Radiology Case Reports 19 (2024) 2864–2867

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Volvulus of a wandering spleen in a pediatric patient ☆

Dolores Ferrara[a,*], *Gianfranco Vallone, MD*[b], *Silvana Russo, MD*[c], *Eugenio Rossi, MD*[d], *Ciro Riccio, MD*[e], *Andrea Maria Rinaldo, MD*[e], *Raffaele Zeccolini, MD*[f], *Massimo Zeccolini, MD*[a], *Francesco Esposito, MD*[g]

[a] *Department of Radiology, "A.O.R.N. Santobono-Pausilipon" Children Hospital, Naples, Italy*
[b] *Life and Health Department "V. Tiberio", University of Molise, Campobasso, Italy*
[c] *Pediatric Surgery Unit, "A.O.R.N. Santobono-Pausilipon" Children Hospital, Naples, Italy*
[d] *U.O.S.D. Diagnostic Imaging P.O. Pausilipon, "A.O.R.N. Santobono-Pausilipon" Children Hospital, Naples, Italy*
[e] *Department of Advanced Biomedical Sciences, University of Naples Federico II Unina, Naples, Italy*
[f] *Department of Precision Medicine, University of Campania "Luigi Vanvitelli", Naples, Italy*
[g] *U.O.S.D. Division of Emergency Imaging Department "A.O.R.N. Santobono-Pausilipon", Children Hospital, Naples, Italy*

ARTICLE INFO

Article history:
Received 11 February 2024
Revised 29 February 2024
Accepted 5 March 2024

Keywords:
Volvulus
Wandering spleen
Torsion
Ultrasound

ABSTRACT

Wandering spleen is a rare condition in children that is often caused by the loss or weakening of the splenic ligaments. Its clinical presentation is variable; 64% of children with wandering spleen have splenic torsion as a complication. A 13-year-old boy who had been showing abdominal pain in the hypogastric region accompanied by vomit and an enormous tumefaction in the suprapubic region came to our observation. Considering the ovoid morphology at ultrasound exam, the echostructure and the marked reduction of parenchymal vascularization, suspicion for torsion of an ectopic spleen arose. Ultrasound evaluation has a primary role in the diagnosis of a suspected wandering spleen and, to avoid potentially life-threatening complications, immediate surgery is often times required.

Introduction

Wandering spleen (WS) was first described by Van Horne, a Dutch physician, in 1667 on autopsy [1]. Wandering spleen, also referred to as aberrant, floating, ptotic, displaced, or prolapsed spleen, is characterized by excessive mobility of the spleen, which presents an elongated pedicle and is displaced from its usual position in the left upper quadrant. Embryologically absent or malformation of one or more suspensory liga-

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: dorianaferrara@hotmail.it (D. Ferrara).

https://doi.org/10.1016/j.radcr.2024.03.004

ments and the laxity of the abdominal musculature weakened by pregnancy are possible causes of this pathology [2].

Herein, we describe a case of wandering spleen in a 13 years old boy.

Case report

Coming to our observation at the Emergency Department was a 13-years-old boy who had been showing abdominal pain in the hypogastric region accompanied by vomit and an evident tumefaction in the suprapubic region for about 7 days.

During the objective examination, the patient presented a tractable abdomen that was painful on deep palpation with the evidence in the hypogastric region of a poorly mobile tumefaction in the deep planes extending from the pubic symphysis to the umbilical region with with overlying skin of regular appearance (Fig. 1).

Laboratory tests showed elevated values of CRP (125.58 mg/L; normal range: less than 0.3 mg/dL), increased direct bilirubin (1.12 mg/ dL; normal range: less than 0.3 mg/dL) and CK (236 U/L; normal range: 22 to 198 U/L). The remaining laboratory values were in the normal range.

At first a direct X-ray of the abdomen was performed and it showed only a few slightly gas distended intestinal loops in the left hypochondrium and an absent meteorism in the lower abdominal quadrants. Ultrasound examination was then performed, using My Lab Esaote convex and microconvex probes and it showed in the hypogastric area a homogeneous mass with no evidence of vascular flow on the color Doppler study (Fig. 2).

This formation mainly extended along the cranio-caudal plane and showed a maximum diameter of 20 cm and a clear plane of cleavage (Fig. 3).

