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Case study

An overwhelming post-splenectomy infection (OPSI)

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ABSTRACT

We present the case of a 25 year-old man who had a splenectomy five years previously following a road traffic accident (RTA). He presented to our A&E department one evening with fever and upper abdominal pain associated with nausea & vomiting. Clinical examination was unremarkable. Initial investigations revealed only pyuria. He was treated symptomatically along with parenteral antibiotic and admitted to the general medical ward. Within few hours he deteriorated rapidly with septic shock, multi-organ failure, disseminated intravascular coagulation (DIC) and eventually cardio-respiratory arrest. Despite all resuscitative measures he died within few hours of admission.

Splenectomized patients are prone to develop severe infection, including sepsis and meningitis, due to OPSI, or overwhelming post-splenectomy infection. Presentation may be mild, but the course is rapid and the prognosis is very poor, even in young people. It is important that splenectomized patients receive vaccines according to guidelines, take antibiotic prophylaxis and are educated to seek medical attention at the earliest sign of even minor infections.

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BACKGROUND

Our patient presented with an overwhelming sepsis in a background of an immuno-suppressed state due to an absent spleen. The impaired antibody production and impaired clearance of opsonized particles contributes to an increased lifelong susceptibility of these patients to serious and often fatal infections.[1,2,3]

We report here a fatal septicemia in a splenectomized young person, who did not attend for a repeat of a vaccine, which could have either prevented, or at least ameliorated, the disease.

CASE PRESENTATION

A 25-year old UAE national presented to our A&E department with a 6 hr history of upper abdominal pain associated with nausea, vomiting and fever. He was fully conscious with normal mentation. The patient was involved in a road traffic accident 5 years prior to this presentation and had undergone laparotomy and splenectomy. For a few hours in the post-operative period he received a polyvalent pneumococcal vaccine. He also gave a history of a knee arthroscopic procedure following a sport injury two weeks earlier. Other history and systemic enquiries were unrevealing. Apart from a temperature of 39.2°C, he was haemodynamically stable and other examination was unremarkable; specifically there were no signs of meningitis. Investigation revealed no abnormality apart from pyuria without any casts or bacteriuria. Blood and urine samples were collected for culture and sensitivity.

Initially the admitting doctor diagnosed a urinary tract infection, rather than OPSI, and hence the patient was started on high dose of parenteral Ciprofloxacin. Re-examination in ER revealed a fully conscious but distressed patient, febrile with complaints of nausea and retching, but with stable vital signs. Rebound tenderness was noted in the right hypochondrium along with an old laparotomy scar. There was no neck stiffness or any skin rash. His other blood results and biochemistry were normal apart from a high platelet count of 528,000. Chest & abdominal x-rays were normal. Further inquiry revealed no recent or remote travel, no exposure to animals or birds, and no other family member was sick.

The reviewed diagnoses of the admitting doctor rested between subacute intestinal obstruction and/or acute cholecystitis. An urgent surgical consultation was requested, which ruled out any obstruction. The patient was admitted under medical care with close surgical observation.

A few hours after admission, when the nurse went for a routine vital signs measurement, she found the patient cold, clammy and was vomiting. Blood pressure was found to be low at 80/50 mm of Hg. The on-call doctor was called who found the patient in state of shock with blanching erythema in both flanks. He was immediately started with intravenous chlorpheniramine, epinephrine, fast IV fluid and hydrocortisone. A lack of response, along with epigastric pain, prompted an immediate surgical and medical consultant review.

Bloods were sent for coagulation screening, C-reactive protein (CRP), lactate and a repeat of the biochemistry and full blood counts.

The patient's condition rapidly deteriorated with desaturation, for which he had to be intubated and mechanically ventilated. His antibiotics were changed to ceftriaxone, vancomycin and Tazocin. (Drotrecogin alfa 'Activated Protein C' was not available). Provisional diagnosis now was septic shock, probably due to *Streptococcus pneumoniae* in an immuno-compromised patient (caused by his splenectomy). The rash was probably due to disseminated intravascular coagulation (DIC) syndrome. Acute bacterial meningitis was still considered a possibility.

