CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 38 (2017) 131–135

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Torsion of huge wandering accessory spleen. Case report and review of literature

Salah Termos*, Ahmad Redha, Riad Zbibo, Abdulla Alduwaisan, Majd AlKabbani, Nidal Elyousif, Mohammad Alali

Hepatobiliary and Transplant unit, Department of Surgery, Amiri Hospital, Kuwait

ARTICLE INFO

Article history:
Received 16 June 2017
Received in revised form 15 July 2017
Accepted 16 July 2017
Available online 22 July 2017

Keywords:
Case report
Accessory spleen (AS)
Wandering accessory spleen
Torsion of accessory spleen
Splenectomy

ABSTRACT

INTRODUCTION: Accessory spleens are found in 10–15% of the population, and are even more prevalent in patients with hematological disorders (Rudowski, 1985). It infrequently may become symptomatic due to torsion, spontaneous rupture or hemorrhage which may lead to death. Torsion of an accessory spleen is extremely rare, and requires prompt medical attention [2] (Coote et al., 1999).
PRESENTATION OF CASE: We report the case of a 27-year-old Mediterranean lady with thalassemia trait, who presented to the emergency department with an acute surgical abdomen due to torsion of a giant accessory spleen, measuring 13 cm. She was diagnosed with the aid of ultrasound and computed tomography (CT) scan and was treated surgically through resection of the spleen.
DISCUSSION AND CONCLUSION: Torsion of an accessory spleen is not common, and is the surgical indication in about 0.2–0.3% of splenectomies (Mortele et al., 2004). It has variable clinical presentations, and is a difficult preoperative diagnosis due to lack of specificity of symptoms. Accessory spleens are usually smaller than 3 cm, with few cases being reported as larger than 10 cm larger accessory spleens have a higher rate of torsion. Knowledge of this pathology, and familiarity with its radiological findings are fundamental to accurately diagnosing and manageming this challenging condition.

1. Introduction

An accessory spleen is also known as a supernumerary spleen, splenunculi or splenule. It is a congenital foci or ectopic mass of healthy splenic tissues that is found separate from the main body of the spleen [4], They are found in 10–15% of the general population, and in even greater frequency in patients with hematological disorders. They may mimic enlarged lymphadenopathy, cysts and tumors in other abdominal organs (pancreas, adrenal gland & kidney) and occasionally may become symptomatic and clinically relevant due to torsion, spontaneous rupture or hemorrhage which may lead to death [1–3,5].

2. Case presentation

In our manuscript, we report the case of a 27-year-old Mediterranean Lebanese femalewho presented with a few hours history of severe diffuse abdominal pain, mainly in the left upper quadrant. Her pain was sudden in onset, aching in nature and radiating to the left intra-scapular area and left shoulder, associated with nausea and 3 episodes of non-bilious and non-bloody vomiting. Her symptoms did not respond to pain killers or antiemetics. Past medical and surgical history was positive for gastroesophageal reflux disease, hiatal hernia and gastritis.

On presentation, the patient was distressed with severe pain. She looked pale but was hemodynamically stable.Her vital signs showed a temperature of 37.5 Celsius, heart rate of 95, and a blood pressure of 100/70. Physical exam of the abdomen showed marked upper abdominal tenderness mainly over the epigastric area and left hypochondrium, with voluntary guarding. Her laboratory investigations revealed a white blood cell count of 13.6 $10\hat{9}$/L, Hemoglobin 8.7 g/dL and a MCV of 67. The remainder of her laboratory investigationswere within normal ranges.

Radiological investigation started with an ultrasound, which revealed a well-defined, oval, hypoechoic solid mass measuring $12 \times 5 \times 3$ cm. It was found to be avascular (absent Doppler signal) located in the left flank between the left kidney and spleen. A minimal amount of fluid was noted in the left subphrenic space [Fig. 1]. An enhanced CT scan of abdomen and pelvis was performed which showed a 13×6 x3 cm oval hypodense and non-enhancing mass with regular contour in the left upper quadrant, adjacent to the stomach and anteromedial to the native spleen with no evidence of communication. Surrounding fat Stranding with minimal fluid

* Corresponding author at: 25 Arabian Gulf Street, Kuwait City, 13041, Kuwait.
E-mail addresses: dr.termos@hotmail.com, salahtermos@gmail.com (S. Termos).

