CASE REPORT

Splenic cyst and its management in a 21-month-old boy: a rare complication of invasive meningococcal disease

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SUMMARY

Splenic complications of invasive meningococcal disease (IMD) are well recognised, though cyst formation is rare, particularly in paediatric populations. The best approach to their management is not yet established. This case outlines the management of a splenic cyst in a 21-month-old boy following severe IMD. The case took place in the context of an acute emergence of serogroup W prompting significant media attention and subsequent change in vaccination practice at a jurisdictional level in Australia. The patient was critically unwell early in the illness, then later a collection in the left upper quadrant was detected, shown on ultrasound to be a 11.6×7.7 cm splenic cyst. In this case, the cyst was managed by ultrasound-guided drainage tube insertion. The residual collection was small and stable on subsequent imaging.

BACKGROUND

Splenic cyst formation is a rare complication of invasive meningococcal disease (IMD) and there are few published cases about their management, particularly in paediatric populations. The case took place in the context of a rise in cases of meningococcus serogroup W. This was of significant interest to Australian vaccination programmes as it coincides with several jurisdiction-based efforts to vaccinate against meningococcus. We hope to add to the available data and relay our approach to other centres.

CASE PRESENTATION

The patient was a previously well 21-month-old boy with no medical problems and vaccinations that were up to date as per the Australian schedule, which includes *Neisseria meningitidis* type C only. He presented to his local secondary hospital emergency department with evidence of rapidly evolving severe sepsis. The onset of symptoms only preceded his presentation to hospital by 12 hours, with initial fever and relative lethargy followed by 2 hours pallor and cool extremities. The patient received 100 mg/kg ceftriaxone and 0.15 mg/kg dexamethasone 24 min after arriving at triage but required intubation and inotropic support within 2 hours. He was subsequently transferred interstate to the Royal Children's Hospital, Melbourne, for ongoing paediatric intensive care. There, he was commenced on extra-corporeal membranous oxygenation (ECMO) 12 hours after his initial

presentation due to profound haemodynamic instability despite aggressive inotropic support. The early course of the illness was complicated by acute kidney injury (AKI) requiring 7 days of continuous veno-venous haemofiltration with a peak creatinine of 268 mmol/L, as well as peripheral necrosis to all four limbs and associated bilateral lower limb compartment syndrome treated with fasciotomies. Following a challenging early disease course, the patient was successfully weaned off ECMO on day 5 of illness and extubated day 11 of illness. He completed a 7-day course of intravenous ceftriaxone. Blood culture taken prior to antibiotics grew *N. meningitidis*, identified as serogroup W.

On day 34 of illness, he was noted to have mild clinical splenomegaly. An immune deficiency screen undertaken as part of his invasive meningococcal disease (IMD) workup noted Howell-Jolly bodies—not previously noted on his initial blood film—with a splenic ultrasound then arranged to investigate further. The ultrasound detected an 11.6×7.7 cm cystic mass in the left upper quadrant, with an irregular, thickened wall, no discernible vascularity and no evidence of viable splenic tissue (figure 1). A CT scan was arranged next for a more detailed assessment which confirmed this to be an isolated lesion with a cystic structure. The patient was also having significant difficulties with feed intolerance during this time for which he was dependent on nasogastric supplementation.

DIFFERENTIAL DIAGNOSIS

A classification system of splenic cysts has been suggested (figure 2).^{1–3} Non-parasitic primary cysts are more common in paediatric populations with endemic parasitic cysts more common in Central America and Africa, mainly caused by *Echinococcus granulosus*.^{4,5} In the case of this patient, it is possible that the splenic cyst was a congenital malformation which predisposed to IMD, though we feel it is more likely this was a sporadic case with secondary cyst formation in the setting of severe sepsis. Without a biopsy specimen to aid in this classification, we cannot be sure though we do note a lack of evidence of hyposplenism on the patient's original blood film at presentation.

TREATMENT

Intervention was warranted from both diagnostic and management perspectives with risk of rupture

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Figure 1 Coronal CT section of the patient's splenic cyst.

or superinfection. This was in addition to standard management of hyposplenism with vaccination against meningococcus (B and A, C, W, Y), *Haemophilus influenzae* type b and pneumococcus, as well as prophylactic antibiotics. Given the rarity of the problem and the paucity of research in the area, a multidisciplinary approach was taken with engagement of the general surgery, haematology, immunology, gastroenterology and interventional radiology teams. A collaborative decision was made for percutaneous, ultrasound-guided drainage. Other options considered included, open or laparoscopic surgical procedures which were thought to be avoidably invasive and transgastric endoscopic drainage for which there were no available local experts.

The ultrasound-guided drainage was performed by an interventional radiologist (EB) with access to the cyst achieved with a 16-gauge Jelco cannula. An Amplatz guidewire was then passed to the collection to facilitate serial dilatation of the tract before the insertion of a 10.2fr Cook MAC-LOC drain tube. There were no complications of the procedure. This drained 350 mL of brown-green, opaque fluid, with numerous neutrophils, consistent with an inflammatory process. There were no malignant cells seen. The glucose level was 2.1 mmol/L, protein level 48 g/L and lactate dehydrogenase level 38 365 units/L. The fluid was

sterile and negative for *N. meningitidis* 16S rRNA gene nucleic acid detection.

