CASE REPORT

A chronic alcoholic man with high fever, neck rigidity and loss of consciousness: remember the Austrian syndrome a commonly unrecognised invasive pneumococcus triad

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SUMMARY

Austrian syndrome is a rare medical condition characterised by the triad of pneumonia, meningitis and endocarditis due to *Streptococcus pneumoniae*. Native aortic valve insufficiency is the most common cause of cardiac failure in these patients, requiring valve replacement. We report a 52-year-old chronic alcoholic man who presented with fever, neck rigidity and loss of consciousness. Lumbar puncture revealed central nervous system infection while chest X-ray showed pneumonia. Blood and cerebrospinal fluid cultures revealed *S. pneumoniae*. Transoesophageal echocardiography revealed aortic endocarditis with severe valve insufficiency. The patient underwent aortic valve replacement and was finally discharged after completion of 6 weeks intravenous antibiotic treatment. Nowadays, Austrian syndrome is seen infrequently in the antibiotic era. However, clinicians should be aware of this syndrome as its early recognition and prompt combined medical and surgical treatment could reduce morbidity and mortality due to this potentially catastrophic clinical entity.

BACKGROUND

Austrian syndrome is defined as the triad of pneumococcal meningitis, endocarditis and pneumonia. It is commonly seen in middle-aged men with chronic alcohol abuse, where splenic dysfunction may predispose these patients to severe systemic infection by encapsulated organisms like *Streptococcus pneumoniae*.^{1 2} The clinical presentation of these coexisting infections was first described in 1862 by an Austrian pathologist, Richard Heschl. In 1957, Robert Austrian reported eight cases, including six who died because of rupture of the aortic valve.³ Herein, we present a case of Austrian syndrome complicated with aortic valvular destruction, acute cardiac failure and emergency valve replacement. Therefore, we aim to emphasise that early diagnosis is essential for a prompt and timely treatment that may result in good prognosis. In addition, we aim to underline that routine vaccination for *S. pneumoniae* in high-risk groups can also significantly reduce morbidity and mortality due to this potentially catastrophic clinical entity.

CASE PRESENTATION

A 52-year-old Greek Caucasian man was admitted to the Department of Medicine because of 3-day history of high-grade fever with rigours (39°C) and altered level of consciousness. His medical history was unremarkable apart from chronic alcohol abuse for many years but he denied ever intravenous or nasal illicit drugs. At the time of his presentation, his vital signs were as follows: temperature 39.1°C, heart rate 118 beats/min, respiratory rate 24 breaths/min, blood pressure 115/70 mm Hg, and oxygen saturation 88% on room air. Physical examination revealed neck rigidity and auscultation of lungs revealed rales above left lower lobe in the infrascapular area. Moreover, auscultation of heart revealed an aortic murmur which was early diastolic, decrescendo (3 out of 6, Levine scale), heard over the Erb's point (fourth intercostal space on the left sternal border) radiating both to the cardiac apex and the second intercostal space on the right sternal border.

INVESTIGATIONS

The initial laboratory workup showed normochromic normocytic anaemia (haematocrit: 34.3%; haemoglobin: 11.5 g/dL). White cell count was significantly elevated $22.6 \times 10^9/L$ (neutrophils count: $22.8 \times 10^9/L$). Serum urea was 60 mg/dL, creatinine 2.0 mg/dL, glucose 154 mg/dL and lactate dehydrogenase 357 U/L. C reactive protein and erythrocyte sedimentation rate (ESR) were also significantly elevated (22.7 mg/dL and 50 mm/1 hour, respectively). Liver function tests were as follows: total bilirubin, 1.0 mg/dL; aspartate aminotransferase, 46 U/L (upper limit of normal (ULN): 40 U/L); alanine aminotransferase, 94 U/L (ULN: 40 U/L); alkaline phosphatase, 29 U/L (ULN: 140 U/L); and γ -glutamyl transpeptidase 75 U/L (ULN: 32 U/L). Viral serology was negative for HIV and hepatitis B and C viruses. Chest X-ray revealed pneumonia with left lung infiltrates and a small pleural effusion (figure 1). Electrocardiography showed sinus tachycardia. Urinalysis was normal. After blood cultures were obtained, the patient was submitted to lumbar puncture and the cerebrospinal fluid (CSF) revealed elevated white blood cells (1330 with 90% neutrophils predominant),

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Figure 1 Chest X-ray of the patient which revealed left pneumonia and pleural effusion.

elevated protein level (110 mg/dL) and low glucose level (24 mg/dL). CSF Gram staining was negative.

TREATMENT

Empirical treatment for central nervous system infection with intravenous antibiotics (ceftriaxone, vancomycin and ampicillin) and dexamethasone was immediately administered for broad-spectrum coverage against common gram-positive and gram-negative cocci and *Listeria monocytogenes*. Microbiological evaluation revealed the presence of pneumococcal antigen in the urine, whereas blood and CSF cultures after 48 hours grew *S. pneumoniae*. After antibiogram was completed, de-escalation of antibiotics to intravenous penicillin G (24 million units per day) was performed (table 1).

On the sixth day of hospitalisation, the patient deteriorated and had to be intubated because of acute respiratory failure and therefore, transferred to the intensive care unit. Transoesophageal echocardiogram revealed endocarditis causing severe (grade 4) aortic valve regurgitation. Consequently, the patient was submitted to aortic valve replacement and then was transferred back to the Department of Medicine.