http://dx.doi.org/10.1016/j.ijscr.2017.07.037

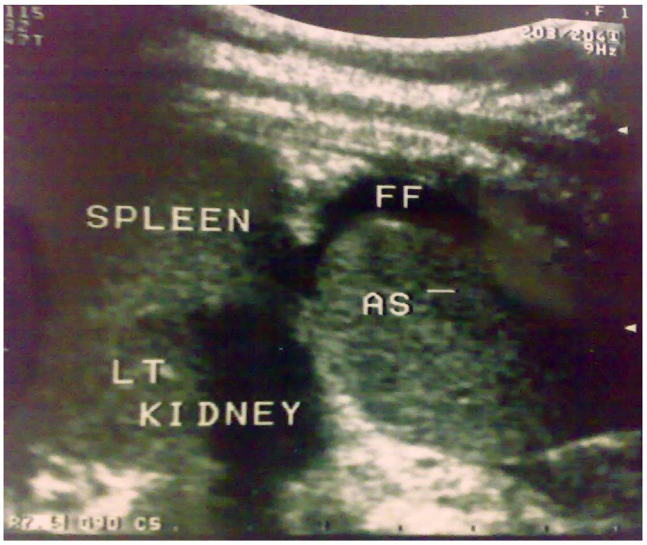

Fig 1. Ultrasound abdomen, showing a well-defined, hypoechoic solid mass measuring 12 × 5 × 3 cm.

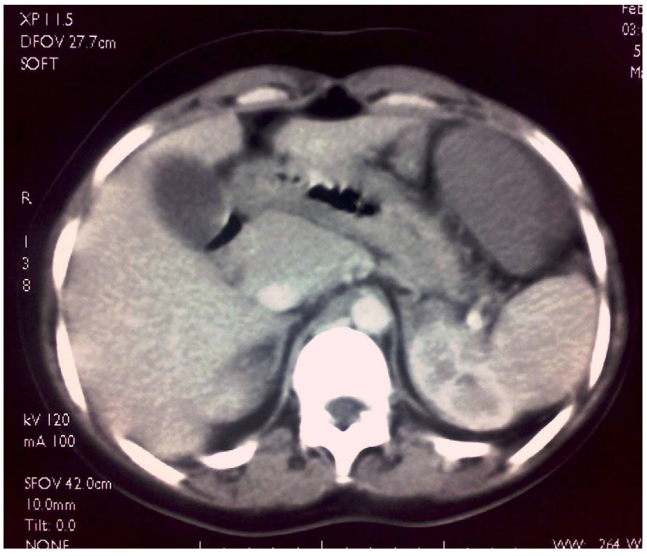

Fig. 2. Enhanced CT scan of abdomen revealing a 13 × 6 x 3 cm non-enhancing accessory spleen larger than the native spleen.

around the mass was also noted [Fig. 2]. A small round mass was also found near the native spleen [Fig. 3].

On the basis of the radiological findings and in view of the intolerable persisting abdominal pain, we decided to proceed with surgical intervention. Surgical access was obtained using a small upper midline incision. A 13 × 6 cm dark blue mass attached to a long mesentery was found wandering under the midline, which was easily manipulated and extracted through the incision. The accessory spleen was dusky with a tense capsule and fibrinous material overlying it [Fig. 4]. It was freely mobile, with evidence of a twist at its own pedicle of mesentery, therefore surgical resection was performed [Fig. 5]. The spleen was in its normal position, with evidence of another small accessory spleen, measuring 1 cm, at the hilum of the normal spleen which was kept in place. Histopathalogical examination showed a 13 × 6 x 3.2 cm mass weighing 196 g containing advanced ischemic changes with hemorrhage of splenic parenchyma. Findings were consistent with splenic infarction. Post-operative course was smooth and uneventful and the patient was discharged home three days later in good

