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Case Report

Emergent double valve replacement in Austrian syndrome☆☆☆

Abstract

Bi-valvular pneumococcal endocarditis in Austrian syndrome, which includes a triad of pneumococcal endocarditis, pneumonia, and meningitis, is a rare but life-threatening disease. We present a case of a woman found to have Austrian syndrome who presented to the emergency department (ED) with dehydration and radiographical signs of lobar pneumonia and quickly deteriorated to fulminant cardiogenic shock in less than four hours. An early echocardiogram in the ED confirmed a diagnosis of bi-valvular endocarditis with severe aortic and mitral valve insufficiency and large vegetations on the valve leaflets requiring emergent surgical intervention with double valve replacement. Assumed meningitis as a part of the triad of Austrian syndrome was confirmed by imaging the day after hospital admission. Early diagnosis of endocarditis by obtaining the echocardiogram in the ED along with emergent surgical intervention allowed for a favorable outcome for the patient.

Severe bi-valvular pneumococcal endocarditis is a serious and rare clinical entity. The clinical course is associated with significant native valve destruction and a poor outcome [1,2]. Concurrent presentation of pneumococcal endocarditis in a triad with pneumococcal pneumonia and meningitis represents “Austrian syndrome,” which is an invasive and aggressive form of pneumococcal disease with high morbidity and mortality [3]. Early diagnosis and timely surgical intervention are the best predictors of an optimal outcome [2,4]. In this report, we present a rare case of Austrian syndrome with a rapidly deteriorating course in the emergency department (ED) that required an emergent mitral and aortic valve replacement to yield a favorable clinical outcome.

A 30-year-old woman with an unremarkable medical history presented to the ED with a 1-week history of generalized weakness, malaise, anorexia, vomiting, diarrhea, neck pain, and myalgias. She denied any fever, chills, cough, headache, or photophobia. She had recently self-detoxified from long-standing cocaine, marijuana, and phencyclidine abuse three weeks before presentation but denied any history of intravenous drug use. She denied recent dental treatment or a history of any invasive procedures. The patient was triaged with an Emergency Severity Index level of 4 of 5 (less urgent), with initial blood pressure of 110/68 mm Hg, pulse rate of 85 beats per minute, respiratory rate of 18 breaths per minute, temperature of 98.3°F, and an oxygen saturation of 98% on room air. She was alert and oriented with no neurologic deficits and no meningeal signs. She had bilateral

crackles at her lung bases. Her heart sounds were normal without a friction rub or gallop, but a harsh, systolic III/VI murmur was audible throughout the precordium. Abdominal examination was normal with no costovertebral angle tenderness. No rash, needle track marks, petechiae, or purpura was found.

Laboratory testing revealed an initial leukocytosis with a white blood cell count of $20.47 \times 10^3/\mu\text{L}$, with 90% segmented neutrophils and 6% band neutrophils; hemoglobin level of 9.2 g/dL, hematocrit of 26.7%, and a platelet count of $21 \times 10^3/\mu\text{L}$. Her prothrombin time was 11.4 seconds, international normalized ratio was 1.16, and partial thromboplastin time was 49.4 seconds. The initial lactate was 3.7 mmol/L. The serum biochemistry including bicarbonate level and anion gap and liver enzymes were in the reference ranges. Initial blood cultures were drawn in the ED and later grew *Streptococcus pneumoniae* sensitive to penicillin (minimum inhibitory concentration, 0.016 $\mu\text{g}/\text{mL}$). The enzyme-linked immunosorbent assay for HIV was negative. A chest radiograph revealed diffusely increased

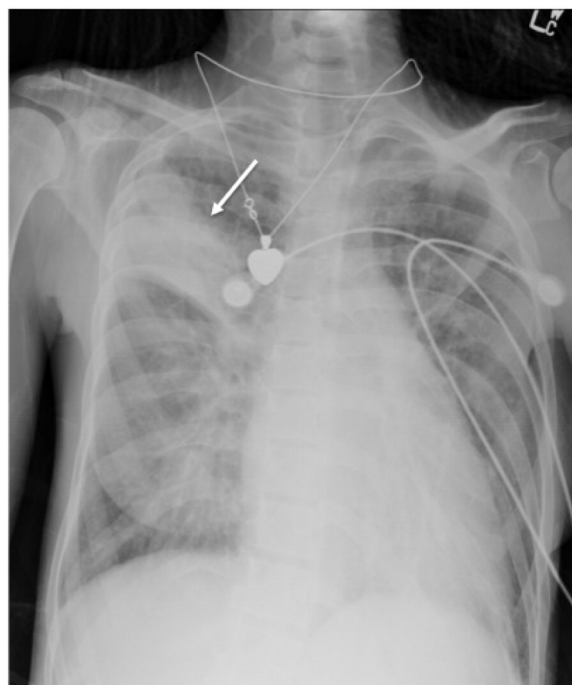


Fig. 1. An anteroposterior (AP) view of chest x-ray taken at the time of patient's presentation shows an area of consolidation in the right upper lobe (arrow) as well as diffuse increased interstitial marking and mild cardiomegaly.

