Egyptian Journal of Surgery Vol 27, No 3, July, 2008

CASE REPORT

POLYSPLENIA IN ADULT PATIENT WITH WANDERING PELVIC AND TORSION SPLEEN

By
Abdulhakim Al-tamimi,[1] Gamal Khudabakhash,[1] Naser Al-Gadasi[2]
[1]Department of surgery, Faculty of Medicine and Health Science, University of Aden, [2]Department of surgery, Faculty of Medicine and Health Science, University of Sana'a, Yemen.

Correspondence to: Abdulhakim Al-tamimi, Email: abotammam1111@yahoo.com

Aim: *To present a rare case of congenital polysplenia in adult patient present with sudden abdominal pain due to twisted wondering spleen in the pelvis.*
Methods: *This is a single case report investigated preoperative and examined clinically where surgical intervention was performed due to rapidly progressive enlarged hypogastric mass.*
Result: *Adult man with polysplenia and minor congenital heart disease presented with twisted accessory spleen wondering in the pelvis with long pedicle.*
Discussion: *Polysplenia syndrome (PSS) is a type of situs ambiguous,usually associated with severe congenital heart diseases and rarely these patient reached adult life. The unique sign of the polysplenia is the presence of multiple spleens, ranging from 2 to 16. We present a case of polysplenia in adult patient with minor heart anomalies and four spleens one of them is very huge and discovered as emergency with torsion wondering pelvic spleen.*

Keywords: *Situs ambiguous, Heterogenous disease, Accessory spleen.*

INTRODUCTION

Polysplenia is a heterogeneous disease that primarily affects the asymmetric organs, including the heart, lungs and bronchi, liver, intestines, and spleen.(1)

The exact cause of polysplenia has not been defined, but it appears to be multifactorial, with some familial predisposition.(2)

Polysplenia is present in less than 0.5% of individuals, usually asymptomatic and located in the left upper quadrant, commonly near the splenic hilum with the pelvic and retroperitoneal location being the rarest sites.(3,4)

Polysplenia is present from birth, although clinical detection may be delayed, depending on the severity of congenital heart disease and gastrointestinal abnormalities.

Wandering pelvic spleen, also known as mobile spleen or ptosed spleen is a rare clinical condition.(5)

THE CASE REPORT

A 26 year healthy male presented with a feeling of heaviness, mild pain and progressively enlarged swelling in the hypogastric region (Fig. 1), no fever or loss of weight. The chest was deformed with clear pigeon shape, no dextrocardia or abnormal heart sounds and mild atrial septal defect (ASD) detected by echocardiography.

On abdominal examination a mobile, visible, firm mass

was felt in the hypogastric region with smooth surface and tenderness on deep palpation, not pulsated and dull on percussion.

Routine hematological investigations were normal. Electrocardiography and echocardiography except ASD otherwise normal.

Ultrasonography of the abdomen revealed hypoechoic mass in the hypogastic region separated from the urinary bladder and compressing the pelvic colon with clear vasculatures (Fig. 2). Kidneys, liver and gallbladder were normal, and there is a small spleen in the left upper quadrant. A CT scan of the abdomen was done on a multislice CT scanner which showed a 12x8 cm hyperdense intensely enhancing abdominal mass lying in the pelvis up to the pelvic brim with normally located small spleen and other small splenules (Fig. 3).

The patient was operated through lower midline incision, big spleen with long twisted pedicle was found in the pelvic cavity extend cephalic above the pelvic brim without splenic notch (Fig. 4), spleenectomy was done and the left upper quadrant was palpated and found two small splenic nodules near the normally located spleen which were removed (Figs. 5,6).

