

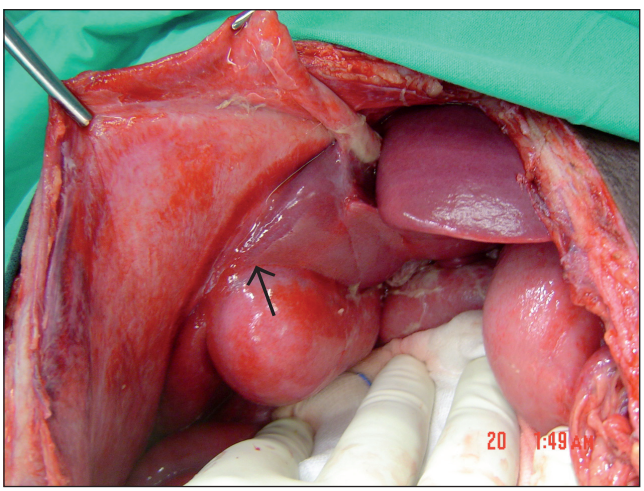

Fig. 1. Fused right liver lobe and right hemidiaphragm found at surgery (arrow).

Discussion

The congenital anomaly can probably be explained on an embryological basis. The liver primordium appears in the third week of gestation as a ventral endodermal outgrowth at the distal end of the foregut.[5-8]

With further development this diverticulum forms the pars hepatica and pars cystica. The pars cystica forms the gallbladder and cystic duct, while the pars hepatica proliferates and grows into the surrounding splanchnic mesoderm. Together they invade the dense somatic mesoderm known as the septum transversum.

The developing liver then bulges into the peritoneal cavity, with the mesoderm of the septum transversum forming its visceral peritoneum. On its cranial surface the septum transversum becomes the tendinous portion of the diaphragm; the liver surface is bare in this area. The adult liver remains suspended by the coronary ligaments.

The abdominal viscera in our patient, including the liver, were covered in places by a thin exudate of fibrinous peritonitis. This was easily removed with an abdominal swab revealing glistening visceral surfaces. We therefore concluded that this fusion could not have been a complication of a subphrenic abscess.

Three cases of ectopic right thoracic liver lobes have been reported. These cases had the supradiaphragmatic liver lobe extensions attached by a pedicle to the intra-abdominal liver and the authors stress that these were not secondary to herniation but congenital.[7]

We postulate that the liver primordium continued its intrauterine growth until it fused with the area of the diaphragm thus resulting in this anomalous union. Variations in fusion of hepatic planes within the liver have been reported.[8]

Our patient was counselled regarding his liver anomaly in order to equip surgeons with prior knowledge should he require surgical intervention in this area in future, most likely for repeat trauma. Others have expressed similar caution in patient preparation for liver transplantation.[8]

Conclusion

To the best of our knowledge this rare anomaly is the first reported case in the English literature. We propose the name 'congenital hepatophrenic fusion' and hypothesise that it results from the uninhibited growth and invasion of the septum transversum by the hepatic primordium.

REFERENCES

1. Trunkey DD. Hepatic trauma: contemporary management. *Surg Clin North Am* 2004; 84: 437-450.
2. Moore EE, Cogbill TH, Jurkovich GJ, Shackford SR, Malangoni MA, Champion HR. Organ injury scaling: spleen and liver (1994 revision). *J Trauma* 1995; **38**: 323-324.
3. Parks RW, Chrysos E, Diamond T. Management of liver trauma. *Br J* Surg 1999; 86: 1121-1135.
4. Skandalakis JE, Skandalakis LJ, Skandalis PN, Mirila PS. Hepatic surgical anatomy. *Surg Clin North Am* 2004; 84: 413-435.
5. Sadler TW. *Langman`s Medical Embryology*. 7th ed. Baltimore: Williams & Wilkins, 1995: 176-182, 254-255.
6. Delatte J-F, Avisse C, Flament J-B. Anatomic basis of hepatic surgery. *Surg Clin North Am* 2000; 80: 345-361.
7. Gray SW, Skandalakis JE, Akin JP. Embryological basis for the treatment of congenital defects. In: Gray SW, Skandalakis JE, eds. *Embryology for Surgeons*. Atlanta, GA: WB Saunders, 1972: 217-224.
8. Savier E, Taboury J, Lucidarme O, *et al*. Fusion of planes of the liver: an anatomic entity merging the midplane and the left intersectional plane. *J Am Coll Surg* 2005; 200: 711-719.

Letter to the Editor

Intestinal malrotation and midgut volvulus coexisting with intussusception

To the Editor: Intussusception and intestinal malrotation are common causes of intestinal obstruction in infants and children. The two conditions may coexist (Waugh's syndrome) but simultaneous occurrence with midgut volvulus is rare.

A 4-month-old boy presented with a 7-day history of vomiting, passage of bloody mucoid stools and a 3-day history of progressive abdominal distension, fever and difficulty breathing.

Physical examination revealed marked dehydration, pallor and a temperature of 38°C. The heart rate was 140/min and the respiratory rate 42/min. The abdomen was distended and tender but no mass was palpable. Bowel sounds were absent. The anal verge was soiled with bloody mucoid stools. A plain abdominal radiograph was not done as it would have delayed intervention. At laparotomy, the findings were ileocolic intussusception involving 20 cm of terminal ileum which was gangrenous, intestinal malrotation with duodenojejunal junction on the right of the midline, midgut volvulus of 180° in a clockwise direction, and the caecum and proximal third of transverse colon were gangrenous (measuring 25 cm). The volvulus was derotated and the intussuceptum reduced. The gangrenous terminal ileum and colon up to the proximal third of the transverse colon were resected. The mesentery of the small intestine was widened and an ileostomy and colonic mucus fistula were fashioned. Postoperative recovery was uneventful. Intestinal continuity was restored after 8 weeks and the child has remained well after 2 years.

Intestinal malrotation coexisting with intussusception (Waugh's syndrome) has been reported; one report noted that intestinal malrotation was present in 40% of patients with intussusception. However, simultaneous occurrence of Waugh's syndrome and midgut volvulus, as in the present case, is rarely reported. A Medline search up to mid-2006 revealed only 3 previously reported cases.

Since the current preferred treatment for uncomplicated intussusception in infants is non-operative pressure reduction, the possibility of Waugh's syndrome presents important implications for further evaluation; such infants may need to be evaluated for intestinal malrotation after successful pressure reduction. However, it has been suggested that absence of abdominal distension and paucity of distal bowel gas on plain abdominal radiographs may point to Waugh's syndrome. This was not observed in our patient.

C. S. Lukong
M. I. Maitama
B. Aminu
P. M. Mshelbwala
E. A. Ameh

Division of Paediatric Surgery
Department of Surgery
Ahmadu Bello University Teaching Hospital
Zaria
Nigeria