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Case Report

Austrian Syndrome: Community-Acquired Pneumonia in an Unusual Triad

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Abstract

Austrian syndrome corresponds to the triad of meningitis, pneumonia, and endocarditis caused by *Streptococcus pneumoniae*, there is no global or local incidence given the infrequency of entity. Scarce cases are published in Latin America, with none of them in Colombia. A case of Austrian syndrome by penicillin-resistant *S. pneumoniae* in an immunocompetent patient is presented. Aortic valve is the most frequent site involved in Austrian syndrome; this patient had an unusual localization of the vegetation on the right coronary artery ostium. The prognosis is poor with a mortality rate of 30% or higher, this patient survived despite systemic complications. Vaccination status impacts in prevention and severity of cases because responsible serotypes are often included in available vaccines. The patient had a serotype covered by available vaccines; however, her vaccination status was unknown. Thus, we present the first case reported in Colombia of Austrian syndrome by a penicillin-resistant *S. pneumoniae*, in a patient with no identified comorbidities or toxicological history, with a successful evolution.

KEYWORDS: Pneumococcal infections, meningitis, pneumococcal. endocarditis, bacterial, pneumonia
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INTRODUCTION

Austrian syndrome was first described in 1862 by the Austrian pathologist Richard Weschl. Later in 1881 Sir William Osler suggested the spread of pneumococcus to the central nervous system as a complication of pneumonia and endocarditis. Some years later Netter emphasized the predisposition due to the aortic valve, and finally the American infectologist Robert Austrian described 8 cases in 1957 revealing that lethality was commonly due to rupture of the aortic valve.[1,2]

Austrian syndrome is composed of the triad of pneumonia, endocarditis, and meningitis, secondary to infection by *Streptococcus pneumoniae*.[3] Considering the introduction of penicillin in 1940 and increasing vaccination against pneumococcus, infections by this pathogen have decreased drastically in years,[4,5] a clear example of this is the change in the incidence of infective endocarditis caused by this pathogen, previously being up to 15% and currently reaching rates less than 3%.[6] Furthermore, this condition is an uncommon event, with less than 100 cases occurring between 1957 and 2017 according to the literature reviewed.[7] Hence, we present the case of a patient with no previous comorbidities who developed Austrian syndrome, with a satisfactory outcome after the interventions performed despite what is described in the literature.

CASE REPORT

The patient gave us her written informed consent to present information about the medical record and diagnostic images. She was a 38-year-old patient with a single personal history of migraine without aura, who was assisted to the emergency department due to four days of moderate-intensity holocranial headache, left otalgia with scant ipsilateral otorrhea, dry cough, general malaise, and fever quantified at 39°C, with two emetic episodes and impaired state of consciousness 24 hours before consultation. Hypotension was documented on admission along with progressive neurological deterioration (Glasgow coma scale of 8/15) with adequate oxygenation indices and ensuring the airway with orotracheal intubation for invasive mechanical ventilation was decided. A simple brain tomography was performed without parenchymal or mastoid findings, and cerebrospinal fluid studies showed polymorphonuclear pleocytosis (32 cells/mm^3) with hyperproteinorrhachia (683 mg/dL) and hypoglycorrhachia (2 mg/dL, with 110 mg/dL of glycemia) being compatible for concurrent bacterial meningoencephalitis with a positive culture for penicillin-resistant *S. pneumoniae*, whence she received management with intravenous ceftriaxone (2 g twice a day) plus vancomycin (1 g twice a day) due to the unavailability of susceptibility test to third generation cephalosporins and quinolones. She also received dexamethasone 10 mg every 6 hours for 4 days scheme as part of the pneumococcal meningitis protocol. The blood laboratory tests were remarkable for mild leukocytosis (14 360 cells/mm^3) with neutrophilia, mild anemia (11.4 g/dL), and elevated C-reactive protein (341 mg/L); liver and renal functions were normal, and the fourth generation HIV antigen test was negative. High-resolution chest tomography

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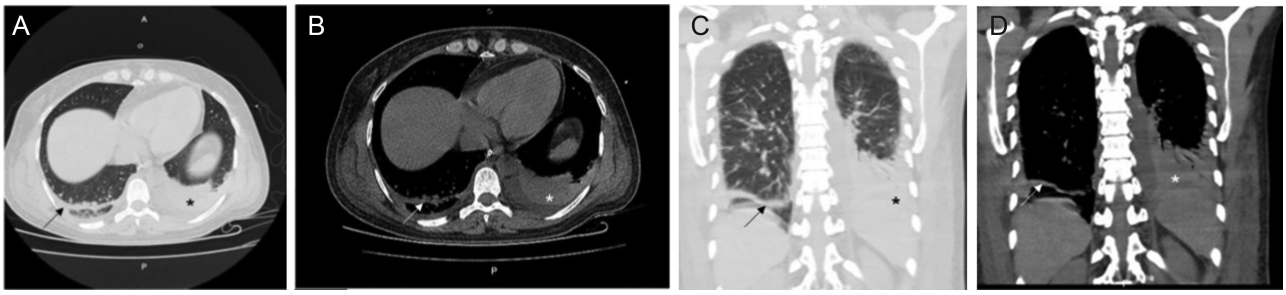

Figure 1. High-resolution computed tomography of the chest. (A-B) Axial section in the lung and mediastinum window shows the presence of atelectasis (arrow) and bibasal pleural effusion, predominantly left (asterisk) with concomitant consolidation. (C-D) Coronal slices using the lung and mediastinum window with the findings referenced in A and B. Source: Authors.

showed left basal consolidation and bilateral pleural effusion compatible with community-acquired pneumonia (Figure 1), while endotracheal tube aspiration cultures were negative. During her stay, she had two peripheral blood cultures positive for penicillin-resistant *S. pneumoniae*, whereby a transthoracic echocardiogram was performed reporting a 4 mm vegetation at the ostium of the right coronary artery, with mild primary aortic insufficiency and left ventricular ejection fraction conserved. In sum, the findings mentioned were compatible with Austrian syndrome. The serotype of *S. pneumoniae* was 19A, which was identified by the Colombian National Institute of Health.

