Torsion of wandering spleen in an infant associated with hamartomatous vascular malformation

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ABSTRACT
Wandering spleen, also known as ectopic spleen, is a rather infrequent entity with an estimated incidence of 0.2%. It is characterized by hypermobility of the spleen, which is caused by the laxity or lack of the ligaments that normally hold the spleen in place in the left hypochondrium. The clinical presentation can take various forms; it can be asymptomatic and inadvertently discovered during radio imaging for a different purpose, or it can present as an abdominal emergency. The course of management is determined by the vitality of the spleen. In view of this, we present the case of a 45-day-old patient who presented to our centre with evidence of intestinal obstruction; however, she was actually suffering from splenic torsion, and a splenectomy was performed.

1. Introduction
Ectopic spleen is quite rare; at present, only approximately 500 cases have been reported, and its estimated incidence is 0.2% [1]. It is 2.5 times more common in males than in females among those younger than 12 months [2]. Since 2003, a total of 44 paediatric cases have been reported worldwide in both the surgical and radiology literature [3]. It is characterized by hypermobility of the spleen caused by the laxity or lack of the ligaments that normally hold the spleen in place in the left hypochondrium [4,5]. Its clinical presentation can take various forms; it can be asymptomatic and inadvertently discovered during radio imaging for a different purpose, or it can present as an abdominal emergency [4]. Consequently, there is a high likelihood of the misdiagnosis of ectopic spleen, which presents a formidable challenge for physicians [6]. The vitality of the spleen determines whether it should be preserved or removed. Consequently, we present the case of a 45-day-old patient with splenic torsion who underwent splenectomy.

2. Case report
2.1. Presentation
A 45-day-old female patient was brought by her parents to our tertiary paediatric surgery centre with bilious vomiting and dark-coloured loose stools for 2 days. An erect abdominal X-ray was performed, revealing air-fluid levels (Fig. 1). On examination, the abdomen was soft, with lower abdominal distension.

The antenatal care period passed smoothly during the first trimester. The mother was diagnosed at the end of the first trimester with gestational diabetes, which was not well controlled. During the third trimester, she developed high blood pressure, and albuminuria was identified during a follow-up visit. The mother is a non-smoker and has no history of abortion or stillbirth. Our patient has 3 healthy older sisters.

The parents are not consanguineous. Additionally, there is no relevant family history of congenital malformations on either parent’s side.

Our patient was born at home through vaginal delivery at 37 weeks, despite the doctors’ advice to the mother to have a Caesarean section because of the risks associated with gestational diabetes. The mother experienced obstructed labour at home; a doctor was contacted and the baby, who weighed 4.9 kg, had to be resuscitated at home. She was then transported to the nearest ED, where she was admitted to the neonatal ICU to be treated for respiratory distress. She was discharged after 3 weeks. She was then diagnosed with a ventricular septal defect (VSD), which is being treated conservatively.

After approximately two weeks of being healthy at home, she was referred to us with a 48-hour history of greenish vomiting and loose, dark stools.
2.2. Operative details

Under general anaesthesia in the supine position, the abdominal cavity was accessed via a right supraumbilical transverse incision, which was later extended for better exposure of a firm mass felt in the left iliac fossa. The mass was tightly adherent to the posterior wall of the bladder. There were adhesions between the mass, the duodenum and the jejunal loops that were dilated, while the ileal loops were collapsed; hence, the incision was extended for better visualization of the surrounding structures to avoid inadvertent injury. Careful dissection and separation of the mass from the posterior wall of the urinary bladder was performed, and complete separation was achieved. All surrounding structures, namely, the ovaries, fallopian tubes, the uterus and the urinary bladder, were carefully inspected and found to be intact (Fig. 2). The kidney was also inspected and found to be in a normal retroperitoneal position. There were two vessels running along the surface of the mass (Fig. 3). They were believed to be the vascular pedicle of the mass and were most likely the splenic vessels arising from the celiac artery.

Because we were not able to confirm the nature of the mass (which was later proven to be an ectopic spleen) at the time of the operation, resection was preferred.

2.3. Pathology

Pathological examination uncovered the true nature of the excised mass, which had a gross size of 4.5 × 4 × 1 cm and a gross friable tan-brownish cut section with areas of cystification. Microscopically, it included capsulated splenic tissue showing wide devitalization and breakdown together with haemorrhage and focal dystrophic calcification, and it was surrounded by dilated hamartomatous blood vessels with cavernous spaces (Figs. 4–5). The report concluded with a diagnosis of a pelvic mass with features compatible with ectopic splenic tissue with hamartomatous vascular malformation. A literature review showed that splenic hamartomatous vascular malformations are seldom encountered, especially in children and newborns. The condition was first reported in 1861 by Rokitansky [7], and approximately 150 cases documented in the literature to date [8], only 20 of which were in children [9].
Fig. 5. Haematoxylin and eosin (×200 original magnification) staining showing breakdown together with haemorrhage and surrounded by dilated haemorrhagic blood vessels with cavernous spaces.

2.4. Postoperative recovery

Postoperatively, the patient had a haemoglobin level of 7.9 g/dl, compared to a preoperative level of 10.5 g/dl (normal range for females aged 31 days - 2 months = 10.7–17.1 g/dl) [10–12]. Nonetheless, she was vitally stable; thus, blood transfusion was considered safely avoidable, but iron supplements were prescribed. In addition, she had normal chemistry and electrolytes, as follows:

- Na⁺ = 142 mEq/L (n = 130–140 mEq/L)
- K⁺ = 4.5 mEq/L (n = 3.5–5.0 mEq/L)
- Creatinine = 0.3 mg/dl (n = 0.2–0.9 mg/dl) [13–18].