Flowmetric examination showed post-stenotic resistance indices with tardus parvus pulse.

Considering the ovoid morphology, echostructure and poor parenchymal vascularization, suspicion for torsion of an ectopic spleen arose so the examination was extended to the remaining upper abdominal quadrants. An uninhabited splenic lodge was in fact then detected. We proceeded to identify the splenic vascular pedicle using the tail of the pancreas as a landmark. From the cranio-caudal scan, we observed the characteristic swirling appearance (Whirlpool sign) of the arterial and venous vessels at the hilum (video) with marked regional venous dilatation.

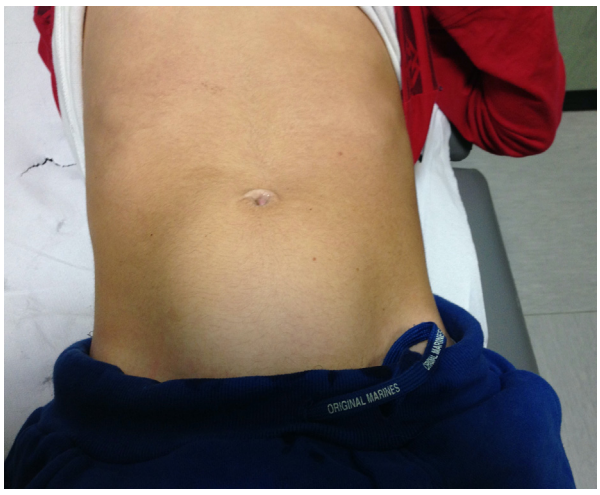

Fig. 1 – Voluminous abdominal swelling in hypogastric region.

On the basis of the ultrasound findings, the diagnostic hypothesis of wandering spleen volvulus was put forward, therefore the patient was urgently transferred to the Surgical Department and the necrotic spleen was removed after ligation of the hilar vessels by laparoscopic technique (Fig. 4).

Discussion

The spleen is an intraperitoneal organ attached to the stomach and posterior abdominal wall via the gastro-splenic ligament (which houses the splenic artery and vein and their respective lymphatic vessels) and the spleno-renal ligament [1]; it is supported inferiorly by the splenocolic and splenophrenic ligament and it comes into contact with the pancreatic tail via the splenopancreatic ligament. These peritoneal ligaments allow the spleen to be held in place in its homonymous lodge located in the left hypochondrium.

The absence or excessive laxity of the suspensory ligaments results in a condition of hypermobility of the spleen defined “ectopic” [2].

WS is a rare condition, congenital or acquired, in which the spleen migrates from the left upper quadrant to ectopic sites (most frequently the hypogastrium) [3].

The overall incidence is less than 0.02% with 2 incidence peaks: between 3 months and 10 years (more frequent in the male sex) and between 20 and 40 years (more frequent in the female sex) [4,5].

Congenital WS is mainly due to a lack of complete fusion of the mesogastrium during the second month of pregnancy, which may result in the absence or improper development of one or more ligaments causing splenic hypermobility.

Adult acquired WS occurs because of trauma, multiparity, connective tissue disorders, and hormonal changes after pregnancy while the pediatric-acquired form is associated with multiple conditions such as renal agenesis, immunoglobulin deficiency, infectious mononucleosis, malaria, Gaucher disease, Hodgkin’s lymphoma, and Di George syndrome [6].

The clinical presentation of WS varies from asymptomatic to an acute abdomen when its pedicle is twisted [7,8]. Most adult patients with WS are asymptomatic and this condition is revealed incidentally during a physical examination as a solid abdominal mass or by diagnostic examinations performed for other reasons.

In pediatric patients, the clinical presentation depends on the degree of torsion of its vascular pedicle: mild torsion may cause chronic abdominal pain secondary to splenic congestion; moderate-grade torsion results in intermittent pain subsequent to the process of torsion and detorsion of the vascular pedicle; at last, severe-grade torsion causes splenic infarction with the onset of a condition of acute abdominal pain [9].