The patient completed 42 days of biconjugate antibiotic management described above, with progressive neurological improvement, ventilatory weaning with the suspension of oxygen therapy, and resolution of bacteremia with negative control peripheral blood cultures. A control chest tomography showed the resolution of left basal pneumonia (Figure 2), and a control transthoracic echocardiogram showed a complete resolution of aortic vegetation. She was discouraged with mild bradypsychia as probably sequalae, without other respiratory or cardiovascular symptoms.

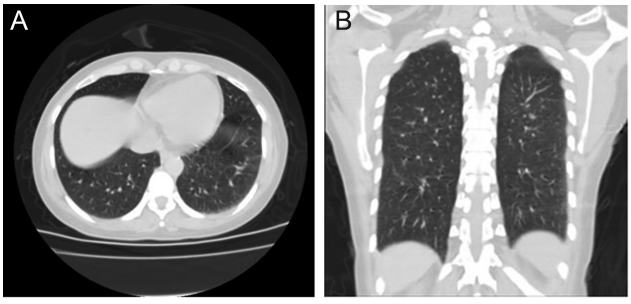

Figure 2. High-resolution computed tomography of the chest after 42 days of antibiotic treatment. (A-B) Axial and coronal section of the lung showing the complete resolution of left basal consolidation, pleural effusion, and atelectasis. Source: Authors.

DISCUSSION

Regarding epidemiological aspects of Austrian syndrome, a higher proportion has been reported in middle-aged men with a history of alcoholism, being the main risk factor that was associated with hyposplenism giving greater susceptibility to infections caused by encapsulated germs;[8] other known risk factors are splenectomy, diabetes mellitus, cirrhosis, and immunosuppression.[9-11] On the other hand, it has been found in case series of patients with infective endocarditis secondary to *S. pneumoniae* that pneumonia was present in almost half of the cases (46%), and Osler's triad in about 26%.[12] In contrast to the previously mentioned, the patient presented was a young adult woman, with no history of alcoholism or other previous comorbidities, which differs from the profile described in the published literature.

The usual clinical presentation is initially given by upper respiratory compromise (otitis or sinusitis) or lower respiratory compromise as pneumonia, with subsequent hematogenous dissemination to the central nervous system and heart.[13] In the case described, the chronology was not clear at all since the respiratory and neurological symptoms began almost simultaneously. The evolution of Austrian syndrome is usually rapid and aggressive,[7] a behavior like the one described on admission, and mortality rates reported up to 32%.[14]

Each component of the syndrome has important characteristics to highlight; in pneumonia, most of the patients manifest fever, chills, cough with purulent expectoration, and pleuritic pain with imaging evidence of non-segmental homogeneous consolidations.[14] However, this patient had left inferior pneumonia and left otomastoiditis, with a non-etiological determination as a pitfall but with a good response at those levels with therapy established. Otherwise, regarding infective endocarditis, only 1.2% present with Austrian syndrome,[11] and in these patients the left native valvular apparatus is more frequently affected, being the aortic valve more commonly compromised up to 49.3% of cases, without ostium compromise been reported.[8,12]

The penicillin resistance in *S. pneumoniae* strains has been increasing worldwide, in Colombia has been reported

MAIN POINTS

- Austrian syndrome does not seem to only affect immunocompromised people, and this could also suggest possible individual susceptibilities not identified.
- Penicillin-resistant pneumococcal disease is an incident problem found in community settings; a good prognosis is supported by appropriate antimicrobial stewardship despite the aggressive course reported in literature.
- Serotypes included in available vaccines could be responsible for disseminated and aggressive presentations such as Austrian syndrome in unvaccinated people. Vaccination programs must be carried out effectively so that they can have a better impact in prognosis.

intermediate resistance of 11% and total resistance of 4%.[15] Notwithstanding, in penicillin-resistant pneumococcal meningitis, the treatment of choice according to Infectious Diseases Society of America are vancomycin plus ceftriaxone when the susceptibility tests to ceftriaxone are unavailable and the minimum inhibitory concentration is >0.1 µg/mL. The role of systemic steroid in meningitis has been reported in terms of survival and neurological complications like hearing loss.[16]

Finally, vaccination against pneumococcus has decreased mortality secondary to this germ; however, these aggressive entities have also been described in serotypes not included in available vaccines with an increased probability of antimicrobial resistance, being pneumonia the usually route of entrance to subsequent dissemination.[10] The responsible serotype of this infection was 19A, a serotype included in the 13-valent pneumococcal conjugate vaccine (PCV13) and 23-valent pneumococcal polysaccharide vaccine (PPSV-23), which we did not know if they had been applied to the patient; these data support the importance of improving population vaccination strategies to avoid exposing them to risk of complications like this. This is the first case described in Colombia of an Austrian syndrome due to a resistant pneumococcus in a young adult patient without underlying risk comorbidities, and because its infrequent presentation, knowledge of the entity becomes more relevant given the high mortality described, the risk of valvular complications and neurological sequelae.

In conclusion, Austrian syndrome is an unusual form of invasive pneumococcal disease, usually conditioned by medical history of risks that increase the probability of dissemination at presentation. Additional features to highlight are the unusual location of vegetation, the fast resolution time of systemic compromise with satisfactory course despite aggressive course at debut, and the importance of vaccination to prevent the aggressive course of these infections.

Informed Consent: Written informed consent was obtained from the patient who agreed to take part in the study.

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