

Complex Cardiac Management in a Patient with Kartagener's Syndrome

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Description

A 32-year-old woman presented with a four-day history of fever and overall feeling unwell. Due to persistent symptoms, blood cultures were taken, revealing a positive result for *Staphylococcus aureus*. Initially, she was treated with meropenem and gentamicin due to a potential history of allergies to cephalosporins and vancomycin. Once methicillin-sensitive *S. aureus* was confirmed, her treatment was switched to oxacillin and gentamicin. Subsequently, a Transesophageal Echocardiogram (TEE) showed an 11 × 5 mm vegetation on the Prosthetic Mitral Valve (PMV), prompting her transfer to a specialized medical center for consideration of repeat mitral valve replacement and further care. At the time of transfer, she was without fever, with a blood pressure of 96/57 mm Hg, heart rate of 90 beats/min, and oxygen saturation of 95% on room air. On cardiovascular examination, she exhibited dextrocardia, with heart sounds heard on the right side of the chest, and a regular rhythm with mechanical S1 and S2. A crescendo-decrescendo murmur was audible at the right lower sternal border and right apex.

Kartagener's syndrome

The patient's medical history revealed Kartagener's syndrome diagnosed at birth, characterized by dextrocardia, bronchiectasis, heterotaxy, ventricular septal defect, and anomalous venous connections to a common atrium. She had undergone six sternotomies previously, starting at 18 months of age for closure of a ventricular septal defect, redirection of anomalous venous connections, and mitral valve repair. Further interventions included mitral valve repair at the age of 5, insertion of a dual-chamber pacemaker for sick sinus syndrome (later removed due to infection), correction of an anomalous right-sided superior vena cava to the left atrium, and replacement of a permanent pacemaker for complete heart block. Additionally, she had undergone mitral valve replacement due to infective endocarditis, complicated by pseudomonas mediastinitis necessitating repeat sternotomy for the insertion of a new mechanical valve and replacement of interatrial baffle and epicardial pacing leads. Upon reaching the end of her

epicardial pacing system's lifespan, she underwent transvenous pacemaker replacement, which was complicated by atrial lead dysfunction, requiring tunneling to connect an existing epicardial atrial lead to the new device.

After transferring, the patient underwent a repeat transthoracic echocardiogram which revealed a minor posterior paravalvular leak and an uncertain lesion on the mechanical mitral valve leaflet, suggestive of a potential vegetation. Despite repeating blood cultures showing no growth during hospitalization, a comprehensive evaluation for potential septic emboli was undertaken. This included computed tomography angiography of the chest, abdomen, and pelvis, revealing a wedge-shaped hypoattenuation in the right kidney, indicating a renal infarct, and a possible mycotic infrarenal aneurysm. No immediate intervention was advised by vascular surgery or infectious disease specialists. A subsequent contrast-enhanced CT scan of the head raised concerns about a Slight Subarachnoid Hemorrhage (SAH), prompting further investigation via Magnetic Resonance Imaging (MRI). The MRI did not confirm SAH or a mycotic aneurysm but identified multiple areas suggestive of microhemorrhages and foci of enhancement consistent with septic emboli to the brain. Consequently, the patient's anticoagulation therapy was switched from warfarin to heparin under the guidance of neurology. Due to these complications, consideration was given to replacing the Prosthetic Mitral Valve (PMV). Following a thorough discussion of risks and benefits, a repeat Transesophageal Echocardiogram (TEE) was performed to reassess the previously reported vegetation. The TEE revealed either significant resolution or embolization of the vegetation, with only a small remaining echodensity observed on the valve. Following a comprehensive evaluation involving various medical specialties, including cardiology, cardiothoracic surgery, neurology, infectious disease, and radiology, a decision was made to pursue medical therapy with close monitoring due to the high risk associated with a repeat sternotomy. The patient successfully completed 8 weeks of treatment with oxacillin and rifampin, supplemented by 2 weeks of gentamicin during the initial phase of treatment, starting from the date of the last positive blood cultures.