Pneumococcal native aortic valve endocarditis with mycotic abdominal aortic aneurysm, paraspinal and iliopsoas abscesses and pneumonia revealing a multiple myeloma


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We report a rare case of multiple myeloma presenting with native aortic valve endocarditis with secondary embolic mycotic abdominal aortic aneurysm, contiguous paraspinal and iliopsoas abscesses, and pneumonia due to Streptococcus pneumoniae in a Chinese man. He was treated with aortic valve replacement, endovascular stenting of aneurysm, image-guided drainage of abscesses, and a 6-week course of endocarditic antibiotic therapy followed by chronic suppressive antibiotic therapy. Cases of multiple myeloma presenting with invasive pneumococcal infection were reviewed.

Introduction

Patients with multiple myeloma are at risk of developing infectious complications due to multifactorial immune defects. Occasionally, the first manifestation of an underlying multiple myeloma may be an episode of acute bacterial infection (Kalambokis et al., 2009). The majority of such cases were caused by encapsulated bacteria, namely Streptococcus pneumoniae, Haemophilus influenzae and Neisseria meningitidis, as humoral immunodeficiency plays a key role in the early phase of the disease. Staphylococcus aureus and other Gram-negative organisms have also been implicated but they are usually responsible for infections occurring later in the disease process where prolonged hospitalization, chemotherapy and significant neutropenia contribute to the overall immunodeficient state (Doughney et al., 1988; Kalambokis et al., 2009). As in other cases of invasive pneumococcal infection, bacteraemia, pneumonia, meningitis, and skin and soft tissue infections are the common clinical syndromes. On the other hand, S. pneumoniae rarely causes infective endocarditis in the post-penicillin era and is responsible for only about 0.9–8% of all cases of infective endocarditis (Lindberg et al., 2005). Here, we describe a rare case of native valve endocarditis with secondary embolic mycotic aortic aneurysm, contiguous psoas and paraspinal abscesses, and pneumonia caused by S. pneumoniae in a patient with undiagnosed multiple myeloma who was treated with surgery and endocarditic antibiotic therapy. A literature review of sporadic cases of multiple myeloma with an initial manifestation of invasive pneumococcal infection was conducted.

Case report

A 64-year-old Chinese man with hypertension under satisfactory control presented with low back pain for 1 week. He was brought to the emergency department by his relatives as his condition deteriorated and he became drowsy on the day of admission. He was unconscious and was in shock with a blood pressure of 80/40 mmHg, pulse rate of 112 min⁻¹ and a temperature of 38.6° C. Soon after admission to the intensive care unit, he developed cardiac arrest and recovered after 17 min of cardiopulmonary resuscitation. Transthoracic echocardiogram showed a prolapsed aortic valve with severe incompetence. Blood
tests on admission revealed leukocytosis with neutrophilia, anaemia, renal failure with hyperkalaemia and hypernatraemia, deranged liver function tests, markedly raised C-reactive protein and raised inflammatory markers. Computed tomography (CT) of the thorax and abdomen revealed multifocal pneumonia, a saccular aneurysm in the infrarenal aorta surrounded by rim-enhancing soft tissue mass measuring 2.8 × 3.4 × 4.5 cm, a cystic rim-enhancing lesion in the right lower psoas muscle and the right paraspinous muscle extending from L4 to S1 and measuring 5.8 × 2.3 × 5 cm. Given the critical condition the patient was in, which warranted urgent surgical interventions and antibiotic administration, lumbar puncture was not performed. Intravenous ceftriaxone and vancomycin were started as empirical treatment for possible concurrent meningitis.

Emergency operations including aortic valve replacement, CT-guided drainage of psoas and paraspinous intramuscular abscesses, and endovascular aortic stenting were performed. Blood culture taken on admission and the aspirated pus yielded *Streptococcus pneumoniae* serotype 19F with a penicillin MIC of 1.0 μg ml⁻¹. Histology of the aortic valve showed foci of prominent neutrophilic infiltrate with myxoid change, and areas of fibrosis, hyalinization and calcification compatible with acute valvulitis with myxoid degeneration. The overall findings fulfilled the modified Duke's criteria (one major criterion of new valvular regurgitation on echocardiogram, three minor criteria of fever, embolic phenomena and positive blood culture not meeting major criterion) for infective endocarditis. Thus, the diagnosis was made of pneumococcal native aortic valve endocarditis with embolic mycotic aneurysm at the infrarenal aorta, contiguous right psoas and paraspinous intramuscular abscesses, and multilobar pneumonia.

In view of the unusually severe presentation of invasive pneumococcal infection, investigations for an underlying immunodeficiency were performed. Antibody to human immunodeficiency virus was negative. Monoclonal IgGκ was detected by serum protein electrophoresis. Bone marrow aspirate showed a prominent plasma cell infiltrate, some of which had atypical morphology. Immunohistochemical staining with CD138 highlighted the plasma cells and kappa light chain restriction was demonstrated. The findings were consistent with plasma cell myeloma. Skeletal survey showed multiple lytic lesions in the skull, right mid clavicle and T8 vertebral body suggestive of myeloma involvement.