OUTCOME AND FOLLOW-UP

A repeat ultrasound was performed 5 days following the intervention showing no identifiable remaining fluid collection, but some solid tissue with minimal vascularity. There was minimal persisting drain output at this stage and the catheter was removed 6 days following its insertion. Of note in this time, the patient's feed tolerance markedly improved and he could transition to full oral feeds over the course of the following month. Follow-up ultrasounds at 2 and 6 months demonstrated a resolving collection. At 6 months, the collection measured 25 mm in diameter with no visible splenic tissue. The imaging was consistent with splenic infarction. Repeat blood films at 1, 2 and 4 months did not show any further Howell-Jolly bodies or other features of hyposplenism. Both the haematology and immunology teams had been involved in the patient's long-term hyposplenism management plan, though currently he is managed by a general paediatrician. He remains on daily prophylactic amoxicillin, is vaccinated against encapsulated organisms as detailed above and is also linked with Spleen Australia⁶ who will ensure the family are up to date with future vaccine and antibiotic recommendations. The patient also has ongoing input by the orthopaedic surgery team following bilateral through-ankle amputations (Syme procedure) 16 weeks following his illness and the plastic surgery team for ongoing management of his hand scarring.

DISCUSSION

The overall incidence of splenic cysts is low with a proposed incidence rate of 0.07%.⁴⁻⁷ Invasive meningococcal disease has been associated with a range of splenic complications in paediatric populations including infarction,⁸ rupture⁹ or necrosis.¹⁰ Although not well documented in the case of IMD, complications of splenic disease include pseudocyst formation¹¹ and secondary asplenia.¹²

The management of splenic cysts has not been studied in previous randomised controlled trials.^{2 13} The suggested threshold for intervention is a cyst diameter of greater than 5 cm or if symptomatic, however, the evidence base for this cut-off is founded on only case-series data.^{1 2 11 14} The major concern is cyst rupture which can be associated with haemoperitoneum, peritonitis, abscess, anaphylaxis, hypersplenism or death.^{7 15} The relative frequency of these complications is not well established.¹⁶ The interventional approaches include percutaneous drainage, complete or partial splenectomy, marsupialisation and unroofing. A laparoscopic unroofing or fenestration procedure was previously preferred in children due to its minimally invasive nature; however, the recurrent rate is high as 50%–88%.^{17 18} The role of marsupialisation is now limited due to its high rates of recurrence.¹⁵ In a 2016 review article of 22 patients with non-parasitic splenic cysts, the team advocated for open cyst excision due to the low rates of recurrence and preserved splenic

Figure 2 Classification system for splenic cysts.¹⁻³

Patient's perspective

Arthur went from happy and healthy on a Friday in late Feb to early Sunday morning being transferred to paediatric intensive care unit (PICU) from interstate in a critical condition, where he was diagnosed with meningococcal septicaemia W strain. The first 2 days after Arthur fell ill while his life was in the balance and he was comatose was an incredibly difficult time for our family emotionally. After nearly 2 weeks in PICU, he was transferred to a ward, knowing he would have some amputations but the levels did not present for another couple of months. During this time, we faced a lot of uncertainty around his gastroparesis which left him with no appetite and not able to keep a lot down while he remained on a nasogastric feed tube. After an ultrasound of his abdomen, followed by a CT scan to confirm, we were advised that Arthur's spleen was gone, most likely died during the acute sepsis stage and in place of the spleen there was a large cyst. This was a troubling time, it was of some relief that there was something that was potentially impacting him being able to eat which could hopefully be resolved with removal. Once the decision was made to externally drain the cyst, it was another surgery among the weekly dressing changes to wounds. The change in Arthur's appetite was overnight. He could tolerate being around food and slowly started eating more and the vomiting subsided. It was a great outcome and the first scan has shown the cyst is not reaccumulating; we will have some follow-up scans down the track to make sure. Meningococcal is a horrific disease, and I am not sure we will ever get over the fact that it is vaccine preventable, yet not offered routinely and discussed during other vaccinations, better still to be on the national schedule. We are eternally thankful and grateful for Arthur's outcome, and the incredible world-class care he has received as, even though he has lost some fingers, both feet and his spleen... some do not survive the horrible disease and many have more extensive life-long impairments both physically and mentally.

Learning points

- Splenic cyst is a possible complication of invasive meningococcal disease.
- The ideal management strategy for splenic cysts remains unclear.
- Our case was managed with percutaneous drainage catheter insertion and was uncomplicated.
- Meningococcal W is an emerging serogroup, potentially preventable by vaccination.

immune function.¹³ Percutaneous drainage is the least invasive of these strategies; however, it is associated with higher rates of recurrence^{13 14} and the subsequent inflammatory reaction may reduce splenic mobilisation making further interventions more challenging.¹⁹ In terms of follow-up, Gezer *et al*¹³ suggests an initial ultrasound at 6 months and 12 months postintervention with subsequent annual ultrasounds if there was no increase in size.

Serogroup W IMD is emerging in several Australian states, following a similar pattern to that seen in the UK, with the same strain (P1.5–2:F1-1:ST11) being isolated.²⁰ In contrast to the vaccination programme in the UK, Australia had only

routinely vaccinated against serogroup C. In 2017, a number of MenACWY vaccine programmes have been implemented, targeting adolescents 15–19 years of age, this including the patient's home state.²¹ South Australia is also trialling a MenB vaccination programme.²²

This case report details a rare pathology in paediatric populations for which there is no standardised course of action. In publishing the case, the team aims to add to the available literature, highlights the management of splenic cysts and the public health implications of IMD.

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