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Austrian Syndrome: Triad of the Past or Harbinger of the Future?

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Abstract

Introduction

Austrian Syndrome is a rare complication of disseminated *Streptococcus pneumoniae* infection, consisting of pneumonia, meningitis, and endocarditis.

Case Description

A 64 year old male with a history of alcohol and tobacco abuse presented with a fever of 101.5° F and acute change in mental status. According to his wife, he had been coughing for 3 weeks. Physical examination noted left ptosis and mydriasis, decreased RUL breath sounds, and an irregular pulse. Chest x-ray confirmed RUL pneumonia and EKG showed atrial flutter. CT brain was negative for mass or herniation. Lumbar puncture was performed and blood cultures were obtained. Vancomycin and ceftriaxone were then initiated. He was intubated and transferred to the intensive care unit.

The cerebrospinal fluid and urine antigen tests were positive for *Streptococcus pneumoniae*, so dexamethasone was started as pneumococcal meningitis treatment. Subsequently, the blood cultures were noted to be positive for gram positive cocci. Transesophageal echocardiogram was ordered to rule out endocarditis, and it showed probable mitral valve vegetation. Later, multiple cranial nerve palsies and a brain empyema developed, with thoracic and lumbar emboli resulting in quadriplegia. The patient expired on hospital day 25.

Discussion

The lungs are the usual portal of entry for pneumococcus leading to pneumonia. There are many predisposing risk factors for invasive pneumococcal disease. Alcoholism is one of the strongest risk factors for pneumococcal endocarditis. The complications of Austrian syndrome include systemic embolization, valve perforation and abscess formation. Transesophageal echocardiography is preferred to transthoracic echocardiography in detecting vegetations. Treatment begins empirically with vancomycin and cefotaxime or ceftriaxone until penicillin sensitivity is resulted. Surgical treatment of the valvular lesions is required in the majority of cases. Austrian syndrome is associated with a 24-63% mortality rate, which makes early diagnosis and appropriate treatment critical. The significance of pneumococcal antimicrobial resistance and the role of vaccination will also be discussed.

Summary

Austrian triad was named after Dr. Robert Austrian in 1957, who presented 8 cases that were associated with aortic valve rupture and 6 of which died.

The first to relate this triad was an Austrian pathologist, Dr. Heschl in 1862, and subsequently by Sir William Osler in 1881 (the year that pneumococcus was discovered).

Lung is the usual portal of entry for *Pneumococcus* (82%). Once encountered, both humoral and cellular responses are initiated to fight the infection.

Invasive pneumococcal disease (IPD) is defined by the isolation of pneumococci from normally sterile sites (pleural fluid, blood and CSF). There are risk factors (local and systemic) that have been identified for IPD.

Once bacteremia is sustained, pneumococcus can seed various organs. Meningitis is a common complication, occurring in 59.5% of the cases in one study.

In the preantibiotic era, pneumococcus was responsible for 15-20% of endocarditis. Since the advent of penicillin in the early 1940s, pneumococcal endocarditis has been seen in 3% of patients with infectious endocarditis (IE).

Many reports have linked pneumococcal endocarditis with alcoholism, transforming Osler's triad into a tetrad.

Pneumococcal endocarditis is associated with a high mortality rate (24-63%). Several causes have been identified:

- The presentation is usually acute. As a result, the patient usually lacks the peripheral stigmata of endocarditis.
- The destruction of endothelial tissue generally occurs rapidly followed by acute valvular insufficiency, congestive heart failure and sometimes, early death.
- Cardiac and extracardiac sequelae are common, specifically hemodynamic instability secondary to valve perforation, abscess formation and systemic embolization.

Streptococcus pneumoniae has a predilection for the aortic valve (74%). However, a pre-existing valvulopathy is not the rule in pneumococcal endocarditis.

Echocardiography plays an important role in the diagnosis of IE:

- TTE is a noninvasive, rapid procedure with excellent specificity for vegetations (98%), but has poor sensitivity (<60%). Besides, TTE alone cannot exclude important aspects of IE, including fistulae, leaflet perforation, infection on prosthetic valves, and periannular abscess.
- TEE, however, has significantly higher sensitivity (76% to 100%) and specificity (94%) than TTE for perivalvular extension of infection. Moreover, it enhances visualization of prosthetic valves, with 86% to 94% sensitivity and 88% to 100% specificity for IE.

