CASE REPORT

Acute torsion of a wandering spleen

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The 'wandering spleen' is a rare condition due to extreme laxity or absence of ligaments that fix the organ in its normal anatomical position within the left upper quadrant. Without early surgical intervention, wandering spleen can lead to torsion and subsequent splenic infarction or rupture. Clinical suspicion plus urgent investigation and intervention are important, so as to salvage the spleen and prevent complications. We present a case of torsion of a wandering spleen in a 21-year-old young woman, who presented with a painful pelvic mass. We also reviewed the literature on this entity.

Introduction

Several authors have described wandering spleen as a rare surgical condition encountered in the past. As implied by the term "wandering", the spleen may be found in unusual locations commonly the lower abdomen or pelvis (due to gravity), and present as a lower abdominal or pelvic mass. Very few case series have been reported, and clinical experience of this condition is rare. The patient may become symptomatic when torsion of the pedicle results in pain, ischaemia, infarction, or rupture.[1] The initial clinical diagnosis may be neglected if this rare entity is not kept in mind. Urgent ultrasonography (USG) could be a very useful initial tool, which reveals absence of the spleen in the left upper quadrant and an abdominal mass with USG features of a spleen.[2] Herein, we present a patient with torsion of a wandering spleen.

Case report

The patient was a 21-year-old Chinese woman with good past health. She presented to the Department of Surgery, Tseung Kwan O hospital on 30 January 2010 with a history

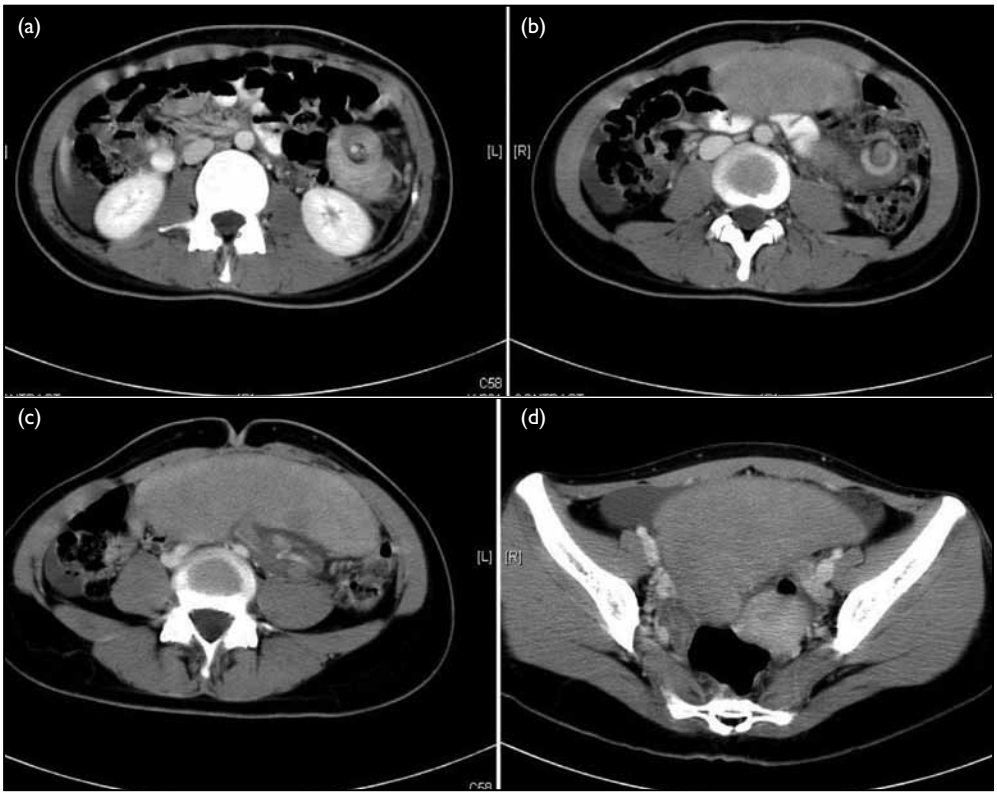

FIG 1. Contrast computed tomography (CT) of the abdomen and pelvis
(a, b) CT films showing 'whirl' sign from twisting of vascular pedicle, (c) vascular pedicle entering the spleen, and (d) a significant part of spleen located inside pelvis

Key words
Pelvis; Spleen; Splenic diseases; Tomography, X-ray computed; Torsion; Wandering spleen

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of epigastric pain for 2 days. The pain was colicky in nature without radiation. She had also noticed an abdominal mass for about 1 year. The size of the mass had remained static, but from time to time it became painful for few days. She had no history of trauma or injury.