Postoperative alpha fetoprotein measured 108 IU/ml (=130.68 ng/ml) [18].

Furthermore, an abdominopelvic ultrasound confirmed the absence of a spleen in the left hypochondriac region, which also helped confirm the diagnosis of an ectopic spleen. Post-splenectomy immunizations, i.e., pneumococcal, meningococcal and haemophilus influenzae, were scheduled.

The postoperative period was rather uneventful, except for the patient’s prolonged stay of 1 week following her operation and her slow recovery. alpha fetoprotein was 108 IU/ml (=130.68 ng/ml) [10,18].

Laboratory work-up is deemed non-specific; however, signs of hypersplenism or functional asplenia as well as elevated inflammatory markers could be considered a vague clue [25].

Different imaging modalities, such as ultrasonography (U/S), nuclear scintigraphy, computed tomography (CT), and magnetic resonance imaging (MRI), may be helpful for achieving a definitive diagnosis of an ectopic spleen [26].

In terms of the management of a wandering spleen, whether complicated or not, surgery is the safe choice because conservative management of an asymptomatic wandering spleen is associated with a complication rate of 65% [27]. Considering that the wandering spleen is usually free of attachments to other organs, a laparoscopic approach may be considered. Although splenectomy is recommended if a gangrenous spleen is encountered, that is not the usual situation; because a healthy spleen may also be found incidentally upon laparotomy, splenectomy is recommended as the preferred course of action to avoid any chances of future complications and to preserve splenic function. The preservation of a healthy spleen is also recommended if complications associated with torsion, such as infarction, thrombosis or hypersplenism, have been excluded; this is especially important in young patients (from birth to the third decade of life) because they may be at risk of post-splenectomy sepsis [28]. Partial splenectomy of a totally strangulated spleen is rather infeasible; nevertheless, partial infarction of a wandering spleen requiring partial splenectomy followed by splenopexy or splenectomy and splenic implantation has been reported in the literature [29].

3. Discussion

Ectopic spleen is a rare condition that can affect adults and children and is usually discovered between the third and fifth decades of age. Among adults, wandering spleen affects females more often than males [19,20]. In contrast, as previously mentioned, it is 2.5 times more common in males than in females among children younger than 12 months of age. Additionally, approximately 30% of cases occur in children, typically in those older than 10 years of age [19].

As a general rule, the spleen is held in its anatomical position in the left upper abdominal quadrant by 3 main ligaments, which are believed to be always present, and 2 other ligaments that are variably present. The three main constantly present ligaments are the splenorenal ligament, located where the peritoneum contacts the lesser sac between the left kidney and the spleen and through which the splenic vessels pass between its two layers; the splenogastric ligament; and the splenocolic ligament. In addition to the aforementioned ligaments, there are 2 other ligaments that are variably present, namely, the splenoportal and splenophrenic ligaments. The abnormal position of a wandering spleen, which is commonly in the pelvis, results from the effect of gravity on the spleen, which is vulnerable due to lax ligaments, congenital malformation, the congenital absence of any of the ligaments, and congenital anomalies in the development of the dorsal mesogastrium, which lead to hypermobility within the abdominal cavity; in such cases, the spleen is only attached to its elongated vascular pedicle, which is the source of most of the resulting complications, such as splenic infarction and infarction [21]. In contrast, among older age groups, various conditions are believed to be associated with the acquired ligamental anomalies that cause a wandering spleen, such as splenomegaly, previous pregnancies and the enlargement or absence of a kidney [4].

In cases of splenomegaly and multiple pregnancies, the laxity of the spleen’s supporting ligaments is amplified by the direct effects of gravity and oestrogen, respectively [22].

Clinical diagnosis is rather challenging because the condition can vary from being asymptomatic, which is the case in 15% of children with wandering spleen [23], to being an abdominal emergency that requires urgent surgical intervention. Symptoms are most commonly attributed to complications related to torsion, which occurs in approximately 64% of cases of paediatric wandering spleen [24]. In addition, symptoms can be attributed to other complications, such as compression of another organ by the spleen or its pedicle. Additionally, there is an increased risk of splenic trauma as a result of obstructed labour or neonatal resuscitation, as might have happened with our case. A palpable abdominal mass outside the left upper quadrant is the most common physical finding in approximately 90% of children with wandering spleen; the mass represents the enlarged spleen that could be torsed. Additionally, approximately 55% of children with wandering spleen present with abdominal pain [23].

Laboratory work-up is deemed non-specific; however, signs of hypersplenism or functional asplenia as well as elevated inflammatory markers could be considered a vague clue [25].

4. Conclusion

A high degree of suspicion is a prerequisite for successfully diagnosing a case of wandering spleen because it is a rare condition with a non-specific presentation, especially in neonates. The spleen should be considered when an abdominal mass is palpated, particularly if there is pain or other abdominal symptoms. In addition, many imaging modalities, especially abdominal ultrasound, can offer great help in reaching a diagnosis. Optimally, preference should be given towards preserving the spleen if it remains viable during surgery; otherwise, splenectomy is required, followed by prompt, appropriate prophylaxis and immunizations.
Splenopexy or gastropexy may be performed in elective situations; however, gastropexy seems to be superior for holding the spleen in a normal anatomical position and avoiding splenic manipulation [25].

Disclosure

The authors declare no conflict of interest.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

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Conflict of interest

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Solafa Amin: pathologist; wrote the report.

Appendix A. Supplementary data

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References