Treatment

In acute bacterial meningitis or endocarditis, recommendations are to start patient empirically on cefotaxime/ceftriaxone and vancomycin, until penicillin sensitivity is obtained. If the pneumococcus is resistant to cefotaxime (MIC >2 µg/ml), then a vancomycin and rifampin combination should be used.

Corticosteroid is beneficial in treating pneumococcal meningitis patients. It reduces the inflammatory response, increases cerebral perfusion, and improves the adults' outcome.

The clinical course of pneumococcal endocarditis is usually acute and aggressive. Local (peri-valvular abscesses, or perforated valves), and systemic (uncontrolled infection, and multiple emboli) complications frequently occur despite adequate antibiotic usage. As a result, surgical treatment of the valvular lesions is required in the majority of cases.

Indications for surgery are as follow:

- Vegetation involvement (persistent vegetation after systemic embolization, anterior mitral leaflet vegetation, particularly with size >10 mm, 1 or more embolic events during 2 weeks of antimicrobial therapy, increase in vegetation size despite appropriate antimicrobial therapy).⁷
- Valvular dysfunction (acute aortic or mitral insufficiency with signs of ventricular failure, heart failure unresponsive to medical therapy, valve perforation or rupture).⁷
- Peri-valvular extension (valvular dehiscence, rupture, or fistula, new heart block, large abscess or extension to the abscess despite appropriate antimicrobial therapy).⁷

Significance of Pneumococcal Antimicrobial Resistance

Currently, 15 to 30% of *S. pneumoniae* worldwide are multidrug-resistant (MDR).

Despite increase in antimicrobial resistance worldwide over the past few decades, mortality rates for IPD have not increased.

Clinical failures often include factors independent of the pneumococcal antimicrobial susceptibility. These include:

- Host factors (e.g., comorbidities; extremes of age; or underlying immunosuppression).
- Pneumococcal virulence (e.g., capsular subtype).
- Mortality rates are also higher in the presence of: multilobar involvement, hypoxemia, renal insufficiency, and the need for ICU care.

Given the above confounding factors, dissecting out the impact of antimicrobial resistance on clinical outcomes is difficult, if not impossible.

Vaccination and its Importance

In the United States, about 39,750 cases of IPD and 4,000 deaths occur annually.

The clinical spectrum from colonization to IPD depends on the pneumococcal capsular serotype. Currently, 94 capsular serotypes have been identified.

Six serotypes (i.e., 4, 6B, 9V, 14, 19F, and 23F) account for >80% of IPD in children and >50% of IPD in adults in the United States. They also account for the majority of IPD in Europe.

Since the introduction of the PCV7 (4, 6B, 9V, 14, 18C, 19F, 23F), the rate of IPD due to PCV7 serotypes has declined significantly in many countries. In the US, it decreased from 64% of invasive and 50% of noninvasive isolates in 1999–2000 to 3.8% and 4.2%, respectively, in 2010–2011.

The PCV7 also has indirect (herd) effects that have led to decreased incidence of vaccine serotypes disease in unvaccinated children and adults.

However, there have been reports of an increase in non-PCV7 serotypes, especially 19A. This phenomenon is termed 'replacement'.

PCV13 adds pneumococcal serotypes 1, 3, 5, 6A, 7F, and 19A to PCV7's serotypes to provide coverage for over 85% of epidemiologically significant pneumococcal serotypes in the United States and throughout the world.

Austrian Triad and Vaccination

Reports of pneumococcal endocarditis often failed to provide adequate data on either the infecting pneumococcal serotype or patients' vaccination histories.

Among the limited data we have, Aronin et al showed that the most common capsular serotypes that were identified causing Austrian triad were 12, 1, and 8. Hence, PCV 13 could potentially prevent a significant number of Austrian syndrome cases.

Future Prospects

The development of effective conserved pneumococcal protein vaccines (CPPV) that would not target the polysaccharide capsule are currently in trials. These may prevent pneumococcal disease and carriage without resulting in the selective pressure that is thought to drive serotype replacement.

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