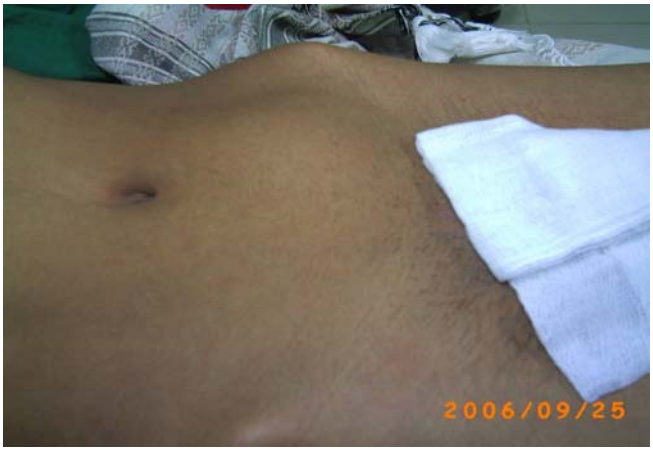

Fig 1

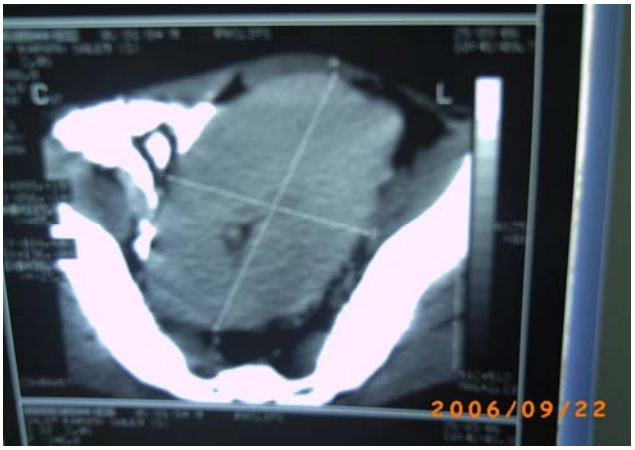

Fig 3

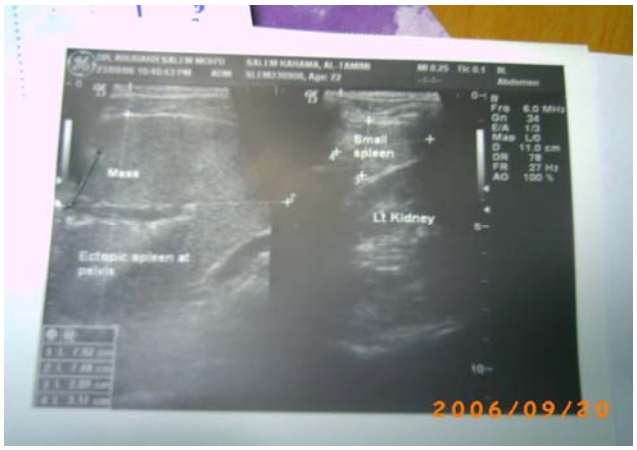

Fig 2

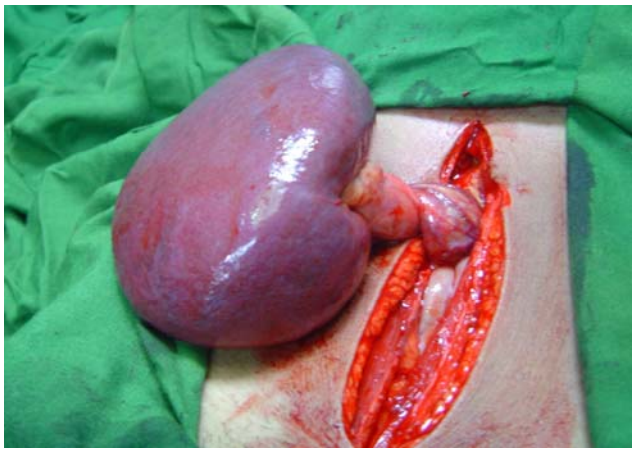

Fig 4

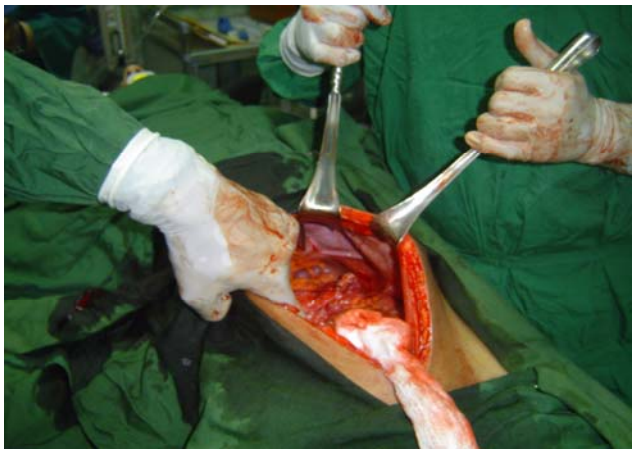

Fig 5

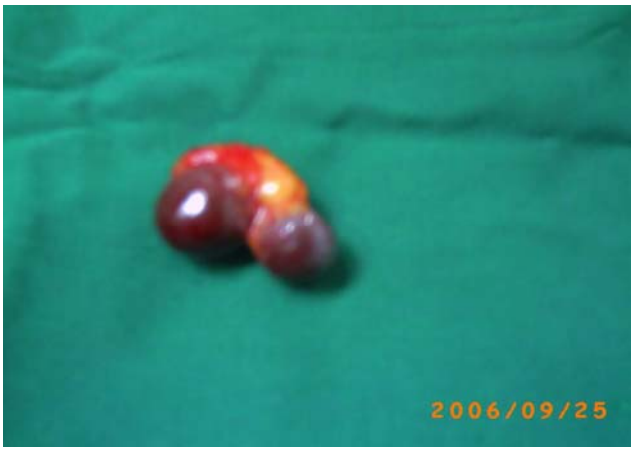

Fig 6

DISCUSSION

Helwig is credited with describing the Heterotaxia(polysplenia) syndrome in 1929.[6]

