Radiology Case Reports 14 (2019) 1209–1213

Available online at www.sciencedirect.com

ScienceDirect

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

Case Report

Wandering spleen with splenic torsion in a child with DiGeorge syndrome

Charlotte S. Taylor, MD, Candace Howard-Claudio, MD, PhD*

Department of Radiology, University of Mississippi Medical Center, 2500 N. State Street, Jackson, MS 39216

ARTICLE INFO

Article history:
Received 15 May 2019
Revised 21 May 2019
Accepted 27 June 2019
Available online 6 August 2019

Keywords:
Wandering spleen
Splenic torsion
Splenic infarct
DiGeorge syndrome
Renal agenesis

ABSTRACT

Wandering spleen is a rare condition, occurring due to either abnormal development of or abnormal laxity of suspensory ligaments. The hypermobility of the spleen predisposes these patients to splenic torsion, which may be a life-threatening complication. The clinical presentation of wandering spleen varies widely from vague pain to an acute abdomen. There are numerous case reports of other congenital anomalies in children with a wandering spleen. We present a case of wandering spleen with splenic torsion in a child with DiGeorge syndrome, which to our knowledge has not been previously reported.

Introduction

Wandering spleen is a rare condition, accounting for less than 0.2% of splenectomies [1,2]. It occurs due to either abnormal development of or abnormal laxity of suspensory ligaments. The hypermobility of the spleen predisposes these patients to splenic torsion, which may be a life-threatening complication. Due to the rarity of this condition and a wide range of clinical presentations, from vague abdominal pain to an acute abdomen, wandering spleen may be initially misdiagnosed as another condition. We present a case of wandering spleen with splenic torsion in a child with DiGeorge syndrome, which to our knowledge has not been previously reported.

Case report

A 7-year-old boy who is nonverbal with DiGeorge syndrome, tetralogy of Fallot (status postrepair), and a single dysplastic kidney with stage III chronic kidney disease presented to an outside emergency department. He was febrile to 100.5°F and had left-sided abdominal pain and guarding. He had a recent laparoscopic appendectomy 1 week prior performed for severe right-sided abdominal pain and the presence of appendicoliths. Pathology returned negative for appendicitis. Computed tomography (CT) of the abdomen was performed and showed geographic areas of hypodensity and stranding of the involving the spleen. Clinicians were initially

Funding: Candace M Howard is supported by the National Institute of General Medical Sciences of the National Institutes of Health under award number 5U54GM115428. The content is solely the responsibility of the authors and does not necessarily represent the official views of the National Institutes of Health.

* Corresponding author.
E-mail address: cstaylor2@umc.edu (C.S. Taylor).

