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Splenic torsion, a challenging diagnosis: Case report and review of literature

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ABSTRACT

INTRODUCTION: Wandering spleen is an unusual condition characterized by hypermobility of the spleen. This is a rare clinical entity and it's more common in childhood under 1 year of age and in third decade of life. In this second peak, it's more frequent in females. Clinical manifestations can vary from asymptomatic to abdominal emergency. Treatment is often surgical.

PRESENTATION OF CASE: We presented a case report of splenic torsion from our hospital and a review of cases described in literature. This is a 40 year-old woman with complaints of upper abdominal pain associated with nausea and vomiting. A marked tenderness and a palpable abdominal mass on left hypochondrium were found as well as a slight increase in inflammatory parameters. A CT was performed and demonstrated findings compatible with splenic torsion. Surgery was performed doing laparoscopic splenectomy; Review of literature was made using the keyword combination: "wandering spleen". The research resulted in 451 articles.

DISCUSSION: The physical examination and CT are fundamental for diagnosis. Surgery was performed and laparoscopic splenectomy was made because infarcted spleen; about the review of literature, the majority of patients were female and the average age at the time of diagnosis was 25.2 years. 69.5% needed splenectomy and 78.6% of surgeries were laparotomic.

CONCLUSION: Splenic torsion is a rare but important differential diagnosis in patients presenting with acute abdomen. Diagnosis should be made promptly before development of life-threatening complications. Surgery is often necessary and splenopexy or splenectomy can be done.

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1. Introduction

Wandering spleen (WS), or ectopic spleen, is an unusual condition characterized by hypermobility of the spleen due to underdevelopment or absence of the ligaments that attach the spleen to the left upper quadrant [1,2]. This is a rare clinical entity, with only about 500 cases reported worldwide and an incidence rate of 0.2% [3].

Clinical manifestations are variable and vary from asymptomatic to abdominal emergency [1]. Due to rarity and various mode of presentation, it has been a diagnostic and therapeutic challenge for the physician [4].

The aim of this article is to present a case of 40 year-old woman that presented in our community institution and review the literature of all the described cases of wandering spleen.

This work has been reported in line with the SCARE criteria [5].

2. Presentation of case

A 40-year-old caucasian woman was referred to our Emergency Department after 48 evolution of an upper abdominal pain associated with nausea and vomiting. Her past medical history was negligible. She was afebrile but tachycardia and, in the abdominal physical examination, a marked tenderness and a palpable abdominal mass on left hypochondrium was found.

In the biochemical parameters we identified haemoglobin of 10.5 g/dL, leukocytosis (22,700/mm³) and a slight increase in a C-reactive protein (32.75 mg/L). A contrast-enhanced abdominopelvic computed tomography (CT) was performed and demonstrated: the absence of spleen in its normal position (spleen was in left flank), no contrast enhancement within the splenic parenchyma and whirl appearance of splenic pedicle

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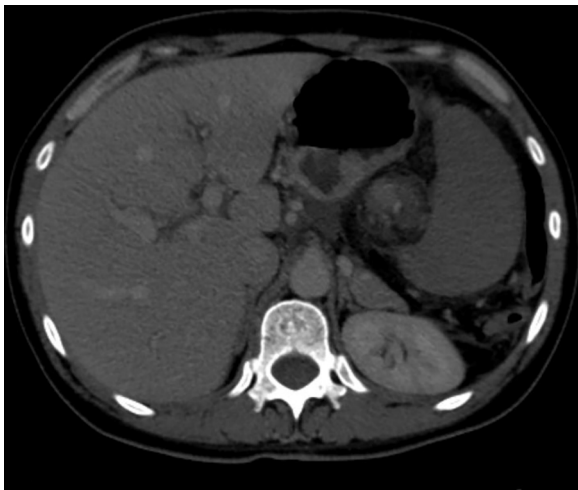


Fig. 1. Axial CT image showing whirl sign and no contrast enhancement within splenic parenchyma.

