Case report

Pneumococcus beyond an Austrian syndrome – A case report

Gonçalo Guerreiro⁎, André Pina Monteiro, Luís Coelho, Pedro Póvoa

AUSTRIAN SYNDROME

Austrian syndrome is a rare entity characterized by Osler’s triad: endocarditis, pneumonia and meningitis, caused by Streptococcus pneumoniae (Austrian, 1957 [1]). This aggressive syndrome is associated with high morbidity and mortality, often due to the involvement of the heart valves and their destruction (Nogué et al., 2019 [2], Araji et al., 2008 [3]).

We present a case of Austrian syndrome in a splenectomised elderly patient with an unusual presentation: septic arthritis complicated by endocarditis, septic cerebral emboli, meningitis and pneumonia. Despite appropriate therapy, the prognosis remained poor and the patient died at day 7.

© 2022 The Author(s). Published by Elsevier Ltd.

ARTICLE INFO

Article history:
Received 21 February 2022
Received in revised form 20 March 2022
Accepted 24 March 2022

Keywords:
Pneumococcus
Invasive pneumococcal disease
Austrian syndrome
Splenectomy
Pneumococcal vaccine

ABSTRACT

Austrian syndrome is a rare entity characterized by Osler’s triad: endocarditis, pneumonia and meningitis, caused by Streptococcus pneumoniae (Austrian, 1957 [1]). This aggressive syndrome is associated with high morbidity and mortality, often due to the involvement of the heart valves and their destruction (Nogué et al., 2019 [2], Araji et al., 2008 [3]).

We present a case of Austrian syndrome in a splenectomised elderly patient with an unusual presentation: septic arthritis complicated by endocarditis, septic cerebral emboli, meningitis and pneumonia. Despite appropriate therapy, the prognosis remained poor and the patient died at day 7.

Introduction

Streptococcus pneumoniae are gram-positive bacteria that commonly colonize the mucosal surfaces of the human upper respiratory tract [4]. In susceptible people however, they can lead to the development of disease through a wide range of infections, including blood and sterile sites - invasive pneumococcal disease (IPD) [5]. Local spread, aspiration or seeding to the bloodstream appear to be the main mechanisms in which invasive diseases develops, but other factors including age, lifestyle traits, comorbidities and even the implied strain have their role [4].

A rare but catastrophic manifestation, a triad of pneumonia, endocarditis and meningitis, was described in 1881 by Sir William Osler. In 1957, Robert Austrian reported 8 cases of Osler’s triad in which 6 patients died, mainly due to aortic valve rupture [1].

Although the incidence IPD has decreased initially due to the introduction of beta-lactam therapy and posteriorly due to universal vaccination [3], it remains a major cause of mortality and morbidity in Europe, especially in immunocompromised patients or with multimorbidity [5].

Case description

We reported a case of an 84 year-old female, partially dependent in daily-life activities for the last 6 months, after a severe SARS-CoV-2 infection.

She had a past medical history of splenectomy due to Immune thrombocytopenic purpura, Rheumatoid Arthritis medicated with methotrexate and corticosteroids and total knee prosthesis bilateral, last revision 8 years ago. Medical records about pneumococcal immunization were not available.

She resorted to an orthopedic surgeon, following a 4-days history of disabling knee pain, fever and anorexia. Upon initial observation, inflammatory signs and ROM limitation were noted. A clinical diagnosis of septic arthritis was made and the patient was referred to the Emergency Room (ER). Blood test on ER admission revealed leucocytosis (WBC 16.6 × 10⁹/L) with neutrophilia (81.2%) and a raised C-reactive protein. She was taken to the Operating Room (OR) and knee arthroscopy, debridement and lavage was performed. Synovial and capsular biopsy, pus samples and blood cultures were collected.

She was admitted to intensive care (ICU) after surgery, with a septic shock diagnosis. Vancomycin and Ceftriaxone were promptly started. As part of the ICU initial screening, a pneumococcal urine antigen test was performed and came back positive. On day 1 of ICU, a preliminary blood culture result reported a gram-positive cocci. Based on this finding, an echocardiogram was performed to rule out endocarditis. However, it demonstrated several mitral vegetations resulting on a partial posterior leaflet destruction, compatible with the diagnosis of endocarditis. Surgical intervention was discussed.
with a cardiothoracic surgeon, but due to the patient’s age and co-
morbidities, she was not eligible for surgical treatment at that time.