https://doi.org/10.1016/j.radcr.2019.06.018

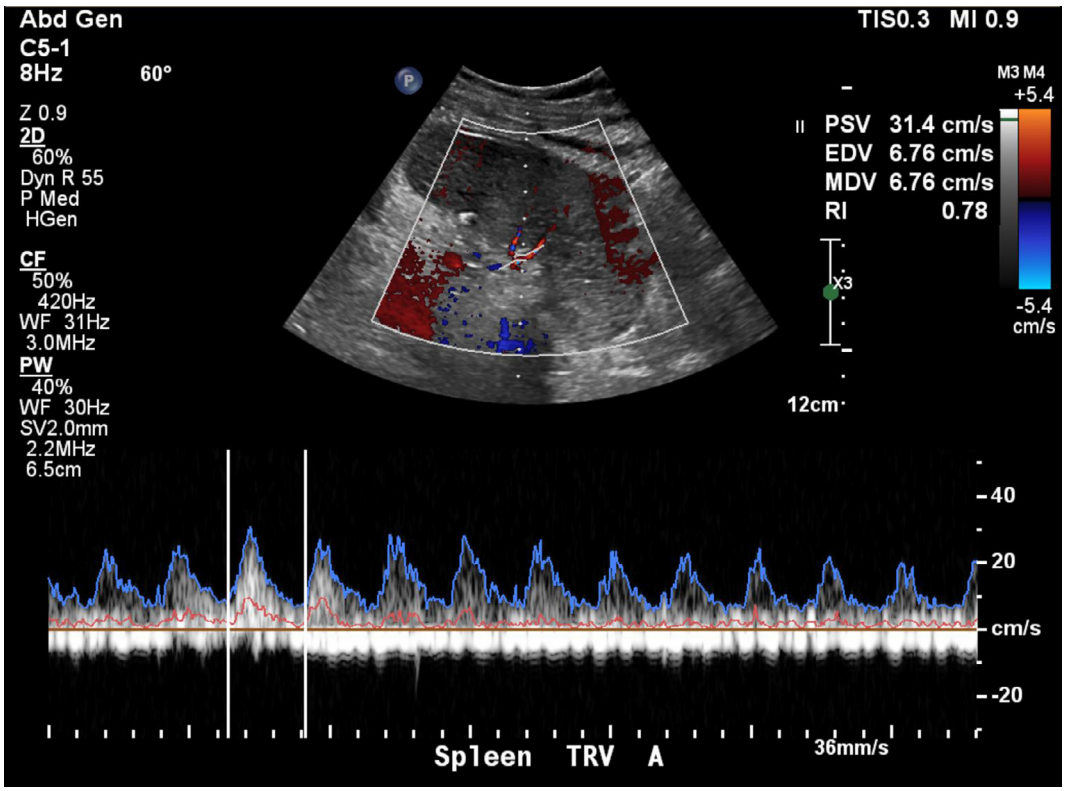

Fig. 1 – Ultrasound of the spleen demonstrates heterogeneous internal echotexture with preserved parenchymal Doppler flow and splenic artery spectral arterial waveforms.

concerned for infection given the patient's immunocompromised state with his DiGeorge syndrome. He was transferred to our pediatric emergency department. Laboratory studies showed a white-cell count of 16,500 mm^3 (reference range, 4300-11,000), hemoglobin level of 6.9 g/dL (reference range, 10.7-13.4), platelet count of 271,000 mm^3 (reference range, 206,000-369,000), and lipase of greater than 5000 units per liter (reference range, 10-175).

Ultrasound of the abdomen performed for pancreatitis workup showed cholelithiasis and biliary sludge. Concurrent ultrasound of the spleen showed heterogeneous echogenicity with preserved parenchymal flow and arterial waveforms on Doppler (Fig. 1). Magnetic resonance imaging of the abdomen revealed abnormal location of the spleen in the left mid abdomen with multiple splenic infarcts. A chest CT from 5 years prior showed normal sub-diaphragmatic location of the spleen (Fig. 2). Also, his spleen was noted to be in a different location on multiple scans (Fig. 3). Thus a diagnosis was made of wandering spleen with splenic torsion.

He was treated preoperatively with intravenous hydration, blood transfusion, and pain control. A preoperative echocardiogram showed an increased gradient in his right ventricle to pulmonary artery conduit, and he underwent ballooning of his conduit prior to surgery. Laparoscopic splenectomy and cholecystectomy was performed the following day, and splenic torsion was confirmed intraoperatively. He improved dramatically postoperatively and was discharged home on penicillin for endocarditis prophylaxis. He was also scheduled for outpatient postsplenectomy vaccinations. At most recent follow-up 1 month after surgery, he was doing well without recurrent abdominal pain or infection.

Discussion

Wandering spleen is a rare entity that primarily occurs in children and young women from ages 20-40 years old [3]. The spleen is abnormally hypermobile, which may be congenital or acquired. A congenitally hypermobile spleen occurs due to maldevelopment of the dorsal mesogastrium, which gives rise to the gastrosplenic and splenorenal ligaments that normally fix the spleen in the splenic fossa. This results in an abnormally long vascular pedicle. Acquired forms occur due to abnormal laxity of the normal gastrosplenic and splenorenal ligaments, which occurs in multiparous women due to hormonal influences in pregnancy. The congenital form is the most common cause in children, and there are multiple reported cases of wandering spleen occurring in patients with other congenital developmental or fixation abnormalities. This includes concurrent renal agenesis, which was present in our patient [4]. While this has not been directly studied, this suggests an association of other congenital anomalies with wandering spleen. There are no previously reported cases of wandering spleen in a patient with DiGeorge syndrome. However, there is a known association of renal agenesis, among

