CASE REPORT

The case of the mysterious vanishing spleen: autosplenectomy complicating pneumococcal sepsis

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SUMMARY

A 57-year-old previously healthy fisherman was admitted in fulminant pneumococcal septic shock, with disseminated intravascular coagulation, requiring aggressive management including bilateral below-knee amputations for ischaemic necrosis. He began to recover and was discharged for rehabilitation, however during his convalescence was found to be hypercalcaemic. No malignancy was found on CT scan, but it was noted that his spleen was absent, replaced by a 4 cm smooth-walled, fluid-filled lesion. This was unexpected as an ultrasound in intensive care 10 weeks previously had demonstrated a normal spleen. Functional hyposplenism was confirmed on a peripheral blood film with evidence of target cells, spherocytes and Howell-Jolly bodies. A diagnosis of autosplenectomy complicating pneumococcal sepsis was therefore made, of which there is just one case previously reported. The patient continues to recover well and was discharged on penicillin prophylaxis after receiving vaccinations for hyposplenism.

BACKGROUND

Functional hyposplenism (FH) is most commonly seen as a complication of homozygous sickle cell disease, where repeated attacks of vaso-occlusion and splenic infarction result in fibrosis and atrophy, a process known as autosplenectomy. FH is rarely associated with coeliac disease, alcoholic liver disease, lymphomas and autoimmune disorders, particularly systemic lupus erythematosus (SLE).¹ Previously there has been just one case of autosplenectomy secondary to pneumococcal disease reported by Eshel *et al*² in an otherwise well 56-year-old woman with pneumococcal meningitis.

FH is diagnosed through a combination of imaging and blood film analysis; typical erythrocyte abnormalities include acanthocytes, target cells, spherocytes and Howell-Jolly bodies.³ If further confirmation is required, decreased splenic uptake can be demonstrated using Technetium-99m scintigraphy.¹

FH is characterised by a defective immune response, particularly against encapsulated bacteria. This leaves patients at particularly high risk of sepsis carrying a poor prognosis of 50%–70% mortality despite aggressive management.¹ In newly splenectomised patients, this is recognised as overwhelming postsplenectomy infection syndrome.⁴ Antibiotic prophylaxis and vaccination is, therefore, imperative once FH is identified. Current guidance from

Figure 1 Medical photography demonstrating the patient's bilateral below-knee amputations and the remaining cutaneous stigmata of his purpura fulminans.

the British Committee for Standards in Haematology (BCSH) advise pneumococcal, *Haemophilus influenzae* type b, meningococcal and influenza vaccinations. They also advocate lifelong penicillin prophylaxis for high-risk groups, including those under 16 or over 50 years old, those with a history of invasive pneumococcal disease or those splenectomised for haematological malignancy.⁵ FH is also associated with thromboembolic events due to thrombocytosis and consideration should be given to venothromboembolic prophylaxis especially in the first month following splenectomy.⁶

CASE PRESENTATION

We present a previously healthy 57-year-old fisherman who was admitted to hospital with a 48-hour history of nausea and vomiting before becoming feverish with left-sided abdominal pain and diarrhoea. His partner brought him to the emergency department (ED) over concerns of physical deterioration and a rash spreading on his legs. On admission, he was in extremis with mottled skin, shut-down peripheries and a widespread non-blanching purpuric rash. Initial assessment identified septic shock with a fever of 38.4°C, tachycardia and oliguria. He was also in respiratory distress, requiring high-flow oxygen therapy.

INVESTIGATIONS AND TREATMENT

On admission, our patient was in septic shock and had a metabolic acidosis with a pH of 7.24, a base excess of −8.5 mmol/L and a lactate of 8.6 mmol/L. Blood tests supported the diagnosis of sepsis with a C reactive protein of 174 mg/L and a positive blood culture for *Streptococcus*

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Figure 2 CT abdomen with contrast demonstrating the lesion in the patient's splenic bed.

pneumoniae. A chest X-ray in ED revealed diffuse patchy consolidation at both lung bases with accompanying bilateral effusions. Following initial fluid resuscitation, high-flow oxygen and intravenous antibiotics in ED, management was quickly escalated to intensive care. The patient required significant cardiovascular support for shock and intubation and ventilation for respiratory distress and hypoxia.

Admission bloods revealed a significantly deranged clotting profile with a severe thrombocytopenia and grossly elevated activated partial thromboplastin time and prothrombin time (>250s and >99s, respectively). D-dimer was immeasurable due to high levels. Disseminated intravascular coagulation (DIC) was confirmed on blood film and the violaceous rash diagnosed as purpura fulminans, a syndrome of haemorrhagic infarction of the skin accompanied by intravascular thrombosis. The coagulopathy was treated supportively with vitamin K and blood products including cryoprecipitate and fresh frozen plasma.

Our patient was also found to have an acute kidney injury stage three with a creatinine of 305 $\mu\text{mol/L}$ (baseline 70 $\mu\text{mol/L}$)

Figure 3 Ultrasound image demonstrating a normal spleen.

Figure 4 Blood film showing the characteristics typical of functional hyposplenism: Howell-Jolly bodies (H), target cell (T) and spherocyte (S). Spiculated erythrocytes also noted: echinocyte (E) and acanthocyte (A).

and an estimated glomerulofiltration rate of 19 mL/min. As soon as he was admitted to the intensive treatment unit (ITU), he was filtered due to acidosis and anuria. However, despite haemofiltration, he developed a persistent hyperkalaemia that was attributed to distal tissue ischaemia of his hands and feet associated with the purpura fulminans. This was initially treated with dobutamine and topical glyceryl trinitrate but unfortunately there was already extensive established necrosis and bilateral below-knee amputations (BKAs) remained the only therapeutic option for this man. [Figure 1](#) demonstrates his finger and leg amputations and the cutaneous stigmata of purpura fulminans approximately 3 months after initial presentation.