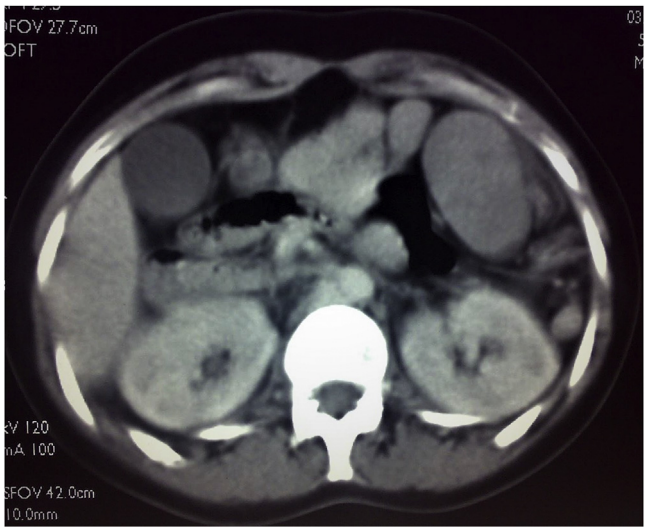

Fig. 3. Another small accessory spleen was also found near the native spleen.

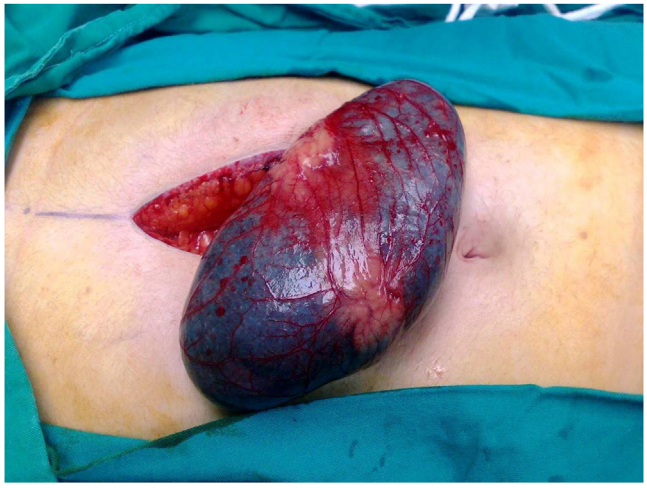

Fig. 4. Ischemic huge accessory spleen.

condition. The patient was later investigated for her low Hb and MCV in the clinic and was found to have thalassemia trait. Follow up ultrasound revealed no abnormal findings.

3. Discussion and conclusion

Accessory spleens are a common pathology, and are considered a variation of normal development of the spleen. It is seen in 10%-20% of patients at autopsy and in 16% of patients undergoing contrast-enhanced CT of the abdomen [6]. According to the literature there is an increased incidence of accessory spleens in patients with hematologic disorders. Accessory spleens are the most common of all splenic anomalies. The vast majority of them are encountered in the vicinity of the splenic hilum, along the splenogastric ligament, splenic vessels, omentum, mesentery, left broad ligament in females or the left spermatic cord in male patients [7,8]. Embryological development of the spleen occurs from the mesenchymal cells migrating to the dorsal mesogastrium. Incomplete fusion of splenic tissue in the fifth week of fetal life gives rise to formation of an accessory spleen. Because the spleen is formed in the dorsal mesogastrium and then rotate to the left side, accessory spleens are always situated on the left side of the abdomen [9].

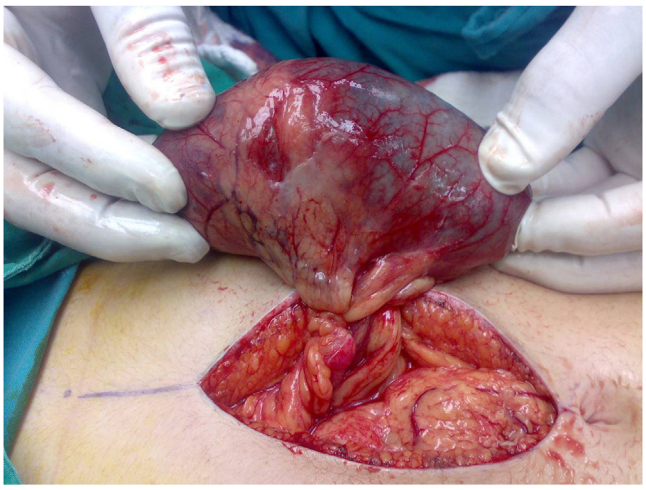

Fig. 5. Twisted long mesentery of the accessory spleen.

Development of the splenic ligaments occurs as demonstrated in the figure [Fig. 5]. A. Gastric and splenic rotation B. Formation of omental bursa and its relation to the spleen. C. Beginning of formation of the greater omentum and its relationship to spleen D. Formation of two major splenic ligaments.