On admission, she had a fever of 38.1°C. Her blood pressure was 106/63 mm Hg and the heart rate was 106 beats/min. A huge, tender left lower quadrant mass arising from pelvis was palpable. Rectal examination yielded nil abnormal and her pregnancy test was negative. Initial blood tests showed white cell count of 17.1 x 10^9 /L and haemoglobin level of 151 g/L, but routine liver and renal function test, and serum amylase level yielded nil abnormal. Initially, the patient was suspected to have an ovarian mass with a complication. Tumour markers (CEA, CA125, HCG, AFP, LDH) were checked but were all within normal limits. Urgent computed tomography (CT) of the abdomen and pelvis (with contrast) showed a large homogeneous soft tissue mass of size 20 cm x 7.5 cm x 15 cm in the lower abdomen and pelvis (Fig 1). The CT also showed the 'whirl sign' of twisting vascular tissue over the left side of the abdomen, and accompanying report noted marked twisting of the small bowel mesentery mimicking volvulus. There was also mild bilateral hydronephrosis, very likely due to an obstructive effect from the mass. The spleen was not visualised in the left upper abdomen.

An operation was jointly arranged with general surgeons and gynaecologists, in view of a preoperative provisional diagnosis of a small bowel volvulus and/or uterine tumour. Operative finding revealed a huge spleen in the pelvic area with torsion of the vascular pedicle starting at the tail of the pancreas (Fig 2). Other internal organs were normal. A total splenectomy was performed, as the organ appeared congested, it was likely infarcted, and not likely to be salvageable.

The patient recovered well after the operation, and triple vaccination (against pneumococcus, *Haemophilus influenzae*, and meningococcus) was given. Histology revealed congestive splenomegaly and torsion of a wandering spleen. The patient developed symptoms of small bowel obstruction on the eighth postoperative day, which resolved on conservative management.

Discussion

Wandering spleen is a rare entity accounting for less than 0.25% of splenectomies. Discussion in the literature is limited to isolated reports and small case series. The aetiology may be congenital or acquired. If congenital, the patient may be born with absence of attaching ligaments. There may be failure of the dorsal mesogastrium to fuse with the posterior abdominal wall during the second month of embryogenesis. This could result in extreme laxity or absence of normal supporting ligaments, namely the gastrosplenic, lienorenal, splenophrenic, splenocolic, splenopancreatic, presplenic fold, pancreaticocolic, and phrenocolic.

Ben Ely et al[3] reported a case in which two sisters both suffered from wandering spleen presenting 3 years apart. Since the disease is so rare and occurred in sisters, the authors suggested a familial association. However, to date there is only one such case report in the literature.

Acquired cases appear mainly associated with parity, which causes weakness of the abdominal wall and a laxity of ligaments normally attached to the spleen. It may also be secondary to splenomegaly, such as occurring in malaria, lymphoma, chronic myeloid leukaemia, and lymphosarcoma.[4]

Women are about 13-fold more prone to this entity than men, age at presentation may be 20 to 40 years, and most affected females are multiparous.[5] In the first year of life there is male predominance (2.5:1), and female predominance thereafter.[6] In paediatric patients, there may be other associated congenital diseases, including prune belly syndrome, renal

游走脾的急性扭轉

游走脾很罕見，主要因為支撐脾臟的韌帶過份鬆弛或缺少，以致不能把脾臟保持在腹腔左上方的正常解剖位置。如果未能及時施以手術，游走脾有機會扭轉，因而導致脾梗死或撕裂。要保留脾臟及防止併發，應對此症高度警惕並及時為病人檢查及作介入治療。本文報告一名患有游走脾的21歲女性，病發時出現盆腔腫塊。本文並會探討此症的文獻報告。

FIG 2. Clinical photo taken during the operation (the patient's head is on the right)
A huge congested spleen in pelvic region with torsion of the vascular pedicle starting at the tail of the pancreas

agenesis, gastric volvulus, diaphragmatic eventration and congenital diaphragmatic hernia.[6,7]