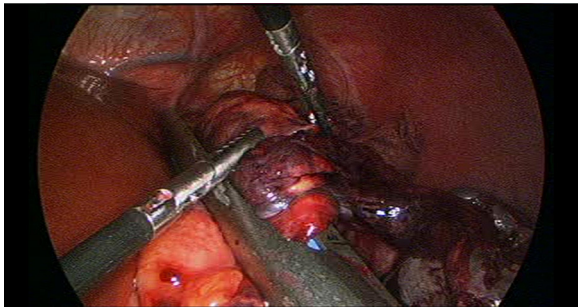


Fig. 2. Intra-operative laparoscopic image showing splenic torsion.

(Fig. 1). This whirl sign strongly suggests a diagnosis of splenic torsion so, surgery treatment was performed immediately. Urgent exploratory laparoscopy confirmed splenic torsion with an infarcted spleen secondary to torsion of a long splenic pedicle (Fig. 2). No perfusion after de-torsion led to laparoscopic splenectomy.

The postoperative period was uneventful. The patient was discharged on the 4th postoperative day without complaints. The patient was already re-evaluated in consultation and she was completely asymptomatic.

3. Review of literature

The research was made under the Pubmed Platform using the following keyword combination: “wandering spleen”.

We collected the cases submitted to surgery, distribute by year intervals, verify average age at the time of diagnosis and calculate the cases submitted to splenectomy or splenopexy, either laparoscopically or laparotomically. The research resulted in 451 articles, which are reviewed individually. We excluded: cases of accessory spleen, cases in animals, articles with no clinical cases and cases with no/partial access. So, of the 451 articles, we excluded 233 and included 218 (Fig. 3).

The Table 1 presents the results from our research organized by intervals of years.

In total we analyse 266 cases of wandering spleen, 204 females (76.8%) and 62 males. The average age at time of diagnosis was 25.2 years. More than half, 185 cases (69.5%), were submitted to splenectomy, 78 to splenopexy (29.3%) and 3 weren't submitted to

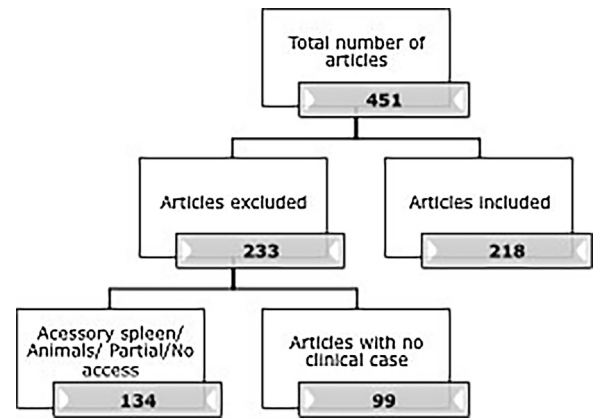


Fig. 3. Diagram showing that, in a total of 451 articles, we excluded 233 and included 218.

surgery (conservative treatment). The majority, 209 cases (78.6%), had open surgery and 54 (20.3%) were intervened by laparoscopy.

4. Discussion

Wandering spleen is a rarely diagnosed clinical entity [1]. It is defined as mobile spleen that is attached only by an elongated vascular pedicle, allowing it to migrate to any part of the abdomen or pelvis [2].

It was first described by Von Horne in 1667 however, one of the first case reports of a wandering spleen in a child was published in 1854 by the Polish physician Józef Dietl that not only prognosticated the life threatening complications of this condition but also considered the relaxation, extension or hypoplasia of splenic ligaments as the major cause to the spleen wander [6,3].

There are some conditions associated with WS: enlargement or absence of kidney, infectious mononucleosis, malaria, Hodgkin's disease, Gaucher's disease and previous pregnancy [7]. This is, for the majority, justified by heavy spleens seen in these pathologies as well as hormonal changes and abdominal laxity verified in multiparous women [3].