On day 2, vasopressor support was suspended. While gradually
reducing sedation, focal neurologic deficits were observed: left
hemiparesis, left conjugate eye deviation and severe aphasia. Cranial
CT scan showed multiple ischemic lesions, compatible with an em-
bolic etiology, suggesting [septic cerebral emboli]. Lumbar puncture
was performed: CSF showed a glucose concentration of 42 mg/dL, a
WBC of 453 cells/µL, a positive pneumococcal CSF antigen test and a
diagnostic of meningitis was made. On day 3, biopsy and blood
cultures came back positive for S. pneumoniae. Antibiotic therapy
was de-escalated according to antibiogram to Cefotaxime. Despite
effective antibiotic therapy, the patient worsens with a decreasing
GCS score (14–11) and recurrent fever. The patient had an unfavor-
able clinical course, with progressive mental status deterioration
and respiratory failure. On day 4 of hospitalization, a new infiltrate
on plain chest radiograph developed, suggesting pneumonia, thus
completing the diagnosis of Austrian syndrome associated with
septic arthritis and embolic stroke.

Given the poor prognosis, the CNS involvement and associated
comorbidities, therapeutic measures were not sufficient and patients
comfort was optimized. She died on day 7.

Discussion

The clinical spectrum of S. pneumoniae infection is wide and
include several entities that are uncommon these days but were
frequent in the pre-antibiotic era. It remains however a common
cause of otitis media, sinusitis, conjunctivitis and the main cause of
community-acquired pneumonia and bacterial meningitis in
adults [5].

Pneumococcal Septic Arthritis is usually described as “rarely
encountered” but it can account up to 6% of all bacterial arthritis in
some series [3]. It usually arises from hematogenous seeding of a
joint with a primary focus being identified only in 52% of the pa-
tients and without any extra-articular disease up to 16% [6]. Ac-

cording to Ross, Saltzman, Carling and Shapiro (2013) major risk
Factors for adult pneumococcal arthritis include rheumatoid arthritis
(22%), alcoholism (20%), osteoarthritis (14%), prosthetic joints (14%),
coronal disease (10%) and corticosteroid use (9%). Classic risk fac-
tors associated with Austrian Syndrome also include alcoholism,
immunosuppression (e.g. asplenia, corticosteroids users, transplant
recipients) and other comorbidities including diabetes mellitus [2].

In this patient, the primary cause of septic arthritis was not
identified. No signs of other local infections were reported during
primary examination. She was admitted to the ICU under deep se-
dation and mechanical ventilation, so clinical history was not di-
rectly obtained from the patient. Even though the articular source
was surgically controlled and the patient was under adequate anti-
biotic therapy, the infection spread to multiple sites including lungs,
heart and central nervous system.

S. pneumoniae is currently an infrequent cause of severe in-
fectious endocarditis (IE), however, a preliminary result of Streptococcus
cereus, a genus of bacteria that include S. viridans group and S. bovis, forced us to rule out endocarditis [7]. Once en-
docardial infection is established, the course is typically aggressive:
the presentation is generally acute, there’s a rapid valvular de-
struction and extracardiac sequelae, including hemodynamic in-
stability and systemic embolization, as reported in our case, are
common [3].

Regarding central nervous system involvement, acute ischemic
stroke is the most common neurologic complication of IE. The pat-
tern seen on CT, with multifocal infarction, suggested a cardioem-
bolic origin. Management in the setting of IE differs, and
anticoagulation [antiplatelet agents are contraindicated [8]. Me-
ningitis is a relatively rare complication of IE and the reverse may
also occur. However, after bloodstream invasion, the release of in-
flammatory mediators caused by infection facilitates pneumococcal
crossing of the blood-brain barrier and the development of me-
ningitis [9].

Despite aggressive therapeutic management, Austrian syndrome
remains associated in literature with high morbidity and mortality.
The difficulty of early recognition of heart involvement (lack of
stigmas and symptoms already explained by lung and CNS involve-
ment) contribute to the prognosis. Much of the literature on Austrian
syndrome emphasizes the need for early surgical intervention,
quoting a reduction in death rate from 60% to 32% when early sur-

gical approach is adopted [10].

In our case, due to all the comorbidities, the patient was not
eligible for surgical treatment. The damage made by IPD lead us to
reduce the therapeutic effort, and the patient died at day 7.