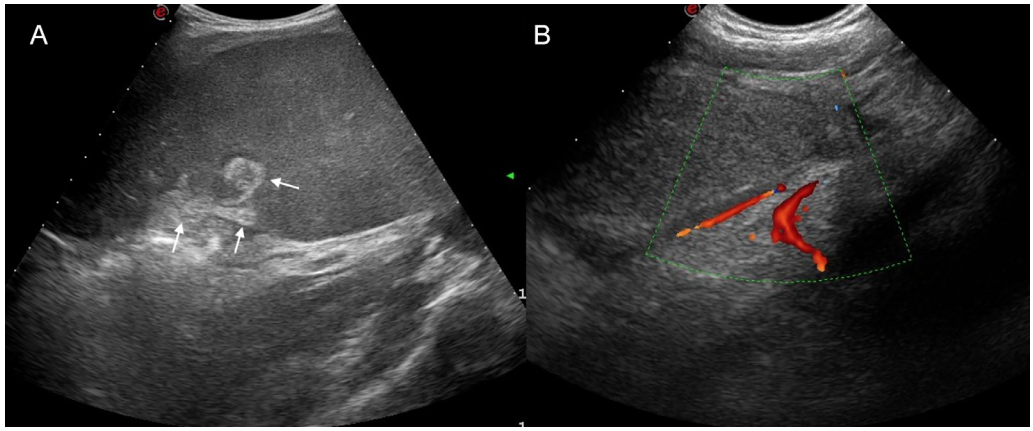

Fig. 2 – (A) Whirled and hyperechoic appearance of splenic vessels in the hilar region (arrows). (B) Nonhomogeneous spleen without vascular signals on color Doppler investigation.

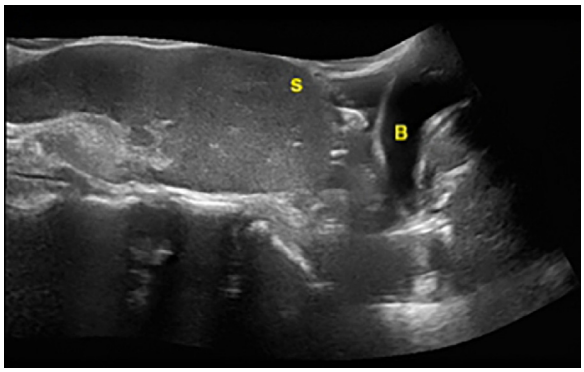

Fig. 3 – Enlarged and congested spleen with long axis oriented along the cranio-caudal plane lower pole is detectable in the supravescical area. S, spleen; B, bladder.

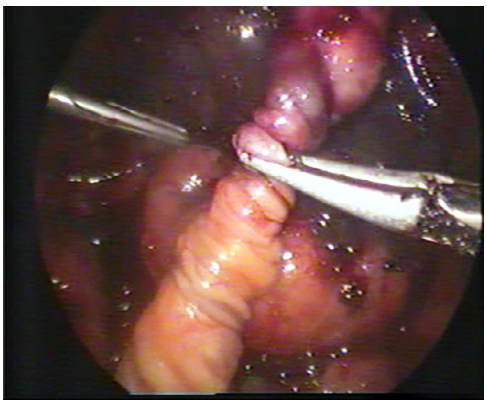

Fig. 4 – Laparoscopy. Splenic pedicle was rotated showing a characteristic Whirlpool sign.

The presence of an ectopic spleen should also be considered in children and young women who present episodes of recurrent pancreatitis with no other obvious cause [10]. This circumstance, which is very rare, often has a subclinical course as it results in localized inflammation in the pancreatic tail.

Sometimes laboratory tests are nonspecific, showing pancytopenia as a manifestation of hypersplenism or functional asplenia [11]. The first imaging technique for making diagnosis of a WS or accessory splenic nodule torsion is the ultrasound; additional contrast enhanced ultrasound (CEUS) increases sensitivity to allow diagnosis of vascular patency and parenchymal viability [12]. The spleen appears as a mass located in the lower abdominal quadrants or in the pelvic cavity with hypoechogenic or heterogeneous echo structure, instead of occupying its anatomical lodge. In addition to its ectopic location, the possible hypoperfusion and torsion of splenic vascular pedicle (Whirlpool sign) on color Doppler can be assessed. Venous congestion could lead to splenomegaly, which worsens the degree of pedicle torsion. CT abdomen with endovenous contrast medium injection is routinely performed as a second-level examination. Commonly it confirms the uninhabited splenic lodge with ectopic, megalic spleen, hypoperfusion and whirling appearance of splenic vessels which appear spontaneously hyperdense without postcontrast opacification. To guarantee the fair perfusion of the organ, the correct, rapid therapeutic management must be executed.

