Torsion of the Spleen and Associated ‘Prune Belly Syndrome’

A Case Report and Review of the Literature

J. J. HEYDENRYCH, D. F. DU TOIT

SUMMARY

Splenic torsion as a complication of wandering spleen is rare. We report the clinical findings, diagnostic problems and treatment of a 1-year-old Coloured child (with classic ‘prune belly syndrome’) in whom the spleen had undergone torsion, thus simulating an intra-abdominal abscess. The postoperative course was uneventful.


In 1885 Prochownik first described torsion of the spleen. In 1933 Abell reviewed the world literature, collecting 95 case reports and adding 2 of his own. Since 1940 an additional 33 reports have been published, but no case of splenic torsion associated with ‘prune belly syndrome’ could be found in the world literature. ‘Prune belly syndrome’ is a syndrome in which the lower part of the rectus abdominis and the lower medial part of the oblique muscles are absent. This is usually associated with dilated bladder and ureters, small dysplastic kidneys and undescended testes.

Before 1944, only 2 of 1000 splenectomies performed in the Mayo Clinic were for visceroptosis of the spleen. In 1974 Carswell reviewed the world literature and found 350 reports of wandering spleen. He added 11 reports of patients treated during a 30-month period; 8 were children, but no case of ‘prune belly syndrome’ is mentioned in this review. Three of Carswell’s 11 patients presented with torsion of the spleen; the youngest patient with splenic torsion reported in the literature was 6 months old.

The purpose of this article is to report an apparently unique case of ‘prune belly syndrome’ in which the spleen had undergone torsion and simulated an intra-abdominal abscess.

CASE REPORT

A 1-year-old Coloured female child with classic signs of ‘prune belly syndrome’ was referred to our unit because of a visible mass in the abdomen. At the age of 1 month the baby had had bilateral cutaneous ureterostomies performed for severe hydronephrosis and mega-ureters. Because of the poor social background the child had been admitted to hospital on several previous occasions when she presented with pneumonia, gastro-enteritis, rashes and parasitic infestation of the gastro-intestinal tract.

On examination the child had loose stools associated with mild dehydration and a prominent red mass (5 x 10 cm) in the mid-abdomen, extending partly into the left upper quadrant. The respiratory, cardiovascular and central nervous systems were normal.

Abdominal examination revealed flabby redundancy of the abdominal wall, multiplicity of wrinkles (prune belly) and flank bulges. Because of the thin parietes, peristalsis and loops of bowel were clearly visible under the skin. The costal angle was wide and the sternum very prominent. The abdominal mass was adjacent to the left ureterostomy opening and the overlying skin was red. The mass was very tender on palpation, was not mobile and there was unmistakable rebound tenderness.

The bowel sounds were normal and no murmurs were audible over the mass or the rest of the abdomen.

Rectal examination confirmed the tenderness elicited on abdominal palpation. Mobility of the mass was limited in all directions and the edge was round and smooth, with no palpable notch. No blood or mucus was detected in the faeces. A radiograph of the abdomen showed that the proximal colon was distended, suggesting an obstructive lesion of the distal colon. A barium enema ruled out intussusception but demonstrated extrinsic pressure on the colon at the splenic flexure.

The stools contained Giardia lamblia, Ascaris ova, Klebsiella, and Candida albicans; the urine contained an abundance of pus cells. The haemoglobin level was 9.7 g/100 ml, and the white cell count 13 000/µl; there was neutrophilic leucocytosis and an erythrocyte sedimentation rate of 55 mm/h (Westergren). Red cell morphology, liver function tests and electrolyte levels were normal. The intravenous pyelogram demonstrated bilateral mega-ureters and hydronephrosis, but no displacement or distortion of the kidneys.

At laparotomy the mass was found to be the spleen which had undergone torsion. Numerous areas of gangrene were visible on its surface but no abscess was detected. Overt perisplenitis caused the flimsy anterior abdominal wall to stick to the surface of the spleen. The perisplenitis had initiated a corresponding inflammatory response of the wrinkled abdominal wall, thus simulating an intra-abdominal abscess. The splenorenal ligament was absent and the splenic pedicle was twisted through 360°. Splenectomy was performed and the child made an uneventful recovery, except for a persistent urinary tract infection.