Accessory spleens can be solitary or multiple; they are usually asymptomatic and diagnosed incidentally by radiological imaging. Typically, accessory spleens appear on CT scans as well-marginated, round masses that are usually smaller than 2 cm and enhance homogeneously on contrast-enhanced images. When accessory spleens are smaller than 1 cm, their attenuation may be lower than that of the spleen because of partial volume effects. Their most frequent location (22%) is posteromedial to the spleen; anterolateral to the upper pole of the left kidney; and lateral, posterior, and superior to the tail of the pancreas. Our patient was diagnosed to have thalassemia traits, supporting previous claims of an increased incidence in patients with hematologic disease. Although it may be possible for several accessory spleens to be found at the hilum, it is rare to find accessory splenic tissue in more than two locations [10]. Our case was unusual, as the patient had two splenules with variable sizes and located in different sites. One of them was also a wandering accessory spleen, with no fixed location, that had twisted at its mesentry. Familiarity with these characteristic features may differentiate them from other pathologic findings in the upper abdomen [11].

The vascular pedicle of an accessory spleen is commonly related to the splenic hilum. On dynamic CT, supplying vascular branches arising from the splenic artery were found in about 43.3% of cases [12]. It may also be related to the tail of the pancreas, gastrosplenic ligament, and small bowel mesentery or to the vessels of the gastric fundus [13]. If the vascular pedicle of the native spleen is long, it may be located anywhere in the peritoneal cavity and there have been reported cases of 'wandering spleens' [14]. Our patient had a native spleen in its anatomical position, so a wandering spleen was excluded. She had a long pedicle attached to her larger accessory spleen, extending from the region of gastric fundus at the gastro-omental area, which made for easy surgical access (Fig. 6).

Accessory spleens have a silent clinical course and may remain undetected for life. Clinical importance arises when splenectomy is considered, or when spontaneous torsion of an accessory spleen occurs. Torsion of the spleen is an extremely rare clinical presentation, and it represents the indication in about 0.2-0.3% of splenectomies [3]. It is a serious complication that occurs mainly