WS has two peaks of incidence: in childhood, especially bellow first year old as the commonest age, followed by the third decade of life [7,1]. WS is 7 times more common in females than males after age 10 and 2.5 times more common in males than females under the age of 1 year [1]. Also in this exhaustive research we verified preponderance for the feminine gender (76.8%) in the third decade of life (average age 25.2 years), just like the patient we present.

Clinical manifestations vary from asymptomatic to abdominal emergency [1]. The most common presentation sign in adult patients is a mass (as we report) with or without subacute abdominal or gastrointestinal complaints, or with acute abdominal findings. The most common presentation in children is acute abdominal pain [7].

Symptoms may remain limited or absent for long periods of time. Nevertheless, complications related to torsion or compression of abdominal organs are quite common and may cause progressive splenic infarction and necrosis of pancreatic tail [7,15,16]. Initially, irreducible torsion produces venous congestion and the spleen becomes edematous and enlarged. If the torsion is progressive, results in infraction of arterial supply, acute ischemia, strangulation, necrosis and splenic rupture [1].

Other complications include pancreatitis, bowel obstruction, gastric volvulus and gastric and duodenal compression however, splenic infarction is, undoubtedly, the most common complication [8].

Table 1
Representation results about the research in pubmed with keywords: “wandering spleen”.

Year	Total articles	Articles excluded	Articles included	N (cases nr)	First author	Gender (Female –F; Male – M)	Average Age (years)	Splenectomy vs Splenopexy vs Conservative	Laparotomy (LPT) vs Laparoscopy (LPS) vs Conservative
1903–1930	4	4	0	0	X	X	x	X	X
1931–1960	19	18	1	2	Abell I.	2F	40	2 Splenectomy	2 LPT
1961–1990	93	77	16	19	Binns JH., Simpson A., Miller EL., Smulewicz JJ., Lernau OZ., Isikoff MB, Sorgen RA., Koppikar MG., Stringel G., Sheflin JR., Vermynen C., Angeras U., Witz M., Samuel N., Smevik B., Seashore JH.	15F	21.5	17 Splenectomy	19 LPT
1991–2000	85	46	39	49	Herman ZW., Nemcek AA Jr, Herman TE., Risques J., Risques J, Balik E., Gunning KA, Dawson JH., Wong SK., Melikoglu M., McFee RB., Lopez-Menendez C., Papakyriacou K., Ellis H., Calik A, Moll S., Sarimurat N., Ng T., Saadaoui H., Moorthy K., Uc A, Cohen MS., Gurski RR., Hirose R., Hon TY, Raissaki M, Baglaj M., Benoist S., Kanthan R., Lane TM., Haj M., Vural M., Ravid A., Spector JM., Ugolini G., Nawaz A., Nomura H., Zimmermann ME, Emir H.	4M 40F	23.2	2 Splenopexy 34 Splenectomy	41 LPT
2001–2010	138	57	81	108	Peitgen K., Habib E., Benevento A., Miele V., Sayeed S., Steinberg R., Chan KC., Githaiga JW., Kim SC., Gilman RS., Dalpe C., Brown CVR., Moran JC., Taori K., Balliu PR., Al-Mashat FM., Lu CC, Ajello A., Fearn C., Corcione F., Deux JF., Alimoglu O., Tucker ON., Cavazos S., Parvaiz A., Martinez-Ferro M., McHeik JN., Puglisi F., Hedeshian MH., Saxena AK., Karmazyn B., Schaarschmidt K., Eposito C., Taori K, Lin CH., Thambidorai CR., Lopez-Tomassetti Fernandez EM., Kleiner O., Fukuzawa H., Takayasu H., Castellon-Pavon CJ, Liu HT, Soleimani M, Tan HH, Lacreuse I, Upadhyaya P, Barbaros U, Falchetti D., Palanivelu C., Amer HZ, Cho SA, Misawa T., Lebron R., Feroci F, Sodhi KS, Bem Ely A., Singla V., Alshukry SM, Wani S, Menéndez P, Iqbal M., François-Fiquet C., Prieto-Montaña J., Lien C.H., Parlakgumus A., Dirican A., Biondi A., Di Crosta I., Turhan A., Dillman J.R., Montenovolo M.I., Yasuda H, Carmona J., Saouab R., Maschio M., Hussain M., Sanchez R., Okasaki T., Ghazeeri G., El Bouhaddouti H., Broadis E.	9M 75F	17.7	14 Splenopexy 1 Conservative 62 Splenectomy	7 LPS 1 Conservative 82 LPT
2011–2016	112	31	81	88	Somalwar A.M., Chu J., Maamouri N., Irak K., Parisinos C.A., Ityel H., Cripps M., Magno S., Sucandy I., Bouassida M., Lam Y., Parrondo MG, Kapan M., Lemke J., Bekheit M, Ayaz U.Y., Yucel E., Fabiano P, Cecchetto G., Le K., Mehta A., Fonseca A.Z., Anyfantakis D., Pérez-Legaz J, Leci-Tahiri L, Udare AS, Priyadarshi R.N., Zarroug A.E., Lianos G., Tapar V., Yilmaz O., Centonze A, Tran S., Misra A.P., Holst JM1, Heydari MB, Leci-Tahiri L, King CR, Bhanumathi V., Memari M., Lombardi R., Tchidjou HK, Bouzaidi K, Ihedioha U., Richman M., Karakus SC, Goyal R.B., Sharma A., Blouhos K., Gorski U., Kaabar N., McBrearty A., Clark J.K., Maksoud S.F., Riaz ul Haq M., Katsura S., Rellum R., Faridi M.S., Ho CL, Hao F., Aswani Y., Mohammadi A., Puranik A.K., Torri F., Jiang M., Jude N.N., Singh S., Köseoğlu H, Sato M, DeJohn L, Sojo-Rodríguez V., Ahmadi H, Makhlof NA, Chauhan NS, Omata J, Radillo L, Din S, Touarsa F, Dème H, Cantone N, Zorron R	33M 72F	23.8	46 Splenopexy 70 Splenectomy	26 LPS 65 LPT
Total	451	233	218	266	X	204F 62M	25.2	185 Splenectomy 78 Splenopexy 3 Conservative	209 LPT 54 LPS 3 Conservative