Department of Paediatric Surgery, University of Stellenbosch and Tygerberg Hospital, Parowvallei, CP

J. J. HEYDENRYCH, B.SC., M.SC., M.B. CH.B., M.MED. (SURG.), Principal Paediatric Surgeon
D. F. DU TOIT, M.B. CH.B., F.C.S. (S.A.), F.R.C.S., Registrar

Date received: 11 October 1977.
Twenty-four hours postoperatively there was a distinct thrombocytosis (platelet count 1 000 000/μl), but 1 week later the thrombocyte count had fallen to 650 000/μl without treatment. Routine immunoglobulin analysis showed no derangement of the gamma, G, M or A fractions at the time of discharge from hospital.

**DISCUSSION**

**Diagnosis**

Torsion of the spleen is seldom diagnosed pre-operatively and the mass is often confused with ovarian masses, a mechanically obstructed bowel or other intraperitoneal swellings. To diagnose acute torsion a high index of suspicion is sometimes more helpful than sophisticated laboratory investigations. Carswell, in a report on 11 cases of which 6 were diagnosed pre-operatively, maintains that the diagnosis can be confidently made when the three signs described by Gindrey and Piquard are elicited. These are: (i) palpation of a firm, ovoid abdominal mass with a notched border; (ii) special mobility of the mass, especially when movement is painless towards the left upper quadrant but painful in any other direction; and (iii) resonance to percussion in the left upper quadrant.

Chronic or recurrent torsion of the spleen may give rise to splenomegaly or hypersplenism which, in turn, results in thrombocytopenia and leucocytopenia. The pre-operative diagnosis of splenic infarction may be established by the demonstration of leucocytosis, thrombocytosis, burr cells and Howell-Jolly bodies in the peripheral smear.

In the difficult case, selective angiography may help to confirm the diagnosis, because no splenic blush will be visible when the splenic artery is totally occluded. A radiograph of the patient in the upright position will disclose bowel only under the left diaphragm. A sophisticated and more helpful investigation is a technetium scan of the spleen, which will also determine the site of the ectopic spleen. Uptake of radioactive material by the spleen, which will also determine the site of the ectopic spleen. In the difficult case, selective angiography may help to confirm the diagnosis, because no splenic blush will be visible when the splenic artery is totally occluded. A radiograph of the patient in the upright position will disclose bowel only under the left diaphragm. A sophisticated and more helpful investigation is a technetium scan of the spleen, which will also determine the site of the ectopic spleen. Uptake of radioactive material by the spleen, which will also determine the site of the ectopic spleen.

The diagnosis must also be thought of when there is a history of fever, abdominal pain, vomiting and splenomegaly.

The spleen develops embryologically from the dorsal mesogastrium just above the tail of the pancreas. When fully developed, it lies along the 9th-11th ribs. The upper end is opposite T9, approximately 3,5 cm from the midline, and the lower pole extends as far forward as the mid-axillary line. Peritoneal folds which form during 'embryological migration' of the spleen to the left upper quadrant are responsible for fixation of the spleen in its normal position, but in some patients these attachments are fairly long and permit abnormal mobility or torsion.

Disturbances of the attachment mechanism may cause elongation of the splenic pedicle which gives rise to so-called 'displaced', 'floating', 'ectopic' or 'wandering' spleen, with or without torsion of the pedicle.

Most authors agree that wandering spleens, with or without torsion, are rarely encountered in clinical practice. Carswell's experience in this respect is unique, as 'wandering spleens' constituted the second commonest cause for splenectomy at Mulago Hospital. Eight of the 11 patients were children and in 6 a correct diagnosis was made pre-operatively. This high degree of accuracy is probably related to the relative frequency of this condition in Uganda.

In the present case the infarcted spleen simulated an intra-abdominal mass. The thin parietes became adherent to the splenic capsule and the splenic notch faced posteriorly, so that none of the signs imputed to wandering spleen were applicable. The fact that the mass was situated adjacent to the ureterostomy opening strongly suggested an abscess secondary to previous surgery or leakage of urine into the peritoneal cavity. Weinreb points out that acute torsion of the spleen can mimic appendicitis, obstruction of bowel, peritonitis, acute cholecystitis or hypersplenism. Shende et al published a case report in which the diagnosis of splenic infarction was made pre-operatively because burr cells and Howell-Jolly bodies were demonstrated in the red blood cells. There were moderate neutrophilic leucocytosis, a raised erythrocyte sedimentation rate (55 mm/h) and a haemoglobin level of 9,7 g/100 ml, but red cell morphology was normal in our patient.