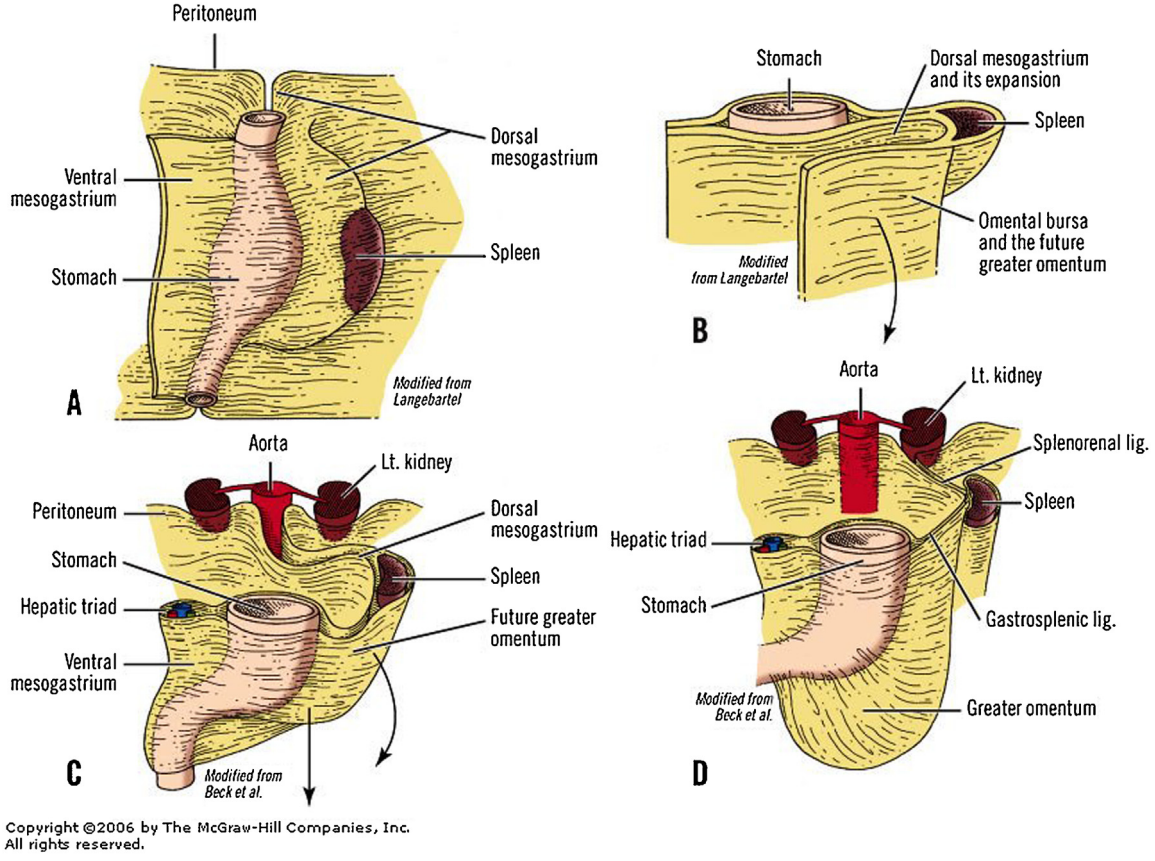

Fig. 6. Embryological development of splenic ligaments [15].

Table 1
Accessory spleens treated laparoscopically.

	Author	Sex	Age	Clinical Presentation	Size	Location
1	Mendi 2006	F	12	Recurrent LUQ pain	<3 cm	Splenic Hilum
2	Yousef 2010	M	12	Acute LUQ Pain	3.5 cm	splenic flexure
3	Lhuaire 2013	M	66	Recurrent abdominal pain	3 cm	Greater Omentum
4	Perin 2014	F	17	Asymptomatic wandering accessory spleen	6 cm	Pelvic Cavity
5	Ozeki 2015	F	31	Left Abdominal pain	3 cm	Greater Omentum

Table 2
Largest sizes in the English literature treated by laparotomy approach.

	Author	sex	Age	Clinical Presentation	Size	Location	Diagnosis	Approach
1	Valls 1998	F	13	Acute Abdomen	6 cm	Tail of pancreas	US/CT	Laparotomy
2	Grinbaum 2005	F	21	Left Upper Abdominal Pain	9 cm	Greater Omentum	US/CT	Laparoscopy
3	Yagmur 2008	M	34	Upper Abdominal Pain	10 cm	Left Colon	US/CT	Laparotomy
4	Impellizzeri 2009	M	12	Acute Abdominal Pain	8.5 cm	Root of Mesentery	US/CT	Laparotomy
5	Ishibashi 2012	F	3	Right Flank Pain (Situs Inversus)	7 cm	Greater Omentum	US/CT	Laparotomy
6	Bard 2014	F	20	Left Abdominal Pain	17 cm	Spleen	CT	Laparotomy
7	Koichi 2015	M	5	Left Lower Abdominal Pain	8 cm	Greater Omentum	US/CT	Laparotomy

in accessory or wandering spleens larger than 6 cm, manifested by acute abdominal findings. Venous return is disrupted first, causing hemorrhagic infarction of the parenchyma, which may lead to spontaneous rupture and death [13,15]. The scenario in our case was typical for torsion and vascular compromise manifested in the form of severe, progressive and non-relenting abdominal pain [16,17].

Intermittent torsion-detorsion may produce recurrent bouts of abdominal pain caused by short-lasting ischemia of the accessory spleen, or from direct mechanical irritation of surrounding organs. In the literature, there have been cases in which splenopexy had been reportedly used, mainly for wandering spleens with no evidence of infarction. Our patient however, had a normal native spleen and twisted accessory spleen with expressed ischemic changes, rendering this technique unsuitable [18].

Most of the reports describing torsion of accessory spleens present with an acute abdomen occur in childhood, with few cases being described in older patients [19]. It must be considered as a differential diagnosis when encountering acute abdomen in children with a palpable mass [9]. Our patient was 27 years old, and no mass was able to be palpated due to guarding. Usually, preoperative diagnosis of accessory spleen torsion is a difficult one to make, even with modern imaging techniques [14]. But due to the high index of suspicion and appropriate imaging modality, this patient's diagnosis was established before surgery.