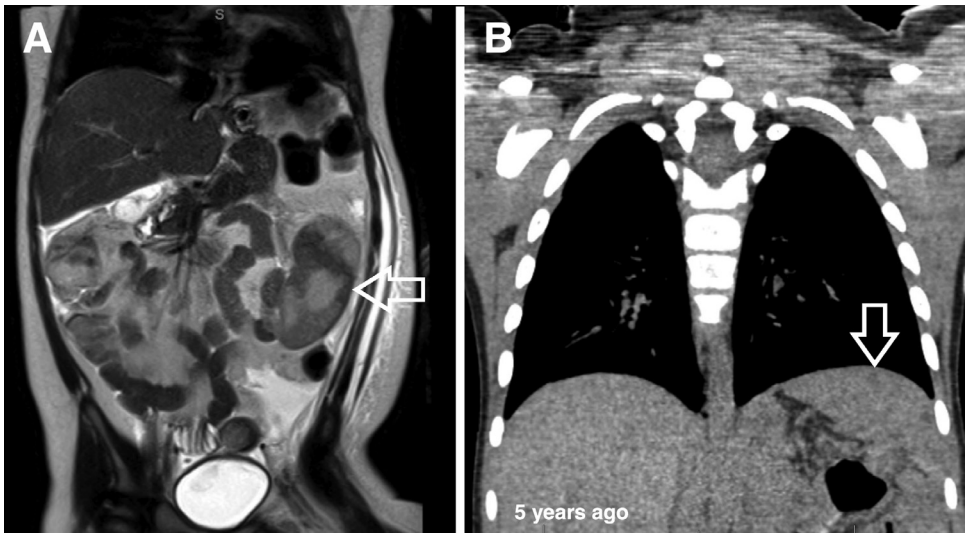

Fig. 2 – Coronal T2 weighted MRI (A) shows abnormal location of the spleen in the left mid abdomen with multiple splenic infarcts. Coronal chest CT (B) from 5 years prior shows the spleen in normal sub-diaphragmatic location.