Most of the cases of wandering spleen reported by Carswell occurred in children, in 2 of whom a persistent mesentery was noted on the descending colon; in another the caecum was abnormally mobile. Solanke reported excessive caecum mobility in over 60% of cadavers (adults and children) of West Africans, as opposed to 2% of Whites. The high incidence of wandering spleen in children lends credeence to the theory that visceroptosis of the spleen is caused by congenital weakness of the attachment mechanism. In the adult, acquired conditions, such as multiparity, trauma, malarial splenomegaly and splenomegaly associated with splenic tumours, may cause visceroptosis and torsion. The findings in our patient endorse the view of congenital weakness, because in addition to the abdominal wall maldevelopment the splenorenal and phrenosplenic ligaments hardly existed, being represented by some abnormally long flimsy strands of fibrous tissue.

The main complication of wandering spleen is torsion of the splenic pedicle. The pedicle may twist many times, resulting in infarction of the spleen, fever, abdominal tenderness, rigidity of the anterior abdominal wall, shock, degenerative cyst formation, or, in severe cases, gangrene of the spleen. Traction of the spleen on the pancreas may result in recurrent attacks of acute pancreatitis with shock, as demonstrated in one of our previous cases.

**Treatment**

We believe surgery is the treatment of choice in any case of wandering spleen, because torsion is a major complication. However, this view is not endorsed by Shende et al, who believe that removal of an asymptomatic wandering spleen is unnecessary because torsion is rare. The mortality of splenic torsion is 20% when it occurs in pregnancy, and of Abell's 97 patients 60 required emergency surgery. Collins cites 2 cases of wander-
ing spleen in which treatment was conservative. One patient required emergency splenectomy later and the second developed chronic torsion and hypersplenism which necessitated surgery. Splenectomy was performed in all but one of Carswell's cases. In the remaining patient a 'reefing' operation was performed because the author believed that the spleen has a protective function in malaria-stricken areas. He does not explain why the reefing operation was contraindicated in the other cases, but draws attention to the fact that in 2% of cases splenectomy may predispose to serious infection in children under 4 years of age.

The increased mortality which follows detorsion and replacement of the spleen has convinced most authors that surgical treatment should be undertaken as soon as the diagnosis of wandering spleen is established. Our previous experience and the findings in the present case prompted us to add our strong support to this view.

We should like to thank the Superintendent of Tygerberg Hospital for permission to publish, and Mrs C. Hunter for the typing.

REFERENCES


Inguinal Hernia Containing a Paramesonephric (Müllerian) Duct in an Adult Male

A Case Report

G. J. KANE

SUMMARY

This article describes the extremely rare contents of a right indirect inguinal hernial sac in an adult male, namely a paramesonephric duct. Radiological investigation at operation by means of 'salpingography' demonstrated the connection of this 'uterus' with the prostatic utricle. 'Salpingo-hysterectomy' was performed. Sex typing of the patient determined him to be a male. His family history revealed siblings with intersex features; the prenatal determination of sexual identity is discussed and reference made to the fate of the mesonephric (Wolffian) and paramesonephric (Müllerian) remnant.


The persistence of a large, uterus-like paramesonephric duct in a male is in itself clinically unusual, but when it also forms part of the contents of a hernial sac it must be considered a rarity.

CASE REPORT

A 24-year-old White man was admitted to Addington Hospital, Durban, on 24 October 1962 with a diagnosis of a right indirect inguinal hernia and right undescended testis which had been present since birth. In addition to the fact that the right side of the scrotum was not developed and the right testis was impalpable, the patient had glandular hypospadias with chordee. The left testis was present in the scrotum and there was no gynaecomastia.

With the patient under general anaesthesia a right inguinal incision was made and a large indirect (oblique) hernial sac was found. The hernial sac was invaginated on its lateral aspect by the spermatic cord excluding the