The polysplenia syndrome (PSS) is a type of situs ambiguous characterized by left isomerism, conformed by a group of visceral anomalies of unknown etiology,the presence of multiple aberrant splenic nodules and wide range of organic malformations may exist.[7,8] The unique sign of the polysplenia is the presence of multiple spleens, ranging from 2 to 16 - four in this case. This congenital anomaly is rarely encountered in the elderly age (about 2.5:100 000 live births but only 5% of patients survive beyond 5 years of life).[9,10]

This syndrome is generally diagnosed in the newborn or infant period, because the majority of patients have severe congenital cardiac anomalies. Most of these patients have severe cyanosis and most of them die before the age of 2 years. Less than 5% of patients survive beyond 5 years and these patients can be incidentally detected on routine radiological examination.[11]

Pelvic spleen or splenia ectopica is a rare condition that often presents as an adnexal mass in females with the definitive diagnosis being made intraoperatively.

Wandering accessory spleens may mimic tumors, such as pancreatic tumor, adnexal tumor, abdominal tumor, retroperitoneal tumor, adrenal tumor, or testicular tumor, according to its location.[8,12] Pelvic spleen may also present with recurrent pain and a pelvic mass. The enlarged mass may present with symptoms of bladder compression and torsion is a recognized complication of this condition.

The major complication of a wandering spleen is acute, chronic or intermittent torsion caused by its increased mobility. Symptoms and signs of splenic torsion are notoriously variable: chronic abdominal discomfort probably due to splenic congestion or ligamentous pressure, intermittent pain presumably due to spontaneous torsion and detorsion, and less often severe abdominal pain from acute torsion and infarction, which produce marked congestion and capsular stretching.[13,14,15] Physical examination may demonstrate a tender mass.[14]

Accessory spleens are usually smaller than 3 cm. However, in this patient it is larger than 10 cm and surgery was indicated when there are symptoms, such as pain, rupture, infarction, or vascular torsion.[16]

REFERENCES

1. Abell, Irvin. Wandering spleen with torsion of the pedicle. Ann. Surg. 1938;98:722-32.

2. Adkins EH. Ptosed spleen with torsion of pedicle. Ann. Surg. 1938;107:832-5.

3. Kato Y, Murayama K, Taniguchi N, Yamaguchi S, Hashimoto H, Kaneko S, et al. A case of accessory spleen presenting as retroperitoneal tumor. Hinyokika Kiyo 1998;44:711-4.

4. Churei H, Inoue H, Nakajo M. Intrapancreatic accessory spleen: case report. Abdom Imaging. 1998;23:191-3.

5. Berg C, Geipel A, Kamil D. The syndrome of left isomerism: sonographic findings and outcome in prenatally diagnosed cases. J Ultrasound Med. 2005;24:921-31.

6. Chen SJ, Li YW, Wang JK. Usefulness of electron beam computed tomography in children with heterotaxy syndrome. Am J Cardiol. 1998;81:188-94.

7. Ditchfield MR, Hutson JM. Intestinal rotational abnormalities in polysplenia and asplenia syndromes. Pediatr Radiol. 1998;28:303-6.

8. Gayer G, Apter S, Jonas T. Polysplenia syndrome detected in adulthood: report of eight cases and review of the literature. Abdom Imaging. 1999;24:178-84.

9. Winer-Muram HT. Adult presentation of heterotaxic syndromes and related complexes. J Thorac Imaging. 1995;10:43–57.

10. Rose V, Izukawa T, Moes CA. Syndromes of asplenia and polysplenia: a review of cardiac and non-cardiac malformations in 60 cases with special reference to diagnosis and prognosis. Br Heart J. 1975;37:840–52.

11. Peoples WM, Moller JH, Edwards JE. Polysplenia: a review of 146 cases. Pediatr Cardiol. 1983;4:129–37.

12. Gilljam T, McCrindle BW, Smallhorn JF. Outcomes of left atrial isomerism over a 28-year period at a single institution. J Am Coll Cardiol. 2000;36:908-16.

13. Sty JR, Conway JJ. The spleen: development and functional evaluation. Semin Nucl Med. 1985;15:276–98.

14. Raissaki M, Prassopoulos P, Daskalogiannaki M, Magkanas E, Gourtsoyiannis N. Acute abdomen due to torsion of wandering spleen: CT diagnosis. Eur Radiol. 1998;8:1409–12.

15. Swischuk LE, Williams JB, John SD. Torsion of wandering spleen: the whorled appearance of the splenic pedicle on CT. Pediatr Radiol. 1993;23:476–7.

16. Hofstaetter C, Plath H, Hansmann M. Prenatal diagnosis of abnormalities of the fetal venous system. Ultrasound Obstet Gynecol. 2000;15:231-41.