Case Report

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Complicated congenital splenic cyst: Saved by a splenunculus

Nina Karia, Kokila Lakhoo

ABSTRACT

A 12-year-old girl presented with a large congenital splenic cyst complicated by *Salmonella* organisms. After failure of conservative management and percutaneous drainage, a splenectomy was performed. An incidental splenunculus was preserved. On follow up the splenunculus had increased to normal splenic size and there was no evidence of Howell-Jolly bodies, suggesting normal splenic function.

Key words: *Salmonella*, splenectomy, splenic cyst, splenunculus

INTRODUCTION

Congenital splenic cysts are rare in children; however, if unrecognised the disease may carry high mortality of up to 100%.[1] A splenic cyst can be divided into primary or secondary depending on the presence of epithelial lining or not respectively (based on Fowler's[2] and subsequently Martin's classification[3]). Primary splenic cysts can be further sub-divided into parasitic and non-parasitic. Congenital cysts are lined with mesothelium[4] and are classified within non-parasitic, primary splenic cysts.

Congenital cysts are more commonly noted in girls,[5] are usually asymptomatic and diagnosed incidentally by imaging. However symptoms may develop once complicated by infection, haemorrhage and trauma. Infection is particularly important in patients who are immunocompromised.[1]

Department of Paediatric Surgery, Children's Hospital Oxford, Oxford UK

Address for correspondence:
Dr. Kokila Lakhoo,
Department of Paediatric Surgery, Level 2 Children's Hospital Oxford, Headley way Oxford, OX39DU United Kingdom.
E-mail: Kokila.lakhoo@paediatrics.ox.ac.uk

CASE REPORT

Our case focuses on a 12–year-old girl who presented to a district general hospital with a vague 1 week history of vomiting, upper abdominal pain radiating to the LEFT shoulder, pyrexia and fluctuating GCS (Glasgow Coma Scale). Three weeks prior to this presentation she had been on holiday abroad where she had a brief period of watery diarrhoea, abdominal pain and high temperatures which temporarily resolved. She had no significant past medical history of note and her vaccinations were up to date. On arrival to the tertiary centre she was noted to have a fluctuating GCS ranging between 9 and 13, BP 101/ 29, HR 180, RR 50, and a temperature of 41°C. Palpation of the abdomen revealed a tender left upper quadrant with moderate hepatosplenomegaly and no evidence of peritonism.

On further examination there was decreased air entry in the left lung base, a non-radiating 3/6 soft systolic murmur over the mitral region and no peripheral stigmata of infective endocarditis, focal neurology, rashes or features of meningism. Laboratory investigations revealed a haemoglobin of 9.8, platelet count of 20 and deranged clotting. Her LFT and WCC were normal but her CRP was raised to >320 and she had a metabolic acidosis with a bicarbonate value of 16.4 and a base excess of -7.9. The chest radiograph revealed a left lower lobe collapse and an urgent ultrasound scan confirmed a large splenic collection mainly in the upper two thirds of the spleen with some preservation of splenic tissue in the lower pole. [Figure 1]. A percutaneous drain was inserted under ultrasound guidance and produced 2.7 L of infected fluid. Cyst fluid, blood and stool cultures confirmed *Salmonella enteritidis* and she was started on ceftriaxone, gentamicin and metronidazole as per drug sensitivities. Despite treatment she remained pyrexial and a CT scan was performed [Figure 2].

The CT scan revealed a 13 × 10 cm collection with

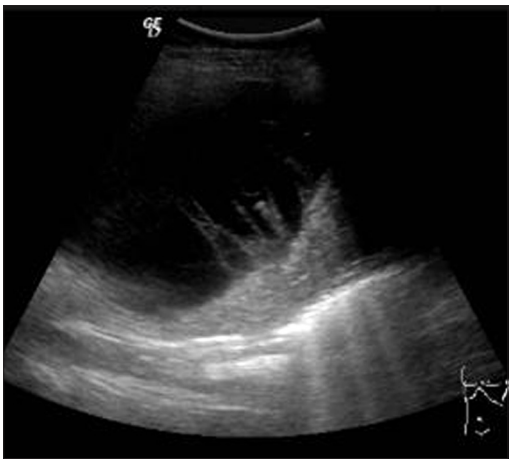


Figure 1: US of spleen showing large cyst with septae.

thick septations not amenable to further radiological drainage. In view of this finding an exploratory laparotomy was performed.

Laparotomy findings revealed a large multiseptated cyst in the upper pole of the spleen. The anterior wall of the cyst was de-roofed and 2 L of infected fluid was drained and a large drain left *in situ*.

Postoperative recovery was uneventful. There was significant clinical improvement which correlated to laboratory markers and subsequent negative cultures.