The main complications of a splenic torsion are compression of neighboring organs by splenomegaly, splenic infarction and/or rupture, thrombosis, and hemorrhage [13].

For patients with mild symptoms in the absence of complications, conservative treatment is preferred.

Instead, in case of complications, partial or total splenectomy by laparoscopic or laparotomic surgery is indicated [14].

Conclusions

Spleen torsion is a rare complication that can constitute a true surgical emergency given the risk of ischemia and necrosis of the organ. After clinical evaluation, the first imaging technique for making diagnosis is the ultrasound evaluation of the abdomen which shows an uninhabited splenic lodge, an ectopic spleen, the characteristic “Whirlpool sign” of splenic

vascular pedicle and reduced/absent hilar and parenchymal flow.

CEUS and CT with contrast medium are reserved for doubtful cases and proper surgical framing.

Early diagnosis can prevent the onset of complications such as parenchymal necrosis and rupture reducing morbidity and mortality associated with these complications.

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Patient consent

Informed consent was obtained from all individual participants included in the study.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2024.03.004.

REFERENCES

[1] Goyal RB, Gupta R, Prabhakar G, Praveen M, Tariq AM. Salvage splenopexy for torsion of wandering spleen in a child. APSP J Case Rep 2014;5(1):4.

[2] Ayaz UV, Dilli A, Ayaz S, Api A. Wandering spleen in a child with symptoms of acute abdomen: ultrasonographic diagnosis. Case report. Med Ultrasound 2012;14(1):64–6.

[3] Fonseca AZ, Ribeiro M, Contrucci O. Torsion of a Wandering Spleen treated with partial splenectomy and splenopexy. J Emerg Med 2013;44(1):e33–6.

[4] Ityel H, Granot Y, Vaknine H, Judich A, Shimonov M. Splenic torsion of a wandering spleen. Ist Med Assoc J 2011;13(2):119–20.

[5] 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(7):1446–9.

[6] Hedeshian MH, Hirsh MP, Danielson PD. Laparoscopic splenopexy of a pediatric wandering spleen by creation of a retroperitoneal pocket. J Laparoendosc Adv Surg Tech A 2005;15(6):670–2.

[7] Karmzyn B, Steinberg R, Gayer G, Grozovski S, Freud E, Kornreich L. Wandering spleen-the challenge of ultrasound diagnosis: report of 7 cases. Clin Ultrasound 2005;33(9):433–8.

[8] Chen JW, Yeh DM, Peng SH, Chen GS, Tseng YH, Lin CW, et al. Sonographic diagnosis of a subclinical wandering spleen: role of the decubitus position. J Ultrasound Med 2012;31(3):483–7.

[9] Karakas HM, Tuncblick N, Okten OO. Splenic abnormalities: an overview on sectional images. Diag Interv Radiol 2005;11:152–8.

[10] Varga I, Galfiova P, Adamkov M, Danisovic I, Polak S, Kubikova E. Congenital anomalies of the spleen from an embryological point of view. Med Sci Monit 2009;15(12):RA269–76.

[11] Gayer G, Zissin AS, Atar E, Portnoy O, Iezchak Y. CT findings in congenital anomalies of the spleen. Br J Radiol 2001;74:767–72.

[12] Aguirre Pascal E, Fontanilla T, Pérez I, Muñoz B, Carmona MS, Minaya J. Wandering spleen torsion—use of contrast-enhanced ultrasound. BJR Case Rep 2017;3(1):20150342.

[13] Soleimani M, Mehrabi A, Kashfi A, Fonnouni H, Buchler MW, Kraus TW. Surgical treatment of patients with wandering spleen: report of six cases with a review of the literature. Surg Today 2007;37:261–9.

[14] Sty JR, Conway JJ. The spleen: development and functional evaluation. Semin Nuel Med 1985;15:276–98.