The detection and characterization of accessory spleens are important in three clinical scenarios:

1. An accessory spleen may mimic lymphadenopathy and tumors in other abdominal organs such as the pancreas, the adrenal gland, or the kidneys.
2. Accessory spleens occasionally may become symptomatic due to torsion, spontaneous rupture, hemorrhage, and cyst formation.
3. A surgeon's awareness of their presence may be important when the intention is to remove all functional splenic tissue in cases of hematological diseases [20–23].

For an accessory spleen that requires surgical intervention, magnetic resonance (MR) angiography may be helpful due to the enhanced detailing of the anatomical structure and may influence the surgical approach (removal vs splenopexy). MR imaging has been reported to demonstrate the hemorrhagic components occurring secondary to infarction or venous ischemia. Magnetic susceptibility effects of blood degrading products such as deoxyhaemoglobin or methemoglobin, as well as the retraction of the clot determine the signal intensity on MR. The MR sequences that are sensitive to susceptibility effects are helpful in demonstration of hemorrhage. Angiography and scintigraphy were used in some elective cases. With 99mTc-denatured RBC scans used mainly to avoid unnecessary surgery, such as in the case of intrapancreatic accessory spleens, or 99mTc-sulfur colloid scintigraphy to detect ectopic splenic tissues for more detailed preoperative evaluation especially in smaller accessory spleens [24–26]. Our case was an urgent one that was treated on an emergent basis. We initially started our investigation with an ultrasound, followed by an enhanced CT scan to confirm the diagnosis. Following this, we decided to proceed for surgery and not to pursue any further imaging studies, as the patient was suffering from significant signs and symptoms of splenic infarction.

Laparoscopy has been tailored to treat smaller sized accessory spleens, which were diagnosed preoperatively, or as a diagnostic laparoscopy for acute surgical abdomen with obscured diagnosis [Table 1]. Laparotomy is used more frequently, mainly in larger accessory spleens. We collected the recent cases done in a table, showing the size and the mode of operation [Table 2]. In our surgery, we performed a small upper midline incision about 10 cm smaller than the twisted accessory spleen [Fig. 4]. We recommend adjusting the incision size and site according to the preoperative findings. The patient received post-splenectomy triple vaccinations two weeks following the procedure and was asymptomatic on her follow-up visit.

Torsion of the accessory spleen pedicle is an uncommon entity especially in adult populations. It is rarely diagnosed preoperatively, and review of the literature showed only a few cases of large accessory spleens. Our case was a 27-year-old lady with a giant twisted accessory spleen, measuring 13 cm, which was treated with surgical resection using a small incision. Awareness of this entity and familiarity with typical radiological findings are mandatory for proper evaluation and planning prior tosurgical management.

Funding

We report no involvement of sponsors.

Ethical approval

The case report was approved by the local ethic committee of Amiri teaching hospital. LEC-project number 17-2017.

Consent

Written informed consent was obtained from the patient and is available upon request. No patient identifying material was used in this manuscript.

Conflicts of interest

The authors declare no conflict of interest.

Acknowledgments

The work has been reported in line with the SCARE criteria [27].