OUTCOME AND FOLLOW-UP

The patient was finally discharged after completion of 6 weeks of intravenous antibiotic treatment post-aortic replacement with

penicillin G, and normalisation of biochemical and haematological parameters. As the patient was not previously vaccinated for *S. pneumoniae*, vaccination for *S. pneumoniae* and influenza (annually) was recommended due to the history of chronic alcohol abuse. So far, 1 year after discharge, the patient remains in good health.

DISCUSSION

A variety of microorganisms can cause infective endocarditis; staphylococci and streptococci account for the majority of cases. Nevertheless, streptococci, other than *Bovis* and *Viridans* group (such as *S. pneumoniae*) account for <5%.⁴ Of note, the clinical characteristics of patients with pneumococcal infective endocarditis include the following: frequent involvement of the aortic valve; the infection may involve cardiac valves that were considered morphologically and functionally normal prior to onset of the infection; onset of infection and the clinical progression may be acute and causes severe valve damage and embolic complications.^{3 5} Pneumococcal infective endocarditis typically follows pneumonia in alcoholic patients and may be complicated by concurrent meningitis, a clinical entity referred classically as Austrian syndrome.^{1 6–11} Accordingly in our case, the patient had a history of chronic alcohol abuse and presented with an invasive pneumococcal infection manifesting with pneumonia, meningitis and endocarditis of the aortic valve. Notably, he deteriorated over a short period of time and needed to be urgently transferred to the intensive care unit.

In a large series of patients with infective endocarditis due to *S. pneumoniae*, pneumonia was also present in almost half of cases (46%) while the typical triad of Austrian syndrome was observed in a quarter of them (26.1%).⁵ Of interest, the frequency of this potential catastrophic clinical triad varies from 6.7% to 42% of cases with pneumococcal infective endocarditis.^{3 12} So far, around 70 patients with Austrian syndrome have been reported in the literature. In general, Austrian syndrome involves middle-aged male patients and almost half of them require valve surgery. It seems to carry poor prognosis as the estimated case fatality rate is over 20% and the only risk factor associated with better outcome is early valve replacement.^{1 9 13}

Chronic alcohol consumption is a well-known predisposing condition for invasive pneumococcal disease. Potential explanations include malnutrition, immunological defects such as impaired leucocyte chemotaxis, dysfunction of the reticuloendothelial system and risk of lung aspiration.^{3 12}

Unfortunately, increasing numbers of isolates of *S. pneumoniae* have become resistant to penicillin and may be simultaneously resistant to other beta-lactams as well as other antimicrobial agents. First-line treatment for infective endocarditis and meningitis is high-dose penicillin while in penicillin-resistant cases, high doses of ceftriaxone or cefotaxime are reasonable. Moreover, in ceftriaxone-resistant or cefotaxime-resistant cases, addition of vancomycin or rifampicin is suggested.^{4 14} Although, antibiotic susceptibility to *S. pneumoniae* was not frequently available in the literature in patients with Austrian syndrome, undoubtedly clinical course was worse in cases with penicillin-resistant pneumococci.⁸

The incidence of bacterial meningitis has been decreased after the implementation of effective vaccines. Pneumococcal vaccination is recommended for all children and adults who are at increased risk for developing pneumonia or invasive pneumococcal disease, especially if there are underlying host conditions that compromise antibody responses.¹⁵ Two types of pneumococcal vaccines are approved for use: a pneumococcal

Table 1 Antibiogram of *Streptococcus pneumoniae*

	MIC	Sensitive (S)/ Resistant (R)
Ceftriaxone	0.032	S
Chloramphenicol	<2	S
Clindamycin	0.094	S
Moxifloxacin	0.5	S
Linezolid	1.0	S
Meropenem	0.023	S
Penicillin	0.012	S
Rifampicin		S
Trimethoprim-sulfamethoxazole	<10	S
Vancomycin	0.38	S

MIC, minimum inhibitory concentration.

polysaccharide vaccine that includes 23 purified capsular polysaccharide antigens and a pneumococcal protein-conjugate vaccine that includes capsular polysaccharide antigens linked to a non-toxic protein that resembles diphtheria toxin.¹⁵ In our case, the patient had an unremarkable medical history. Nevertheless, due to chronic alcohol abuse, previous pneumococcal vaccination should be strongly suggested that could prevent adverse outcomes of this fulminant infection. Attending physicians should therefore adhere to immunisation recommendations in high-risk groups.

In conclusion, although pneumococcal infective endocarditis is rare, Austrian syndrome should be suspected in high-risk patients with invasive pneumococcal infection. Clinicians should be aware of this syndrome as its early recognition and prompt combined medical and surgical treatment could reduce morbidity and mortality of this serious clinical entity. In addition, routine vaccination against *S. pneumoniae* should be strongly suggested in patients with underlying comorbidities like our patient according to standard immunisation schemes.

Learning points

- ▶ Austrian syndrome is rare but should be investigated extensively in every case of invasive pneumococcal infection among high-risk groups.
- ▶ Early diagnosis and appropriate combined medical and surgical treatment should be considered promptly.
- ▶ Antipneumococcal vaccination should be strongly suggested in high-risk groups because of the catastrophic complications of invasive pneumococcal disease.

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