Despite all the above efforts, our patient deteriorated rapidly and within few minutes developed a cardio-respiratory arrest from which he could not be revived.

INVESTIGATIONS

The patient's blood culture grew *Streptococcus pneumoniae* sensitive to ceftriaxone. His prothrombin and activated partial thromboplastin times were prolonged and platelets went down, indicating DIC. C-reactive protein was 200 mg and blood lactate was high.

DISCUSSION

The human spleen is located in the upper left part of the abdomen and normally measures about 1x3x5 inches, and weighs approximately 150 g.[4] Splenectomized patients are likely to suffer from severe infections, such as sepsis and meningitis, which is known as overwhelming post-splenectomy infection

syndrome (OPSI) in Europe and the USA. The clinical course is rapid, the symptoms are serious and the prognosis is very poor. Splenectomy removes the splenic macrophages that filter the phagocyte bacteria and other blood-borne pathogens.[1] OPSI is usually caused by encapsulated bacteria *Streptococcus pneumoniae*, *Haemophilus influenzae* (HIB) and *Neisseria meningitides* and more than half of those infected with these encapsulated bacteria die.[5] Other pathogens causing OPSI may include bacteria such as *Escherichia coli*, *Pseudomonas aeruginosa*,[6] *Capnocytophaga canimorsus* and others.

The first description of OPSI was published by King and Schumaker in 1952.[7] The disease may start as a minor flu-like illness that rapidly escalates into a fulminant infection.[6] It is most common in the first two years after splenectomy, but may occur decades later.[8] The estimated incidence rate among patients with splenectomy is 0.18–0.42% per year, with a lifetime risk of 5%.[1] OPSI carries a 50–70% mortality rate.[9] Of the patients who die as a result of OPSI, more than 50% succumb within the first 48 hours of admission to the hospital.[2]

The main risk factors are the age at which splenectomy occurs, with children being particularly at risk (splenectomy usually deferred until five years of age); the reason for splenectomy (risk higher with malignancies, thalassemias than idiopathic thrombocytopenic purpura (ITP));[3] and the time interval from splenectomy (most cases occur within two years).[10]

Newer guidelines, particularly those from UK, have emphasized that most infections after splenectomy could be avoided, thus reducing morbidity and mortality, through measures that include appropriate and timely vaccination, antibiotic prophylaxis, education and prompt treatment of the infection.[11] Hence the polyvalent pneumococcal vaccine should be given either two weeks before or two weeks after splenectomy, and repeated in five years or as dictated by levels of antibody titer. Some guidelines recognize the potential superiority of the 7-valent, conjugated pneumococcal vaccine but had no current recommendation for its use. The *Haemophilus influenzae* type b/meningococcal group C (Hib/MenC) conjugated vaccines should be given at least two weeks before or two weeks post splenectomy, but there is currently no recommendation for revaccination. If a person has been previously vaccinated against these organisms with conjugate or polysaccharide vaccines, there is no indication for revaccination. All splenectomy patients should receive influenza vaccinations yearly. The recent British guideline states that lifelong prophylactic antibiotics are still recommended.[11] Finally, all patients who develop the infection, despite all prophylactic measures, should be admitted for prompt treatment with a broad spectrum of systemic antibiotics.[1] The risk of overwhelming, rapid death warrants extreme caution in these patients. Blood cultures are likely to remain positive for a time despite antibiotics early in the course because the concentration of bacteria is extremely high. Early therapy, coupled with excellent supportive care, is the only hope for recovery. Patients and their families should be educated about the nature of this disease and seek medical evaluation early, rather than attempting self-diagnosis and self-therapy.

LESSONS LEARNT

- Each hospital should keep a splenectomy/asplenia & hyposplenism registry. At our hospital (Dibba, Al-Fujairah, UAE) there is now a designated nurse who is responsible for organizing the registry and the vaccination schedules for the post-splenectomy patients.
- Guidelines (local & international) should be strictly followed both for treatment and prophylaxis.
- Education of medical staff, patients and relatives cannot be overemphasized. Patients may wear a bracelet or pendant indicating their health status to health care givers.

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