The patient was treated with a 6-week course of intravenous antibiotics consisting of ceftiraxone and vancomycin, with the vancomycin being stopped after the causative organism and its MIC had been determined. He was maintained on chronic suppressive antibiotic therapy (500 mg amoxicillin orally every 8 h) and remained well. Reassessment CT scan performed 50 days after the drainage showed complete resolution of the intramuscular abscesses and absence of leakage from the endovascular aortic stent. Transthoracic echocardiogram showed a functioning prosthetic aortic valve without paravalvular leakage, abscess or vegetation. Chemotherapy and localized radiotherapy for multiple myeloma were started 1 month after completing antibiotics. The patient was stable at follow-up 9 months after completing antibiotic treatment.

**Discussion**

The close relationship between multiple myeloma and invasive pneumococcal infection has long been recognized. Among the various types of haematological malignancies, multiple myeloma carries the highest risk for the development of invasive pneumococcal infection with an estimated odds ratio of 62.83 (Wong et al., 2010). The proposed immunological mechanisms included functional hypogammaglobulinaemia due to immunoparesis and the consequent attenuated antibody response to antigenic stimuli, defective complement activation, altered neutrophil and dendritic cell functions, and renal failure (Nucci & Anaissie, 2009). On rare occasions, pneumococcal infection might unmask previously undiagnosed multiple myeloma. The present case was a rare example of such cases in which native valve endocarditis with septic embolic phenomena was the initial manifestation of the underlying disease.

As in other cases of pneumococcal endocarditis where left-sided infection predominated, our patient also suffered from an aortic valve endocarditis. The overall mortality rate in pneumococcal endocarditis might be as high as 50 % and the course was often rapidly fatal due to extensive valvular destruction and haemodynamic compromise (Lefort et al., 2000; Ugolini et al., 1986). Our patient was in severe shock and developed cardiac arrest soon after admission because of valvular dysfunction. Without the prompt multidisciplinary surgical interventions, our patient would have likely succumbed to the acute infective episode. Concerning the optimal duration of antibiotic therapy after endovascular stenting, there is as yet, to our knowledge, no conclusive evidence in the literature. While most authors agree that at least 6–8 weeks of initial antibiotics is necessary, life-long suppressive therapy carries both pros (prevention of relapse) and cons (potential side effects and emergence of antimicrobial resistance), and should be prescribed on an individual basis (Rojas et al., 2010). In our patient, chronic suppressive therapy was used to prevent infection of the prosthesis which could lead to leakage from or rupture of the aorta.

Besides infective endocarditis, other forms of invasive pneumococcal infection as the presenting sign of multiple myeloma have been reported (Kalambokis et al., 2009). Including our patient, a total of 13 cases of invasive pneumococcal infection were identified as the first manifestation of an underlying multiple myeloma (Table 1). The mean age was 64.9 years and the male:female ratio was 7:6. Except for one patient with bladder cancer and one with prostate cancer, most patients had no serious
Table 1. Characteristics of patients with myeloma presenting with infections caused by *S. pneumoniae*