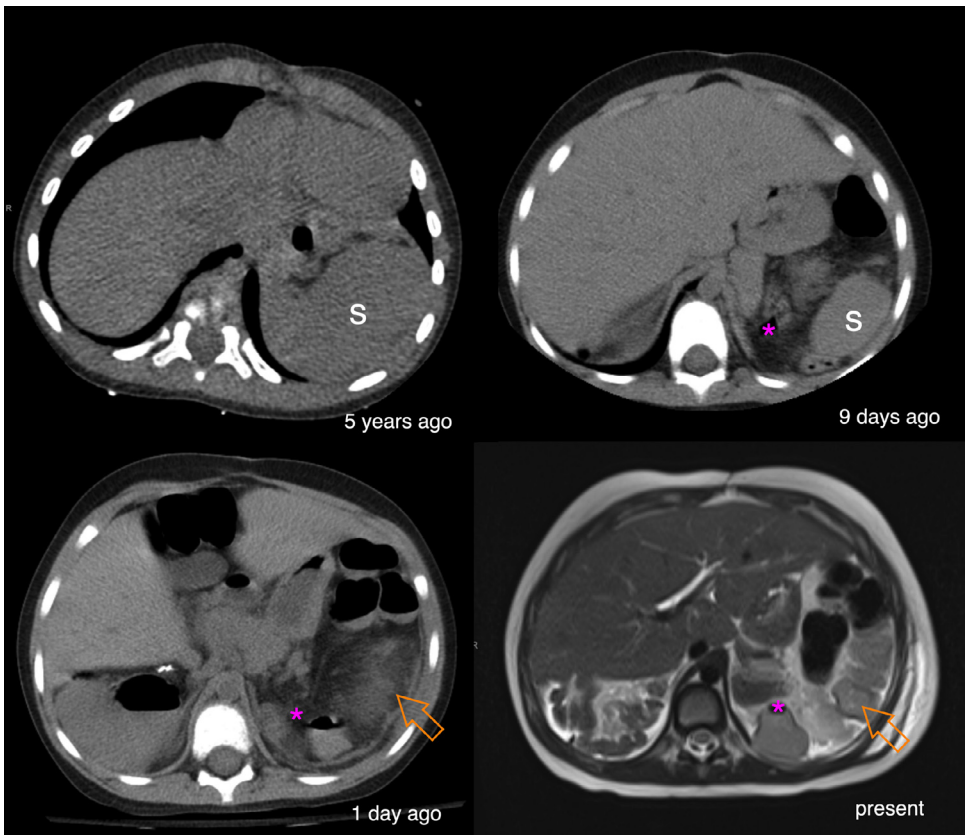

Fig. 3 – Axial CT through the abdomen without contrast (A-C) and axial T2 weighted MRI of the abdomen (D) at different time-points. The spleen (s) is in the expected anatomic location in the left upper quadrant but in slightly different locations 9 days and 5 years prior (A,B). Imaging after the patient's initial laparoscopic surgery 1 day prior and present day (C,D) show an empty splenic fossa (orange arrows). Left kidney is absent from the left renal fossa (*).

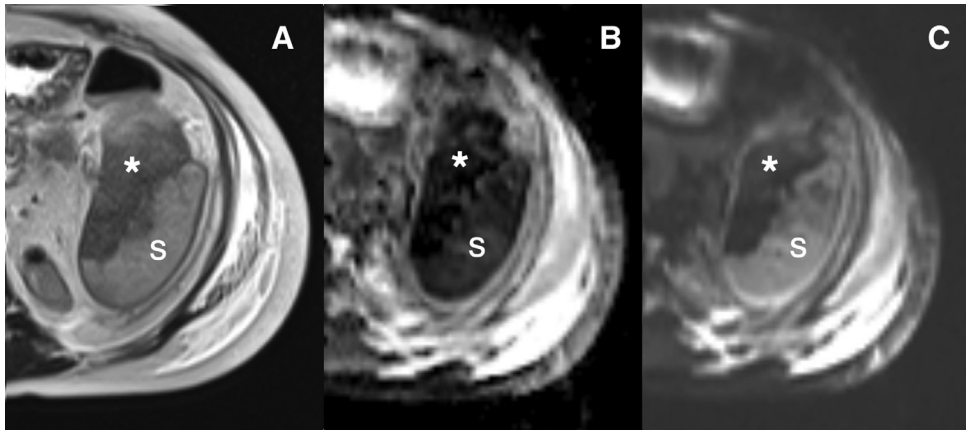

Fig. 4 – Splenic infarcts on MRI. Axial T2 weighted (A), ADC map (B), and DWI (C) sequences at the level of the mid-abdomen. The normal viable splenic tissue (s) is mildly T2 hyperintense and mildly restricts diffusion, similar in signal as nodal tissue. Infarcted spleen (*) is hypointense on all 3 sequences. The normal restricted diffusion of the spleen should not be confused as infection or other pathology.

other urogenital anomalies, with DiGeorge syndrome [5]. We hypothesize that our patient's underlying genetic defect gave rise to renal agenesis, which may have some association with wandering spleen.

When symptomatic, the most common presenting signs and symptoms of wandering spleen in children are abdominal pain and palpable mass. When complicated by splenic torsion, symptoms vary based on the degree of torsion. Mild torsion may present as chronic abdominal pain due to congestion. Moderate torsion will present with intermittent severe pain due to intermittent torsion and detorsion. Severe torsion results in splenic infarcts and will present as an acute abdomen. In our patient, his abdominal pain that lead to his appendectomy was likely a manifestation of intermittent splenic torsion. The insufflation of his abdomen with his laparoscopic appendectomy allowed his wandering spleen more freedom to further twist on its pedicle, leading to infarction and a more severe presentation as an acute abdomen 1 week later.

