Case Studies

Pseudomonas aeruginosa bacteraemia in an adult with cystic fibrosis and acute appendicitis

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Abstract

Despite their high bacterial load, bacteraemia is rare in patients with cystic fibrosis (CF). We report an adult with CF who developed Pseudomonas aeruginosa bacteraemia during an episode of acute appendicitis. The Pseudomonas aeruginosa isolated from the blood culture was confirmed by molecular typing to be the same transmissible strain responsible for the patient’s chronic pulmonary infection. We hypothesise that this patient’s bacteraemia was caused by Pseudomonas aeruginosa in swallowed sputum, crossing the inflamed appendiceal wall and entering the blood stream.

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1. Introduction

Bacteraemia in patients with cystic fibrosis (CF) is rare, even in those who are febrile during infective exacerbations [1,2]. This is thought to be due to potent humoral and cellular defence mechanisms in response to chronic pulmonary infection. We present a case of Pseudomonas aeruginosa bacteraemia in a patient with CF and acute appendicitis. We hypothesise that the source bacteraemia was P. aeruginosa in swallowed sputum that crossed the inflamed but intact, appendiceal wall and entered the blood stream.

2. Case history

A 25 year old female with CF presented to the Emergency Department with a 12 hour history of abdominal pain, pyrexia and elevated blood glucose measurements. She had been diagnosed with CF (ΔF508 homozygous) as an infant due to failure to thrive and had exocrine pancreatic insufficiency. She received insulin for CF related diabetes mellitus and was chronically infected with a transmissible strain of P. aeruginosa (Manchester strain) [3]. At the time of presentation her lung function was: FEV1 1.3 L (41% predicted) and FVC 1.75 L (46% predicted). Despite extensive physiotherapy input, she had always been reluctant to expectorate her sputum and would usually swallow it. There had been no significant gastrointestinal problems and her BMI was 21. She did not have a totally implanted venous access device (TIVAD).

The patient’s pain was initially generalised across her abdomen and then became localised to the right iliac fossa (RIF). On examination she was pyrexial (38.5 °C) and looked unwell. Palpation of her abdomen revealed guarding and tenderness in the RIF. Urinalysis was unremarkable and a pregnancy test was negative. An abdominal radiograph showed faecal loading but no dilated loops of bowel. Initial blood tests were normal apart from an elevated white cell count (11.7 × 10⁹/L), platelet count (528 × 10⁹/L), CRP (34 mg/L) and...
glucose (29.7 mmol/L). Blood cultures were taken. An abdominal ultrasound scan was undertaken but this was inconclusive as the appendix could not be visualised.

After discussions between the surgical and medical team the patient was admitted to the ward for observation. During this period the patient remained pyrexial, 2 further sets of blood cultures were therefore taken and the intravenous (IV) cefuroxime that had been prescribed was changed to IV tobramycin and imipenem. After a 24 hour observation, the patient was not showing any signs of improvement and the surgeons decided to take her to theatre. The appendix was found to be thickened and inflamed but not perforated. An appendicectomy was performed which went without complication. Subsequent histology confirmed appendicitis.

Post-operatively, the patient made a good recovery although she had a persistent, low-grade pyrexia. After 48 hours Gram-negative rods were identified from the first set of blood cultures. All three blood cultures isolated *P. aeruginosa* after 1 to 3 days of incubation. Although this organism was multi-resistant, it was sensitive to tobramycin and imipenem. The patient completed 10 days of IV antibiotics and made a full recovery. Repeat blood cultures taken 5 days post-operatively were negative after 5 days of incubation. Subsequently the *P. aeruginosa* isolates were sent for molecular typing (pulsed-field gel electrophoresis), this confirmed that they were the same transmissible strain (Manchester strain) responsible for the patients chronic pulmonary infection.

### 3. Discussion

Despite their high bacterial load, a prospective study only found the incidence of bacteraemia in febrile patients with cystic fibrosis to be 1.2% [1]. This may relate to potent humoral and cellular defence mechanisms in response to chronic pulmonary infection [1] or the enhanced reticuloendothelial clearance of immune complexes seen in patients with CF [4]. We hypothesise that this patient’s bacteraemia developed when *P. aeruginosa* in swallowed sputum was able to traverse the inflamed appendiceal wall and enter the bloodstream.

It is known that ingested *P. aeruginosa* can be recovered from the faeces [5]. It is also known that in appendicitis, bacteria can traverse the intact appendiceal wall prior to perforation and the number of bacteria that do this, increase when there is more inflammation and tissue damage present [6]. There is a high rate of portal bacteraemia (45%) seen in patients with confirmed appendicitis but due to the liver being an effective barrier in preventing bacteria entering the systemic circulation, only 7% have systemic bacteraemia [7].

Although *P. aeruginosa* is the most prevalent pulmonary pathogen for individuals with CF [8], there are only 3 reported cases of *P. aeruginosa* bacteraemia in patients with CF. Two of these patients were already critically ill with overwhelming pulmonary infection and respiratory failure (bacteraemia was confirmed post mortem in one patient and 5 days before death in the other) [1,2]. The third patient had bacteraemia diagnosed after presenting with an infective pulmonary exacerbation [9]. He was successfully treated with IV and oral anti-*pseudomonal* antibiotics. Of note this patient was receiving oral corticosteroids. Other factors thought to increase the risk of bacteraemia and fungaemia include other forms of immunosuppression, diabetes mellitus and the presence of a TIVAD [1].

The *P. aeruginosa* responsible for this patients bacteraemia was one of the transmissible strains responsible for the cross-infection identified at many European, Australian and Canadian CF centres [3,10–12]. These strains are often multi-resistant and exhibit a number of unusual phenotypic features [3]. It is not known if these features have an effect on the mechanisms that usually prevent systemic bacteraemia in CF patients or if they affect the response to treatment. Interestingly, despite the organism being multi-resistant the patient responded well to intravenous antibiotics.

In summary bacteraemia is rare in patients with CF but it must be considered in patients who are unwell and febrile, especially if risk factors, including appendicitis are present.

### Conflict of interests

None.

### References