Articles published in pubmed organized by intervals of years. In total we analyse 266 cases of wandering spleen, 204 females. The average age was 25.2 years. 185 were submitted to splenectomy (69.5%), 78 splenopexy (29.3%) and 3 weren't submitted to surgery. 209 (78.6%) had open surgery and 54 (20.3%) were intervened by laparoscopy.

Preoperative diagnosis of wandering spleen is rarely suggested based on clinical findings alone, because of nonspecific symptoms [2].

Laboratory tests are usually nonspecific too. It can usually reveal evidence of hyperesplenism or functional asplenia (by evaluating peripheral smears for Howell-Jolly bodies or other particles) in particular, when associated with torsion of an elongated splenic pedicle [9]. In our case we just found leukocytosis and a slight increase in a C-reactive protein.

Since a clinical diagnosis can be difficult, non-invasive imaging procedures, such as sonography, nuclear scintigraphy, CT and magnetic resonance, are the common diagnostic modalities. Plain radiographs and barium examinations are usually non-specific [9].

The absence of spleen in left hypochondrium and the presence of a soft tissue mass resembling a spleen in the lower abdomen is highly suggestive of WS. Both abdominal sonography as the CT is usually accomplished to diagnose WS [10]. Doppler is excellent to find vascular compromise in torsed pedicle [10] but it is operator dependent as bowel gases can obscure the findings [4,10]. Post-contrast CT scan remains the investigation of choice because it can demonstrate the organ's circulation and the viability of splenic parenchyma [4,9]. This information is crucial to reveal splenic infarction and consequently, to decide the surgery option [9,3].

