Rare disease

Right abdominal mass in a 2 year-old child

Constantinos A Parisinos,¹ Ibrahim Matter,² Jeorge G Mogilner,³ Mark A Rodrigues,⁴ Nadav Slijper³¹Department of Internal Medicine, Royal Free Hospital, London, UK;²Department of General Surgery, Bnai Zion Hospital, Haifa, Israel;³Department of Paediatric Surgery, Bnai Zion Hospital, Haifa, Israel;⁴Department of Radiology, Edinburgh Royal Infirmary, Edinburgh, UK**Correspondence to** Dr Constantinos A Parisinos, c.a.parisinos@gmail.com**Summary**

The authors report a case of a wandering spleen presenting as a right lower quadrant abdominal mass, 2 years post a transabdominal left diaphragmatic hernia repair in a 2-year-old child with a congenital diaphragmatic hernia. The wandering spleen was fixed laparoscopically in an extraperitoneal pouch.

BACKGROUND

Wandering spleen is a rare condition caused by laxity or absence of the splenic ligaments. The causes for this condition are not yet clear. Its non-specific signs and symptoms together with the rarity of this condition hamper the clinical diagnosis. Untreated, it can proceed to severe complications such as splenic torsion and necrosis, a surgical emergency. Early surgical intervention is the best guarantee for preserving the spleen. We hypothesise that a wandering spleen may be associated with a left congenital diaphragmatic hernia (CDH). This association has rarely been reported in the literature.

CASE PRESENTATION

A 2-year-old child was referred to the emergency department with a painless, right lower quadrant abdominal mass. Observations and blood tests including full blood count,

urea and electrolytes, liver function tests and C reactive protein were within normal limits. Examination revealed a palpable, non-tender, smooth right pelvic mass. Medical history included a left CDH (figure 1) that was repaired by laparotomy immediately after birth. The organs that were reduced from the chest were the small and large bowel. Postoperative recovery was uneventful. There was no other relevant past or family history of note.

INVESTIGATIONS

Upon arrival, an abdominal ultrasound (AUSS) was requested, and a wandering spleen, with a patent vascular pedicle was found overlying the right kidney (figure 2).

TREATMENT

Following multidisciplinary discussions, the child underwent a laparoscopic splenopexy in a retroperitoneal pouch.

Figure 1 Chest x-ray before (A) and after (B) laparotomy and left CDH repair. Reduction of intrathoracic abdominal organs is clearly demonstrated.

Figure 2 Abdominal US (AUSS) before (A) and after (B) left splenopexy. (A) AUSS showing the spleen overlying the right kidney in the right iliac fossa. (B) Fixed spleen in the retroperitoneal pouch with patent vascular pedicle following laparoscopic splenopexy.

Owing to the previous operation, the slightly lower position of the diaphragm, and the adhesions in the subphrenic region, the retroperitoneal pouch was created slightly anterior and caudal to the standard location. The peritoneum was incised with a hook and the pouch created by blunt dissection and cautery. After placing the spleen in the pouch, the peritoneum was closed, leaving a window for the vascular pedicle, using Vicryl sutures.

OUTCOME AND FOLLOW-UP

The child proceeded to an uneventful recovery and was discharged on the second postoperative day. During the following 2 months, two separate AUSS investigations demonstrated the spleen in the fixed position, with a patent vascular pedicle (figure 2).

DISCUSSION

Wandering spleen is a rare condition; the largest multicentre retrospective study identified just 14 cases of wandering spleen in France over a 25 year period (1984–2009).¹ Wandering spleen is caused by laxity or absence of the ligaments fixing the spleen in the splenic fossa.

The presentation of a wandering spleen may be completely incidental (15%), but in 90% an abdominal mass can be identified. It may be diagnosed during a routine physical examination (as in the present case), but usually presents with acute abdominal pain caused by torsion of the spleen (65%). Other presentations include abdominal mass, intermittent abdominal pain, bowel obstruction, dysuria, gastric volvulus, bleeding oesophageal varices and with a splenic abscess.¹ When torsion occurs an urgent operation is required to release the spleen and fixate it, if possible, or resect it if the spleen is already necrotic. In elective cases, a splenopexy is recommended, which can be performed either through a laparotomy or by laparoscopy. The fixation options include a retroperitoneal pouch, the use of a mesh or the creation of an omental pouch.

CDH is seen in approximately 1:2000 live births. The herniating organs are usually small bowel, stomach, colon and left lobe of the liver. Herniation of the spleen is fairly rare. The association of a wandering spleen and a left-sided diaphragmatic hernia was first described in 1940² and there have been a further 11 reports of this association published in the literature.^{1 3–11} The migration of the spleen above the diaphragm, or the herniation of adjacent organs during

the neonatal development, may contribute to the absence of its attachments.

A second operation, to fixate the spleen in the left upper abdomen, after a previous left diaphragmatic hernia repair, is expected to be difficult because of adhesions, and the flattening of the left diaphragm, compromising the space in the splenic fossa. Splenopexy in these cases may require the use of a mesh to hold the spleen in place, or the compromise of creating a lower extraperitoneal pouch. Hence, when a diaphragmatic hernia presents later in life with an associated wandering spleen a splenopexy should be performed in the same procedure as the hernia repair.³

We hypothesise that the initial insult affected the full development of the diaphragm and may be responsible for the absence of the fixating splenic ligaments. However, a wandering spleen is not seen in every child with CDH, and this may be partially due to adhesions formed after the CDH repair.

Learning points

- The diagnosis of wandering spleen is extremely difficult to establish because of its rarity and non-specific symptoms.
- An early diagnosis and surgical care are the best guarantee for preserving the spleen. A laparoscopic approach seems to be the treatment of choice.
- We believe that a wandering spleen should be considered in every case of a left CDH. Intervention is necessary, as severe complications such as splenic torsion are common and can have devastating effects.

Competing interests None.

Patient consent Obtained.

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