Clinical presentation in these patients can be confusing, and along with the rarity of this condition, wandering spleen can be misdiagnosed without a strong degree of clinical suspicion. Our patient's underlying immunodeficiency, leukocytosis, fever, and recent surgery initially created a high clinical suspicion for an infection. Our patient's elevated lipase and presence of gallstones on ultrasound was also a confounder in the clinical workup. Presentation of wandering spleen as recurrent pancreatitis has been reported and is likely due to ischemia in the tail of the pancreas [6,7]. In our patient with cholelithiasis, gallstone pancreatitis remained in the differential diagnosis, and cholecystectomy was performed along with splenectomy. Fever and leukocytosis have also been reported in splenic torsion [4]. Another unusual presentation of wandering spleen is bleeding diathesis due to hypersplenism and thrombocytopenia [8,9].

Diagnosis of wandering spleen is primarily an imaging diagnosis. The ectopic location of the spleen, especially if in a different location over multiple imaging studies, is diagnostic of a wandering spleen. The presence of splenic infarcts on CT or MRI, as in our case, should raise suspicion for torsion. The classic whorled appearance of splenic vessels seen on CT cinches the diagnosis of splenic torsion. On MRI, it is important to recognize that the spleen restricts diffusion normally, similar to nodal tissue (Fig. 4). This should not be confused for infection or other pathology. Ultrasound is the initial imaging study of choice in children and may suggest the diagnosis of wandering spleen if the spleen is noted to be in an abnormal location. A sulfur colloid liver-spleen scan can also be diagnostic, showing an ectopic location of the spleen [4]. Splenic infarcts will manifest as photopenic areas [10]. A lack of parenchymal blood flow or loss of arterial waveforms on Doppler indicates severe torsion and vascular compromise. In our case, arterial flow was preserved at the time of ultrasound, likely due to intermittent or incomplete torsion.

Treatment of a wandering spleen is surgical, either with splenopexy or splenectomy. Splenopexy is preferred for a viable spleen to preserve immune function, and splenectomy is reserved for cases of an infarcted spleen. Vaccinations for encapsulated organisms (*H. influenzae,* meningococcal, and pneumococcal vaccines) should be given after a splenectomy.

In conclusion, wandering spleen is a rare cause of abdominal pain, and it can lead to potentially life-threatening complications. Radiologists should be aware of this diagnosis, which will primarily be made on imaging studies. Review of previous studies, if available, is tremendously helpful in making a confident diagnosis of wandering spleen. If splenic infarcts are present, the possibility of splenic torsion should be considered. While not directly studied, certain congenital anomalies may have an association with a wandering spleen. Future research in this area is needed.

Declaration of Competing Interest

Dr. Howard Claudio receives funding from a grant from the National Institute of Health. The authors have no other financial or personal conflicts of interest.

REFERENCES

[1] Whipple HO. The medical-surgical splenopathies. Bull NY Acad Med 1939;15:174–6.
[2] Mayo WJ. A review of 500 splenectomies with special reference to mortality and end results. Ann Surg 1928;88(3):409–15.
[3] Gayer G, Zissin R, Apter S. CT findings in congenital anomalies of the spleen. Br J Radiol 2001;74:767–72.
[4] Brown CW, Virgilio GR, Vazquez WD. Wandering spleen and its complication in children: a case series and review of the literature. J Pediatr Surg 2003;38:1676–9.
[5] Lopez-Rivera E, Liu YP, Verbitsky M, Anderson BR, Capone VP, Otto EA, et al. Genetic drivers of kidney defects in the DiGeorge syndrome. N Engl J Med 2017;376(8):742–54.
[6] Lebron R, Self M, Mangram A, Dunn E. Wandering spleen presenting as recurrent pancreatitis. JSLS 2008;12(3):310–13.
[7] Gilman RS, Thomas RL. Wandering spleen presenting as acute pancreatitis in pregnancy. Obstet Gynecol 2003;101(5 pt 2):1100–2.
[8] Benoist S, Imbaud P, Veyrieres M. Reversible hypersplenism after splenopexy for wandering spleen. Hepatogastroenterology 1998;45(24):2430–1.
[9] Moll S, Igelhart JD, Ortel TL. Thrombocytopenia in association with a wandering spleen. Am J Hematol 1996;53(4):259–63.
[10] Chin JK, McCormick PA, Hilson AJ, Burroughs AK, McIntyre N. Liver/spleen scintigraphy for diagnosis of splenic infarction in cirrhotic patients. Postgrad Med J 1993;69(815):715–17.