Following bilateral BKAs, the patient's condition significantly improved and he was stepped down to ward-based care after a month on ITU. It was after discharge to a community hospital for rehabilitation that he developed generalised abdominal pain and bloods identified hypercalcaemia of 2.89 mmol/L with a normal parathyroid hormone. A CT scan of his thorax, abdomen and pelvis was, therefore, performed to look for an underlying malignant cause but none was found. At this point, it was noted that his spleen was absent. In its place, there was a smooth-walled, fluid-filled 4 cm lesion with no macroscopic evidence of normal splenic tissue ([figure 2](#)). This was at odds to a bedside ultrasound on ITU performed 10 weeks previously which had shown normal abdominal anatomy, including a spleen measuring 8 cm ([figure 3](#)). A contrast-enhanced MRI was performed 1 month later which showed a reduction in size and peripheral enhancement of the remaining part-cystic part-solid mass, confirming the suspicion of autosplenectomy.

Further bloods were subsequently taken and confirmed functional hyposplenism through a combination of blood film findings including target cells, spherocytes and Howell-Jolly bodies (as seen in [figure 4](#)) and thrombocytosis. Spiculated cells (echinocytes and acanthocytes) are also noted. In combination with his imaging, these findings are highly suggestive of autosplenectomy and appropriate management was, therefore, initiated including long-term penicillin V prophylaxis and vaccinations as per BCSH guidelines.

While no macroscopic thrombus was seen radiologically, as with all causes of autosplenectomy, the most likely pathophysiology is infarction and atrophy from ischaemic insults. In our patient's case this is probably due to deposition of microvascular thrombi in the splenic vascular system produced in the coagulopathic cascade of DIC. This hypothesis is supported

Patient's perspective

Written by the patient's partner—pseudonym used for anonymity.

"After falling ill on Good Friday 2016 with sepsis, John fought hard and at times it was touch and go. His condition stabilised, but his recovery slowed and two weeks after he was first admitted to ITU we were informed that a double amputation was required if he was going to get any better. This news was devastating, especially with him being a fisherman, and the future seemed bleak. However everywhere you looked there were brave, inspiring people who coped with so much more than this. The staff at the hospital were amazing and after the surgery he really did start to get better. When his kidney function returned it felt like everything was going to be OK again. After a few weeks he was transferred to a community hospital, which saved me a 60 mile round trip every day. However things took a turn for the worst when he began to develop severe stomach pains. A scan was inconclusive and blood tests pointed to the unimaginable possibly being the cause. When we attended an appointment with the consultant gastroenterologist, we were told more tests were needed, but things were not looking as bad as originally feared. Unfortunately, while waiting to start his mobility rehabilitation, John developed urinary sepsis and was re-admitted to an acute hospital. It was during this time we learned of the damage the infection had done to his spleen. This was a big turning point in him making the recovery he has, as once they started to treat him with daily penicillin he stopped getting infections and began gaining weight and building up his strength. It has been a long road to recovery and John still has a battle on his hands. However after everything he has been through he has held on to his positive attitude, bright outlook and strong determination to get back to catching fish for a living. This, coupled with fantastic support from medical staff, friends and family, has made coping with this nightmare situation possible. Life is a wonderful gift."

further by his complaint of left-sided abdominal pain on admission, possibly indicative of symptomatic acute splenic infarction.

DIFFERENTIAL DIAGNOSIS

Our patient had pneumococcal septic shock confirmed by blood cultures, thought likely to be from a chest source. This was complicated by DIC leading to autosplenectomy and functional hyposplenism, most likely from thrombotic causes.

OUTCOME AND FOLLOW-UP

Our patient continues to make an excellent recovery. He remains on penicillin prophylaxis and gabapentin for neuropathic pain which is well controlled. His leg prostheses were fitted at the end of 2016 and he is now walking up to 15 m independently and continuing his rehabilitation physiotherapy. He has required one further terminalisation of his ring finger amputation due to necrosis of the remaining joint, but beyond this has required no further intervention since discharge.

DISCUSSION

This is a rare case of autosplenectomy secondary to DIC in pneumococcal sepsis. The only similar case ever reported is that by Eshel *et al* in 1991 of a 56-year-old woman who developed FH shortly after a pneumococcal meningitis. This was diagnosed by CT and radionuclide scans during her recovery when she developed marked thrombocytosis (without evidence of DIC) and left hypochondriacal pain. In contrast, although our patient presented with left-sided abdominal pain initially, this was not investigated beyond the normal bedside ultrasound. The diagnosis was incidental following investigation of unexplained hypercalcaemia. This fortuitous diagnosis may well be life-saving due to the prophylaxis initiated against his resulting immunodeficiency.

Learning points

- ▶ Functional hyposplenism (FH) is an extremely rare complication of pneumococcal sepsis. More common causes include sickle cell disease, lymphoma, coeliac disease and systemic lupus erythematosus.
- ▶ FH can be diagnosed through a combination of blood film analysis and imaging, including Technetium-99m uptake scan if the diagnosis is uncertain.
- ▶ Once FH is diagnosed it is imperative to protect the patient from infection. The latest British Committee for Standards in Haematology guidelines advocate vaccination against pneumococcus, meningococcus, *Haemophilus influenzae* type b and influenza in all patients, with the addition of lifelong antibiotic prophylaxis in high-risk groups.

Contributors GM conceived and wrote case report, consented the patient and arranged medical photography. MJ wrote the case report and obtained blood film images. DS reported the patient's imaging, critically revised and approved the final report. JB reviewed the patient's blood film, critically revised and approved the final report.

Competing interests None declared.

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