The patient was discharged on oral ciprofloxacin. However, at 3 week follow up a repeat US scan revealed a residual collection measuring 2.8 × 1.5 × 1.6 cm with total splenic destruction. The decision was taken to perform a total splenectomy. At surgery the destroyed splenic tissue and the residual cyst was excised but a splenunculus noted incidentally was spared. Histological appearance confirmed a congenital

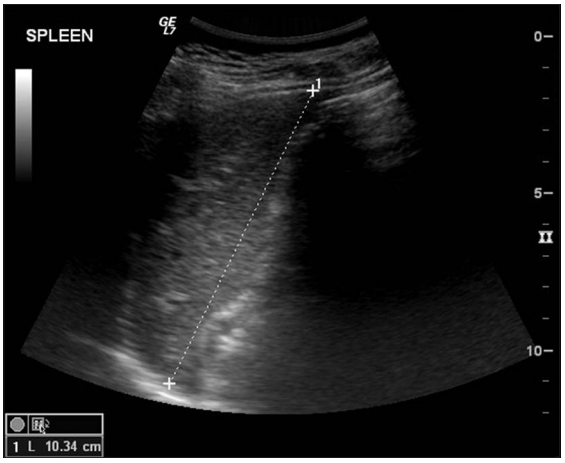


Figure 3: Ultrasound scan 9 month post-splenectomy shows enlargement of the splenunculus to a normal splenic size.



Figure 2: CT abdomen showing a large splenic cyst with some splenic preservation in the lower pole.

epithelial inclusion cyst of the spleen complicated by a secondary infection.

The patient was placed on long-term prophylaxis with Penicillin V for protection from pneumococcal infection. Regular follow-up with ultrasound scan over a 9 month period revealed gradual enlargement of splenunculus to a normal splenic size [Figure 3] and absence of Howell Jolly bodies on blood film. The patient was subsequently taken off penicillin prophylaxis after consultation with our haematology colleague.

DISCUSSION

Congenital splenic cysts are uncommon and one complicated by secondary *Salmonella* infection appears to be a rare phenomenon. Hepatic, biliary and splenic involvements are the most common extraintestinal sites for Salmonella species.[6] If splenic involvement does occur, this appears to occur early in the natural progression.[7] There are often predisposing conditions such as sickle cell which occurs in a third of cases[8] and patients who are immunocompromised. The underlying

pathology in this case may potentially be explained as a result of bacterial seeding via haematogenous spread during transient bacteraemia,[9] whereby the underlying asymptomatic congenital cyst acted as a predisposing factor.

Salmonella, particularly *typhi* species, can initially present following ingestion with a self-limiting diarrhoeal illness as the bacteria penetrates gastro-intestinal lymphoid mucosa. During a 10-14 day incubation period it can often be asymptomatic[9] before presenting with the typical symptoms which appears to correlate to our case above. During the first 10 days stool cultures are often negative and diagnosis can only be made by positive blood cultures. Splenic abscess typically presents with abdominal pain, fevers, splenomegaly, cough and left-sided consolidation and pleural effusions.

Leucocytosis is typically associated;[10] however, in our case the WCC was lower than expected in correlation to other inflammatory markers due to the severity of sepsis on presentation. The anaemia and thrombocytopenia may be as a result of DIC secondary to sepsis or splenomegaly resulting in sequestration. Ultrasound and CT scanning appears to be the gold standard to diagnose splenic cysts; however, they poorly define aetiology and the classification of cyst. Septations in a cyst imply secondary haemorrhage or infection.

The rationale behind conservative management with therapeutic percutaneous drainage in attempt to preserve splenic function is pertinent particularly in children as the spleen plays an immunological role and defends against encapsulated organisms. The problem still remains that percutaneous drainage appears to be a temporary measure as enlargement, rupture or re-infection remains a high possibility.[9] If these measures fail to eradicate infection performing a partial or total splenectomy at this stage has not shown an increase in mortality. In our case partial splenectomy was not an option due to the complete destruction of the normal splenic tissue.

Fortunately in this case study the option of preserving splenunculus has resulted in normal immunological functioning spleen whilst eradicating the diseased spleen.

REFERENCES

1. Nelken N, Ignatius J, Skinner M. Changing clinical spectrum of splenic abscess. A multicenter study and review of literature. Am J Surg 1987;154:27-34.
2. Fowler RH. Cystic tumors of spleen. Int Abstr Surg 1940;70:213-23.
3. Martin JW. Congenital Splenic cysts. Am J Surg1958;96:302-8.
4. Mirilas P, Mentessidou S, Skandalakis J. Splenic Cysts: Are There So Many Types?. Am Coll Surg 2007;204:459-65.
5. Hilmes M, Strouse P. The Paediatric Spleen. Semin Ultrasound CT MRI 2007;28:3-11.
6. Jaussaud R, Brasme L, Vernet-Garnier V, Deville JF. Splenic Abscess complicating Salmonella typhi infection. Euro J Clin Microbiol Infect Dis 2000;19:399-402.
7. Stuart BM, Pullen RL. Typhoid: Clinical analysis of three hundred and sixty cases. Arch Intern Med 1946;78:629-61.
8. Cohen JI, Bartlett JA, Corey GR. Extra-intestinal manifestations of salmonella infections. Medicine 1987;66:349-88.
9. Chulay J, Lankerani M. Splenic Abscess; Report of 10 Cases and Review of literature. Am J Med 1976;61:513-22.
10. Choudhury R, Rajiv C, Pitamber S, Akshay S, Dharmendra S. Management of splenic abscess in children by percutaneous drainage. J Pediatr Surg 2006;41:53-6.

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