<table>
<thead>
<tr>
<th>Reference (year)</th>
<th>Sex/age and underlying condition(s)</th>
<th>Monoclonal paraprotein-light chains (g dl⁻¹)</th>
<th>Clinical syndrome(s)</th>
<th>Site(s) of isolation</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Posner <em>et al.</em> (1978)</td>
<td>M/59 Hypertension</td>
<td>IgG-κ (4.80)</td>
<td>Meningitis, bacteraemia</td>
<td>Blood and cerebrospinal fluid</td>
<td>Antibiotics</td>
<td>Died within hours after admission</td>
</tr>
<tr>
<td>Barasch <em>et al.</em> (1986)</td>
<td>F/50</td>
<td>IgG-λ (2.94)</td>
<td>Bacteraemia</td>
<td>Blood</td>
<td>IV penicillin G</td>
<td>Recovered</td>
</tr>
<tr>
<td>Barasch <em>et al.</em> (1986)</td>
<td>F/70 Hypertension, ischaemic heart disease, atrial fibrillation</td>
<td>IgG-κ (3.16)</td>
<td>Pneumonia, bacteraemia</td>
<td>Blood and sputum</td>
<td>IV penicillin G</td>
<td>Recovered</td>
</tr>
<tr>
<td>Barasch <em>et al.</em> (1986)</td>
<td>F/70</td>
<td>IgG-κ (2.80)</td>
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<td>Blood and sputum</td>
<td>IV penicillin G</td>
<td>Recovered</td>
</tr>
<tr>
<td>Cuesta <em>et al.</em> (1992)</td>
<td>F/47</td>
<td>IgG-λ (7.50)</td>
<td>Left knee septic arthritis</td>
<td>Synovial fluid</td>
<td>Intra-articular lavage with IV penicillin for 4 weeks followed by oral penicillin for 2 weeks</td>
<td>Recovered</td>
</tr>
<tr>
<td>Renou <em>et al.</em> (2007)</td>
<td>M/62</td>
<td>IgG-κ (2.85)</td>
<td>Septic knee arthritis</td>
<td>Synovial fluid</td>
<td>Surgical drainage and IV fosfomycin, vancomycin followed by oral amoxicillin and rifampicin IV vancomycin and ceftriaxone</td>
<td>Recovered</td>
</tr>
<tr>
<td>Aue &amp; Austein (2007)</td>
<td>F/60 Osteoporosis</td>
<td>Not available</td>
<td>Bacteraemia</td>
<td>Blood and bone marrow aspirate</td>
<td>IV vancomycin and ceftazidime</td>
<td>Died of multiorgan failure 4 h after admission</td>
</tr>
<tr>
<td>Bigaillon <em>et al.</em> (2007)</td>
<td>M/88 Carcinoma of bladder</td>
<td>IgG-κ (3.68)</td>
<td>Meningitis, pneumonia, bacteraemia</td>
<td>Blood and cerebrospinal fluid</td>
<td>IV vancomycin and ceftriaxone</td>
<td>Died of multiorgan failure a few days after admission</td>
</tr>
<tr>
<td>Sumrall <em>et al.</em> (2008)</td>
<td>M/75 Adenocarcinoma of prostate, diverticulosis, osteoarthritis, hypertension, dyslipidaemia</td>
<td>IgG-κ (4.64)</td>
<td>Septic knee arthritis, bacteraemia</td>
<td>Blood and synovial fluid</td>
<td>Serial arthrocenteses and IV ceftriaxone for 30 days</td>
<td>Recovered</td>
</tr>
</tbody>
</table>
The monoclonal paraprotein was IgG in 12 (92.3 %) cases and it was not reported in the remaining patient. The mean monoclonal paraprotein level was 4.05 g l⁻¹. Most had concomitant bacteraemia (11/13; 84.6 %). The two cases which did not have a positive blood culture both presented with septic knee monoarthritis. Four (30.8 %) patients had pneumonia, three (23.1 %) had meningitis, four (30.8 %) had septic arthritis, one (7.75 %) had cellulitis and one had endocarditis with embolic mycotic aneurysm and contiguous intramuscular abscesses. The treatment strategy depended on the clinical syndrome and usually included prolonged antibiotic therapy in addition to surgical drainage of localized collection. Intravenous penicillin was commonly used as monotherapy in the past, but ceftriaxone and vancomycin had been increasingly used due to the reduced susceptibility of the organism to penicillin that has developed in recent years. With prompt intervention, most patients (76.9 %) recovered from the acute infection. However, in a significant proportion of these patients (23.1 %), a rapidly fatal course ensued despite appropriate antibiotics and maximal supportive therapy.

As S. pneumoniae continues to cause significant morbidity and mortality among patients with multiple myeloma, preventive measures should be considered in this at-risk group. Traditionally, vaccination against S. pneumoniae has been advocated for immunocompromised patients. However, patients with multiple myeloma were often unable to mount an adequate humoral response to vaccination. As many as 61 % of the patients failed to demonstrate a protective level of antibody titre even after vaccination against S. pneumoniae (Robertson et al., 2000). Thus, in recent years, the use of prophylactic intravenous immunoglobulin has been proposed. In a recent systemic review, it was found that the occurrence of infections among patients with multiple myeloma and chronic lymphocytic leukaemia significantly reduced with the use of intravenous immunoglobulin prophylaxis. However, as there was no overall survival benefit being demonstrated, and there were possible side effects including thrombotic phenomena and renal failure, this use should only be considered on an individual basis (Raanani et al., 2009).

In summary, the present case illustrates that in middle-aged or elderly patients who present with unusually severe or atypical invasive pneumococcal infections, multiple myeloma, along with other common risk factors including alcoholism, human immunodeficiency virus infection, splenectomy, connective tissue disease, corticosteroid use, diabetes mellitus and intravenous drug use (Taylor & Sanders, 1999), should be actively excluded. Aggressive therapy with appropriate anti-pneumococcal antibiotics and surgical interventions should be commenced as soon as possible as the mortality rate of the infection is high if untreated. Further studies should be conducted to investigate the optimal preventative measures against pneumococcal infection in this at-risk group.

### Table 1. cont.

<table>
<thead>
<tr>
<th>Reference (year)</th>
<th>Sex/age and underlying condition(s)</th>
<th>Monoclonal paraprotein-light chains (g dl⁻¹)</th>
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<th>Site(s) of isolation</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present case (2010) M/70 Hypertension</td>
<td>IgG-κ (4.01)</td>
<td>Native aortic valve endocarditis, mycotic abdominal aortic aneurysm, paraspinal and iliopsoas abscesses, bacteraemia, pneumonia</td>
<td>Blood and abscesses</td>
<td>IV ceftriaxone and vancomycin for 6 weeks followed by long-term oral amoxicillin</td>
<td>Recovered</td>
<td></td>
</tr>
</tbody>
</table>

### References