The CT manifestations of torsion in WS are:

- 1) Splenomegaly – non-specific but it is an important sign of torsion;
- 2) Poor or absent enhancement of splenic parenchyma – observed on contrast-enhanced CT, as we identified in our patient, indicates poor perfusion. It is an indirect sign of pedicle torsion and subsequent infarction but it is non-specific and may sometimes be mistaken by other lesions, like cystic lesion or fluid collection;
- 3) Hyperdense splenic pedicle on unenhanced scan – suggests thrombus in the torsed pedicle;
- 4) Rim sign or pseudocapsule sign – signal of chronic torsion and so, chronic ischemia;
- 5) Whirl appearance of splenic pedicle – pathognomonic CT finding of torsion and includes demonstration of a thickened and coiled vascular pedicle [10]. This last sign was present in our case, too.

Operative management (laparotomy or laparoscopy) is the treatment of choice in uncomplicated and in complicated cases because conservative treatment of an asymptomatic WS is associated with a complication rate of 65% [1]. The surgical intervention can be either splenopexy or splenectomy and the surgical option is defined by the vascularity of the spleen [3,17].

At the beginning of the twentieth century, splenopexy became the appropriate treatment to prevent complications of WS if spleen is viable and there's no evidence of infarction, thrombosis or hypersplenism [6,9,11]. Splenopexy, in addition to conserve spleen, prevent any future complications and so, is the method of choice, whenever possible, especially in children.

Splenectomy is necessary if there is splenic infarction, massive enlarged spleen, splenic vessel thrombosis, secondary hyperesplenism, functional asplenia due to torsion or any suspicious of malignancy [9]. Sometimes, pericapsular fluid collection and hemoperitoneum can be present because of inflammatory fluid following splenic infarction and to splenic vessel rupture following torsion, respectively [17]. In our descriptive research we verified that splenectomy occurred in 69.5% of the cases, which shows that, at diagnosis, the spleen had already signs of suffering in a high number of cases. In our patient, because of splenic torsion with an infarcted spleen that didn't recovered after de-torsion, splenectomy was necessary.

If it is not possible giving before splenectomy, after surgery it is mandatory prophylactic antibiotic therapy and vaccination

for encapsulated bacteria like *Neisseria meningitidis*, *Haemophilus influenzae*, and *Streptococcus pneumoniae* [12,17] to prevent postsplenectomy sepsis syndrome [8]. In our institution, after discharged, all the patients without spleen are advised to have always antibiotic at home. This is important in case of infection, for patients take at the first sign of fever, and just after goes to the hospital.

Both surgeries can be done by laparotomy or laparoscopy, depending on surgeon experience. Table 1 shows that there was a progressive increase in laparoscopic surgery over time however, in the last 5 years (2011–2016), more than a third of the cases worldwide were performed by laparotomy (65 from a total of 76 surgeries). In our hospital, currently, splenic surgery by laparoscopic approach is the preferred technique and is used whenever possible, because all the advantages of laparoscopic surgery: less painful, better cosmesis, early ambulation, overall less morbidity and a faster return to work [4].

5. Conclusion

Torsion of wandering spleen is a rare but important differential diagnosis in patients presenting with acute abdomen [12].

Its diagnosis should be made promptly before life-threatening complications develop [2,13].

The best method of confirming the diagnosis seems to be a CT scan but Doppler US imaging is an equally helpful modality [7,13].

De-torsion and splenopexy is the method of choice, whenever possible because, in addition to conserve spleen, prevent any future complications. When there is an infarction spleen, splenectomy is necessary [10].

So, optimal management requires a high level of suspicion, early diagnosis, and prompt surgical intervention, where preservation of the spleen is the goal.

Conflicts of interest

The authors declare that they do not have any personal conflicts of interest.

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Ethical approval

Being this present study a paper review, it is exempt from prior evaluation by the ethics committee.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request".

Author contribution

Study concept and design – Humberto Cristino MD*1 and Pedro Leão, MD, PhD, FACS1, 2, 3 Data collection and writing paper – Charlene Viana MD, MSc*1 and Carlos Veiga MD1 Data analysis and interpretation – all authors.

Guarantor

Pedro Leão, MD, PhD, FACS.

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