Patients may be asymptomatic, present with a pelvic mass[5] or intermittent colicky abdominal pain (presumably caused by intermittent torsion-detorsion or kinking of relevant vasculature). Other non-specific symptoms include nausea, emesis and mild crampy abdominal pain. The symptoms may change with body posture. One case report describes pain decrease on standing, possibly due to untwisting.[8] In two cases, pain was reported to diminish on adopting the left lateral position.[9]

Patients aged less than 12 months most commonly present with an abdominal mass.[6] In all other age-groups, acute abdominal pain due to a complication was the commonest presentation. Rare complications include obstruction of the gastric outlet, duodenum, small intestine, and colon, various types of intestinal volvulus, pancreatic pathology, splenic abscess and others.[2,6,7,10-12] Physical examination findings can vary depending on the location of the wandering spleen and complications. Abdominal USG (with or without Doppler) and CT are useful investigative tools.[3,5,13]

Concerning treatment, if the spleen is viable, open or laparoscopic splenopexy is preferred. The surgery adopted differed in different centres and at different times.[2,6,9,12,14] If not viable (due to torsion leading to infarction), splenectomy becomes necessary, with prophylactic antibiotics to cover the surgery. In paediatric cases, prophylactic antibiotics should be discontinued up to the age of 6 years. Vaccination against pneumococcus, *Haemophilus influenzae*, meningococcus should also be given.

Conclusion

Wandering spleen is a rare clinical condition. With a high index of suspicion, early investigations and surgical intervention, complications may be prevented and splenic function preserved.

References

1. Moran JC, Shah U, Sinder JA. Spontaneous rupture of a wandering spleen: case report and literature review. Curr Surg 2003;60:310-2.
2. Steinberg R, Karmazyn B, Dlugy E, et al. Clinical presentation of wandering spleen. J Pediatr Surg 2002;37:E30.
3. Ben Ely A, Seguier E, Lotan G, Strauss S, Gayer G. Familial wandering spleen: a first instance. J Pediatr Surg 2008;43:E23-5.
4. Waldman I, Suissa L. Lymphosarcoma in an ectopic pelvic spleen. Clin Nucl Med 1978;3:417-9.
5. Wallace S, Herer E, Kiraly J, Valikangas E, Rahmani R. A wandering spleen: unusual cause of a pelvic mass. Obstet Gynecol 2008;112:478-80.
6. Brown CV, Virgilio GR, Vazquez WD. Wandering spleen and its complications in children: a case series and review of the literature. J Pediatr Surg 2003;38:1676-9.
7. Uc A, Kao SC, Sanders KD, Lawrence J. Gastric volvulus and wandering spleen. Am J Gastroenterol 1998;93:1146-8.
8. Lien CH, Lee HC, Yeung CY, Chan WT, Wang NL. Acute torsion of wandering spleen: report of one case. Pediatr Neonatol 2009;50:177-80.
9. Schaarschmidt K, Lempe M, Kolberg-Schwerdt A, Schlesinger F, Hayek I, Jaeschke U. The technique of laparoscopic retroperitoneal splenopexy for symptomatic wandering spleen in childhood. J Pediatr Surg 2005;40:575-7.
10. Di Crosta I, Inserra A, Gil CP, Pisani M, Ponticelli A. Abdominal pain and wandering spleen in young children: the importance of an early diagnosis. J Pediatr Surg 2009;44:1446-9.
11. Nguyen KD, Dhawan M, Patel S, et al. Gastric variceal bleed due to a "wandering spleen". Am J Gastroenterol 2002;97:S150.
12. Balliu PR, Bregante J, Pérez-Velasco MC, et al. Splenic haemorrhage in a newborn at the first manifestation of wandering spleen syndrome. J Pediatr Surg 2004;39:240-2.
13. Raissaki M, Prassopoulos P, Daskalogiannaki M, Magkanas E, Gourtsoyiannis N. Acute abdomen due to torsion of wandering spleen: CT diagnosis. Eur Radiol 1998;8:1409-12.
14. Hirose R, Kitano S, Bando T, et al. Laparoscopic splenopexy for pediatric wandering spleen. J Pediatr Surg 1998;33:1571-3.