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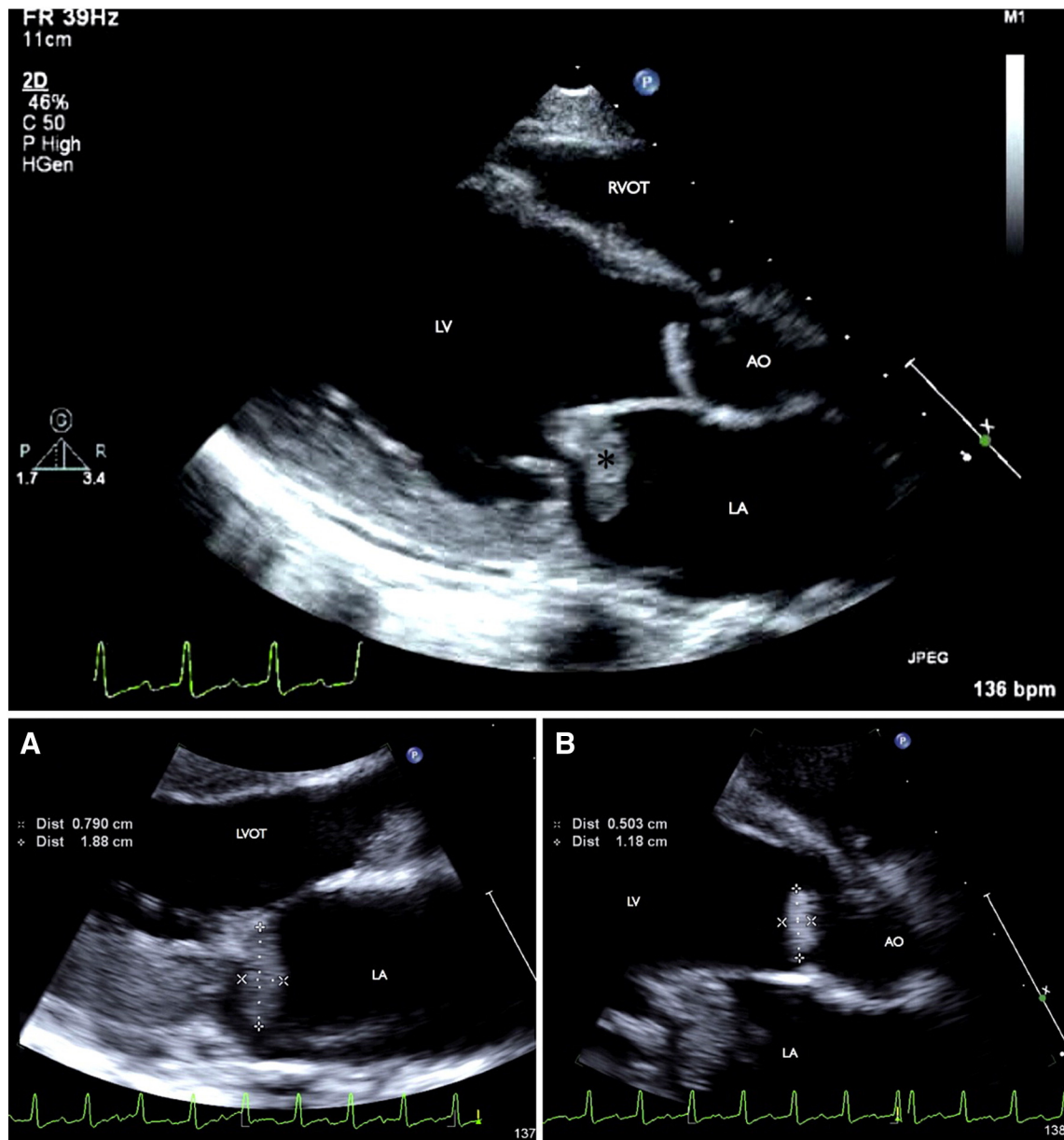


Fig. 2. Two-dimensional echocardiogram: parasternal long-axis view (top) with magnified mitral valve in systole with vegetation of the mitral valve (A) and magnified aortic valve in diastole with pedunculated vegetation of the aortic valve (B). Abbreviations: AO, aorta; LVOT, left ventricular outflow tract; LA, left atrium; LV, left ventricle; RVOT, right ventricular outflow tract; *, vegetation.

interstitial lung markings with right upper lobe consolidation and mild cardiomegaly (Fig. 1).

Oxygen therapy and intravenous administration of fluids were commenced. The initial antibiotic therapeutic regimen, consisting of intravenous vancomycin and ceftriaxone, was chosen empirically for severe community-acquired pneumonia. Despite optimization of medical therapy and aggressive fluid resuscitation, her condition deteriorated over the next 4 hours. She developed shock with a blood pressure of 80/50 mm Hg, persistent sinus tachycardia with a heart rate of 140 to 150 beats per minute, and oliguria. Subsequently, she developed acute respiratory failure with oxygen saturation as low as 80% on 100% nonrebreather oxygen therapy. She required emergent endotracheal intubation and mechanical ventilation.

During the course of her ED admission, the patient underwent a 2-dimensional transthoracic echocardiogram (TTE), which showed a native aortic valve with a large, 12-mm, pedunculated, highly mobile vegetation attached to the noncoronary cusp and severe aortic valve

insufficiency. A pedunculated vegetation of the mitral valve attached to the anterior mitral leaflet and severe mitral valve regurgitation were also seen (Fig. 2). Features of persistent hemodynamic instability as well as the echocardiographic findings of severe aortic and mitral insufficiency and of large vegetations (>1 cm) prompted emergent cardiac surgery. As such, the patient was transferred to the operating room directly from the ED.

In the operating room, cardiopulmonary bypass was established, and an apical left ventricular vent was placed. Intraoperative findings included large masses on the noncoronary cusp of the aortic valve, black-green in color, and approximately 1.5 cm in transverse diameter. A tear in the noncoronary cusp was also noted. Another large green-black, smooth mass was found at the commissure between the posterior and the anterior leaflets of the mitral valve. The posterior leaflet of the mitral valve was unsupported in this particular area. Emergent valve replacements were performed using a 19-mm St Jude (St Jude Medical, St Paul, MN) mechanical prosthesis

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