References

[1] W. Rudowski, Accessory spleens: Clinical significance with particular reference to the recurrence of idiopathic thrombocytopenic purpura, World J. Surg. 9 (3) (1985) 422–430.
[2] J.M. Coote, P.S. Eyers, A. Walker, I.P. Wells, Intra-abdominal bleeding caused by spontaneous rupture of an accessory spleen: the CT findings, Clin. Radiol. 54 (1999) 689–691.
[3] K.J. Mortele, Bart Mortele, S.G. Silverman, CT features of accessory spleen, AJR 183 (2004) 1653–1657.
[4] K.L. Moore, Clinically Oriented Anatomy, 3rd ed., William & Wilkins, Baltimore (BA), 1992, p. 187.
[5] F.J. Pérez Fontán, R. Soler, M. Santos, I. Facio, Accessory spleen torsion: US, CT and MR findings, Eur. Radiol. 11 (2001) 509–512.
[6] E. Timothy, Torsion of an accessory spleen in en elderly patient, Postgrad. Med. J. 66 (1990) 838–839.
[7] J.L. Freeman, S.Z. Jafri, J.L. Roberts, D.G. Mezwa, A. Shirkhoda, CT of congenital and acquired abnormalities of the spleen, Radiographics 13 (1993) 597–610.
[8] Paterson, D.P. Frush, L.F. Donnelly, J.N. Foss, S.M. O'Hara, G.S. Bisset, A pattern-oriented approach to splenic imaging in infants and children, Radiographics 19 (1999) 1465–1485.
[9] K.B. Allen, B.B. Gay Jr, J.E. Skandalakis, Wandering spleen: anatomic and radiologic considerations, South. Med. J. 85 (1992) 976–984.
[10] M.F. Abu-Hijleh, Multiple accessory spleens: case report and literature review, Clin. Anat. 6 (4) (1993) 232–239.
[11] Koenraad J. Mortelé, Bart Mortelé and Stuart Department of Radiology, Brigham and Women's Hospital, Harvard Medical School, AJR 2004; American Roentgen Ray Society CT Features of the Accessory Spleen.
[12] R. Mendi, Evolution of the CT imaging findings of accessory spleen infarction, Pediatr. Radiol. 36 (2006) 1319–1322.
[13] L.B. Dahlin, L. Anagnostake, M. Delshammar, F.T. Fork, S. Genell, Torsion of an accessory spleen in an adult, Eur. J. Surg. 161 (1995) 607–609.
[14] T. Seo, T. Ito, Y. Watanabe, T. Umeda, Torsion of an accessory spleen presenting as an acute abdomen with an inflammatory mass US, CT, and MRI findings, Pediatr. Radiol. 24 (1994) 532–534.
[15] KB, B.B. Gay, J.E. Skandalakis, Wandering spleen: anatomic and radiologic consideration, South Med. J. 85 (10) (1992).
[16] D. Padilla, J.M. Ramia, J. Martin, R. Pardo, T. Cubo, J. Hernandez-Calvo, Acute abdomen due to spontaneous torsion of an accessory spleen, Am J Emer Med. 17 (1999) 429–430.
[17] T.E. Hems, J.F. Bellringer, Torsion of an accessory spleen in an elderly patient, Postgrad. Med. J. 66 (1990) 838–839.
[18] M. Wacha, J. Danis, W. Wayand, Laparoscopic resection of an accessory spleen in a patient with chronic lower abdominal pain, Surg. Endosc. 16 (8) (2002) 1242–1243.
[19] S.N. Mocanu, A. Sierra Vinuesa, C. Munoz-Ramos Trayter, E.A. Castaneda Figueroa, A. Garcia San Pedro, Accessory spleen torsion in a teenager, ANZ J. Surg. 85 (December (12)) (2015) 987–989.
[20] I. Hayward, R.E. Mindelzun, R.B. Jeffrey, Intrapancreatic accessory spleen mimicking pancreatic mass on CT, J. Comput. Assist. Tomogr. 16 (1992) 984–985.
[21] N. Tsuchiya, K. Sato, N. Shimoda, et al., An accessory spleen mimicking a nonfunctional adrenal tumor: potential pitfall in the diagnosis of a left adrenal tumor, Urol. Int. 65 (2000) 226–228.
[22] T. Facon, M.T. Caulier, P. Fenaux, et al., Accessory spleen in recurrent chronic immune thrombocytopenic purpura, Am. J. Hematol. 41 (1992) 184–189.
[23] A. Budzynski, A. Bobrzynski, T. Sacha, A. Skotnicki, Laparoscopic removal of retroperitoneal accessory spleen in patient with relapsing idiopathic thrombocytopenic purpura 30 years after classical splenectomy, Surg. Endosc. 16 (2002) 1636.
[24] G. Gayer, Y. Zissin, CT findings in congenital anomalies of the spleen, Br. J. Radiol. 74 (884) (2001) 767–772.
[25] I. Domínguez, Accessory spleen as a differential diagnosis of intrapancreatic tumors. Case report and review of the literature, Rev. Gastroenterol. Mex. 72 (October–December (4)) (2007) 376–378.
[26] H. Ohta, K. Kohno, N. Kojima, N. Ihara, T. Ishigaki, G. Todo, et al., A case of diaphragm hernia containing accessory spleen and great omentum detected by Tc-99 m phytate scintigraphy, Ann. Nucl. Med. 13 (5) (1999) 347–349, http://dx.doi.org/10.1007/BF03164874.
[27] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, the SCARE Group, The SCARE statement: consensus-based surgical case report guidelines, Int. J. Surg. 34 (2016) 180–186.