Wandering spleen with horseshoe kidney a rare occurrence

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A B S T R A C T

INTRODUCTION: Wandering spleen is a rare clinical entity in itself, with only 2 cases reported thus far when correlated with congenital under-development of the kidney, it usually happens due to under development of its surrounding ligaments. Herein we present a case of wandering spleen with underlying congenital deformity of horseshoe kidney which requires splenectomy due to late presentation.

PRESENTATION OF CASE: A 21 year old lady presented with worsening of chronic abdominal pain for 3 years, associated with nausea and vomiting. Physical examination showed a vague mass located at epigastric region. Consecutively, computed tomography images showed a well-defined, oval, hypoechoic spleen extending from center of abdomen up to epigastric region measuring 15.5 x 13 cm with twisted pedicle. Finally the patient underwent surgical treatment. The intraoperative findings were consistent with computed tomography images. The patient made a full recovery and was discharged well.

DISCUSSION: Wandering Spleen was first described by Van Horne during autopsy back in 1667. Its location is maintained by peritoneal attachments such as lienorenal, splenocolic, splenophrenic, gastrosplenic and phrenicocolic ligaments. Among which, the gastrosplenic ligament and lienorenal ligaments are of greatest significance. Patient with a wandering spleen may present asymptomatic, with a movable mass in the abdomen, or with chronic or intermittent abdominal pain because of partial torsion and spontaneous de-torsion of the spleen as in our case. When feasible especially in young patients, splenopexy should always be the first consideration but however if gross infarct has occurred then splenectomy is inevitable to save the patient.

CONCLUSION: Wandering spleen is a unique surgical entity moreover when appeared in congruence with horseshoe kidney. Its diagnosis should be made in prompt to prevent splenic infarction and to try to salvage with splenopexy especially in younger population. However in patient where splenic torsion with infarction has occurred, splenectomy would be the treatment of choice.

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1. Introduction

Wandering spleen with horseshoe kidney is a rare diagnosed clinical entity, with only 2 reported case thus far [1,21]. It is characterized by the absence or under-development of one or all the ligaments supporting the spleen in its normal position in the left upper quadrant of the abdomen, resulting in hypermobility. It commonly affects children with a predilection for female in the adult population [2]. Clinical presentation is variable, it usually takes the course of an acute torsion with subsequent infarct, which is potentially fatal if not treated promptly. Herein we report a rare case of wandering spleen with horseshoe kidney presenting with acute abdomen. This case report has been reported in line with the SCARE criteria [23,24].

2. Case report

We report the case of a 21 year old lady who presented to emergency department with abdominal pain, she complained of intermittent abdominal pain and early satiety, which persisted for the past 3 years worsening over the past week. She had been visiting the emergency department of various hospitals, but was always treated as dyspepsia and discharged on visit. There was no family history of thalassemia or hematological malignancy, with no history of trauma. She was otherwise in good health. On physical examination, abdomen was soft but there was a deep seated mass detected and located at the epigastric region with a size of 10 x 8 cm, showing firm consistency and non-mobile feature. Percussion over Traube’s space was resonance.

Results of laboratory date were as follows: neutrophils 18.1 10^9/L, white cell count 22.1 10^9/L, creatinine 52 µmol/L, sodium 139 mmol/L, potassium 4.3 mmol/L, urea 3.7 mmol/L, INR 1.6, hemoglobin 9.5 g/dL and platelets 333 10^9/L.

Radiological investigation started with an ultrasound, which revealed a well-defined, oval, hypoechoic solid mass extend-
ing from center of abdomen up to epigastric region measuring 15.5 × 13 cm. An abdominal CT was later obtained to further define the extent and nature of the mass, an empty splenic bed was noted with the mass being the spleen located at the center of the abdomen [Figs. 1 and 3]. The splenic artery and vein are twisted at the distal segments, giving the ‘whirl’ appearance. Horseshoe kidney with mild left hydronephrosis with no identifiable course of obstruction was noted [Fig. 2]. On the basis of the radiological findings and the intolerable pain, we decided to proceed with surgical intervention. Surgical access was obtained using a midline incision. A bluish discolored and congested spleen was identified with an absent lienorenal ligament. It was freely mobile with evidence of twist at its pedicle [Fig. 4], therefore surgical resection was performed. Histopathology shows, extensive hemorrhage of spleen parenchyma associated with congestion consistent with torsion of wandering spleen. Post-operative course was smooth and uneventful, patient discharged home after 3 days and made a full recovery thereafter.