The patient had a previous history of splenectomy and no records
of pneumococcal vaccination were available. The spleen is a lym-
phoid organ with a crucial role in eliminating encapsulated bacteria.
Infections are the most frequent complication and severe infections
may occur. A rapidly progressive fulminant infection, known as OPSI,
overwhelming post-splenectomy infection, has been linked to the
increased risk of spontaneous bacteraemia that these patients pre-

sent, with S. pneumoniae as the main culprit in over 50% of cases.
Incidence of OPSI has been reported to be 5% over the long-term
and mortality can reach up to 50%. The presentation is non-specific, there
is often no obvious portal of infection and it may rapidly progress to
death within 24–48 h [11,12]. Prevention of infection relies on im-
munization, prophylactic antibiotics and patient education, since
most patients are not properly informed about the risks and pre-
ventive measures [11].

Routine immunization has been available since the late 80s and
has changed profoundly the incidence IPD and pneumonia. There are
currently two principal types of pneumococcal vaccines in use:
pneumococcal polysaccharide vaccine (PPV) which is not re-
commended in children less than 2 yo, may require re-vaccination
and promote a less robust immune response compared with pneu-

mococcal conjugate vaccines (PCV), which are highly immunogenic
but are effective against a smaller subset of serotypes [13].

CRediT authorship contribution statement

Each author contributed to the writing of this paper.

Sources of funding

No funding required.

Ethical approval

Yes.

Consent

Report approved by the ethics committee. No close relatives for
sign consent.

Conflicts of interest

No conflicts to declare.

References

[1] Austrian Robert. The syndrome of pneumococcal endocarditis, meningitis and
rupture of the aortic valve. Trans Am Clin Climatol Assoc 1957;68:40–50. [PMC
2248949. PMID 13486606].
[2] Rodríguez Nogué M, Gómez Arraiz I, Ara Martín C, Fray Valle MM, Gómez Peligros
A. Austrian syndrome: a rare manifestation of invasive pneumococcal disease. A
case report and bibliographic review. Rev Esp Quimioter 2019;32(2):98–113.

[3] Velazquez Carlos, Omar Araji, Miguel Barquero J, Perez-Duarte Enrique, Garcia-Borbolla Mariano. Austrian syndrome: a clinical rarity. Int J Cardiol 2008;127(2008):e36–8.

[4] Weiser JN, Ferreira DM, Paton JC. Streptococcus pneumoniae: transmission, colonization and invasion. Nat Rev Microbiol 2018;16(2018):355–67.

[5] Factsheet about pneumococcal disease. European Centre for Disease Prevention and Control; 2021. Retrieved December 28, 2021, from https://www.ecdc.europa.eu/en/pneumococcal-disease/facts.

[6] Ross JJ, Saltzman CL, Carling P, Shapiro DS. Pneumococcal septic arthritis: review of 190 cases. Clin Infect Dis: Publ Infect Dis Soc Am 2003;36(3):319–27.

[7] Murdoch DR, Corey GR, Hoen B, et al. Clinical presentation, etiology, and outcome of infective endocarditis in the 21st century: the international collaboration on endocarditis–prospective cohort study. Arch Intern Med 2009;169(5):463–73. [2009].

[8] Morris NA, Matiello M, Lyons JL, Samuels MA. Neurologic complications in infective endocarditis: identification, management, and impact on cardiac surgery. Neurohospitalist 2014;4(4):213–22.

[9] Mook-Kanamori BB, Geldhoff M, van der Poll T, van de Beek D. Pathogenesis and pathophysiology of pneumococcal meningitis. Clin Microbiol Rev 2011;24(3):557–91.

[10] Atkinson K, Augustine DX, Easaw J. Austrian syndrome: a case report and review of the literature. BMJ Case Rep 2009. [2009, bcr03.2009.1724].

[11] Buzelé R, Barbier L, Sauvanet A, Fantin B. Medical complications following splenectomy. J Visc Surg 2016;153(4):277–86. https://doi.org/10.1016/j.jviscsurg.2016.04.013

[12] Sinwar PD. Overwhelming post splenectomy infection syndrome – review study. Int J Surg 2014;12(12):1314–6. https://doi.org/10.1016/j.ijsu.2014.11.005

[13] Pletz MW, Maus U, Krug N, Welte T, Lode H. Pneumococcal vaccines: mechanism of action, impact on epidemiology and adaption of the species. Int J Antimicrob Agents 2008;32(3):199–206. https://doi.org/10.1016/j.ijantimicag.2008.01.021