Austrian Syndrome: A Devastating Triad

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Abstract

Austrian syndrome is a rare clinical triad of pneumococcal pneumonia, meningitis, and endocarditis first described in 1957. Despite the advent of antibiotics and pneumococcal vaccination, it remains associated with a mortality rate approaching 30–60%, particularly when diagnosis or surgical intervention is delayed. We present the case of a 37-year-old male with an atypical stroke-first presentation of Austrian syndrome. His case underscores the diagnostic challenge, highlights the role of congenital bicuspid aortic valve as a risk factor, and demonstrates the life-saving impact of early valve replacement.

Introduction

Austrian syndrome represents one of the most lethal infectious disease triads, with fewer than 100 cases reported in the modern literature. It is classically seen in middle-aged alcoholic men, but recent reports show expanding demographics, including younger immunocompetent patients. Mortality is high: up to 80% in untreated patients, ~40% with antibiotics alone, and significantly reduced (to ~20%) with combined surgical intervention. The disease poses a diagnostic challenge because symptoms often mimic isolated pneumonia or meningitis, delaying recognition. Our case contributes to the limited data by describing a young patient without traditional risk factors who presented with ischemic stroke, a manifestation rarely reported in this syndrome.

Methodology

We performed a detailed retrospective chart review of this patient's hospitalization, including presenting features, diagnostic imaging, microbiologic testing, and therapeutic interventions. PubMed and EMBASE databases were searched for "Austrian syndrome," "pneumococcal endocarditis," and "stroke in endocarditis." Fewer than 10 reported cases of Austrian syndrome with initial cerebrovascular presentation were identified, highlighting the rarity of this clinical course. Comparative analysis was performed with published case reports and case series to contextualize our patient's presentation and outcomes

Results

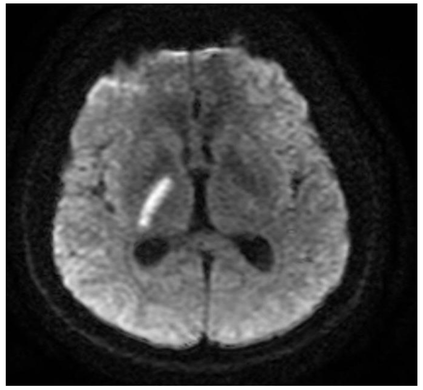
The patient presented with fever (38.2°C), hypoxia (SpO_2 89%), hypertension (190/86), and acute left-sided weakness (NIHSS 18). CTA revealed right ICA occlusion, though he was outside the thrombolysis window and not eligible for thrombectomy. Chest imaging confirmed bilateral pneumonia. Despite antibiotics, he deteriorated neurologically, necessitating intubation. MRI demonstrated multiple embolic infarcts, and CSF findings were consistent with bacterial meningitis. TEE revealed a bicuspid aortic valve with vegetations and severe regurgitation. Blood cultures grew *Streptococcus pneumoniae*. He completed a six-week antibiotic course and underwent surgical aortic valve replacement, leading to significant recovery.

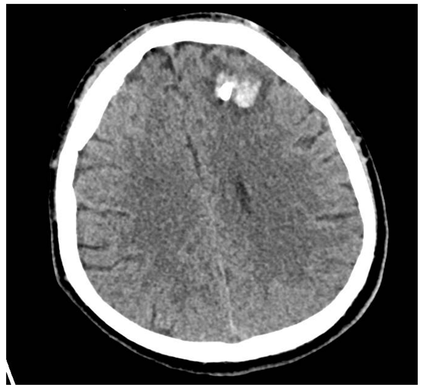
In contrast to most reported Austrian syndrome cases, our patient was young, immunocompetent, and without alcohol or smoking history. His congenital bicuspid aortic valve was likely the key predisposing factor. Stroke as the sentinel event of Austrian syndrome is exceedingly rare - fewer than 10 cases reported worldwide.

Conclusion

This case underscores the need for heightened clinical suspicion of Austrian syndrome in patients with *S. pneumoniae* bacteremia and multi-organ involvement, even in the absence of classic risk factors. It illustrates how congenital structural heart disease can predispose patients to this devastating triad. Importantly, stroke-first presentations, while exceedingly rare, can delay recognition and worsen prognosis. Clinicians should maintain a low threshold for early echocardiography and neuroimaging in septic patients with neurologic deficits. Survival hinges on rapid diagnosis, prompt initiation of broad-spectrum antibiotics, and surgical valve replacement. Our case adds to the limited body of literature documenting young, immunocompetent patients with Austrian syndrome and highlights an emerging need for awareness of atypical presentations.

Imaging





Left: Ischemic CVA in Internal Basal Ganglia region

Right: Hemorrhagic conversion of septic emboli

References

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