*Below pictures are attached to figure and picture document

3. Discussion

This is a 3rd case reported on wandering spleen associated with a horseshoe kidney. Spleen is the largest organ in our reticulo-endothelial system [3]. Its location is maintained by peritoneal attachments such as lienorenal, splenocolic, splenophrenic,
gastrosplenic and phrenocolic ligaments. Among which, the gastrosplenic ligament and lienorenal ligaments are of greatest significance. Gastrosplenic ligament anchors the splenic hilum to the greater curvature of stomach, carrying within it the short gastric and left gastroepiploic vessels, while splenic vessels and tail of pancreas travels within the lienorenal ligament [4].

Wandering Spleen was first described by Van Horne during autopsy back in 1667 [20]. The Spleen is developed from mesodermal cells in the dorsal mesogastrium, during the fifth and sixth week of development, rotation of the dorsal mesogastrium gives rise to gastrosplenic ligament and lienorenal ligament, and thereafter the fusion of the dorsal mesogastrium. Congenital failure of development of these peritoneal attachments results in wandering spleen [5,6–9]. Embryologic basis of congenital laxity is incomplete fusion of dorsal mesogastrium with the peritoneum overlying the left kidney [9]. Acquired caused of laxity of these ligaments can be caused by multiparity, hormonal effects of pregnancy, previous abdominal trauma and splenomegaly [5,8].

Patient with a wandering spleen may present asymptomatic, with a movable mass in the abdomen, or with chronic or intermitent abdominal pain because of partial torsion and spontaneous de-torsion of the spleen as in our case [10]. Most common complication being torsion [11]. Examination may reveal a palpable abdominal mass in any abdominal quadrant but may be absent in 33% of the cases [12]. Clinically the diagnosis should be suspected when we feel a firm tender mobile mass with a notched border. However, it may be obscured in setting of splenic engorgement [13,14]. Preoperative diagnosis of wandering spleen is rarely suggested base on clinical findings alone because of non-specific symptoms. Multiple imaging modality has been used in establishing the diagnosis, which includes ultrasonography, plain radiograph, computed tomography, nuclear scintigraphy, magnetic resonance imaging and angiogram. Of which Duplex Doppler are useful in providing information regarding vascularity of the spleen, however it can be obscured bowel gas [14].

CT scan remains the investigation of choice, manifestation includes: (1) absence of the spleen anterior to the left kidney and posterior to the stomach, (2) a lower abdominal or pelvic mass with homogenous or heterogeneous splenic parenchyma and an attenuation value less than that of normal splenic tissue, (3) whorled appearance of splenic vessels and surrounding fat only, and (4) secondary findings such as ascites and necrosis of the pancreatic tail [15,19,22,23]. As per our case, whirling of the splenic vessels can be seen which is specific of torsion of a wandering spleen.

Treatment have changed over the years, generally splenopexy is favored over splenectomy especially in children and adult up to third decade of life due to the risk of overwhelming post-splenectomy infection (OPSI). However, in settings of spleen infarction, risk of rupture, or with splenic vein thrombosis, splenectomy is the treatment of choice [16–18]. Many surgical techniques are described for splenopexy including: (1) laparoscopic splenopexy. (2) Fixation of the capsule to the left upper quadrant (3) creation of a postero-lateral extra peritoneal pocket at the level of the 12th rib. (4) Mobilization of the splenic flexure with gastroplasty. (5) Use of a polyglycolic mesh bag and hemisplenectomy with fixation of residual spleen. However, splenectomy was per-

**Fig. 3.** Coronal view of spleen with its pedicle.
Fig. 4. Infarct spleen with torsion of pedicle.
formed in our patient in view of extensively torsed and completely infarcted spleen presenting as acute abdomen.

4. Conclusion

Wandering spleen is a unique surgical entity moreover when appeared in congruence with horseshoe kidney, its appearance may be due to embryological anomaly. It should always be considered as a possible cause of acute abdomen in a patient whose spleen is absent from its anatomical position. Its diagnosis should be made in prompt to prevent splenic infarction and to try to salvage with splenectomy especially in younger population. However in patient where splenic torsion with infarction has occurred, splenectomy would be the treatment of choice.

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Consent

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Author contribution

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