

# Subaortic-Right Atrial Fistula after Endocarditis in Hypertrophic Cardiomyopathy

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Infective endocarditis in hypertrophic obstructive cardiomyopathy is rare. Management of this disease is challenging due to the unique features of dynamic pressure gradient over the left ventricular outflow tract and its unpredictable interaction with the management of sepsis. The added complexity of infective endocarditis further complicates an already difficult situation.

A 72-year-old man with hypertrophic obstructive cardiomyopathy presented with acute stroke, fever, and *Staphylococcus aureus* bacteremia. Infective endocarditis of the aortic valve was confirmed. Despite treatment with antibiotics and aortic valve replacement, the patient had recurrent bacteremia and developed a periannular abscess and a subaortic-right atrial fistula, with a resulting fatal outcome.

**Key Words:** Aortic valve replacement • Endocarditis • Hypertrophic obstructive cardiomyopathy • Subaortic-right atrial fistula

## INTRODUCTION

Infective endocarditis (IE) is a rare complication in hypertrophic cardiomyopathy (HCM) and tends to affect patients with basal outflow tract obstruction. The mitral valve (MV), aortic valve (AV), or both<sup>1,2</sup> may be involved, with vegetations attached to the ventricular surface of the MV, specifically the anterior leaflet, the AV cusps, and/or the left ventricle septal endocardium. Such vegetations might be seen during one or both phases of the cardiac cycle.<sup>2</sup> We report a case of AV endocarditis in a previously healthy hypertrophic obstructive cardiomyopathy (HOCM) patient, who presented initially with

an acute embolic stroke, had recurrent bacteremia, and unfortunately succumbed to severe complications and fatality.

## CASE REPORT

A 72-year-old man was diagnosed with HOCM with systolic anterior motion of the MV and a left urethral stone. He was referred to our emergency department due to persistent fever, chills, and dizziness, and was later in and out of consciousness. Brain computed tomography and magnetic resonance imaging revealed right middle cerebral artery ischemic infarction. Laboratory results showed a normal white blood cell count (7060/ul) with a mild left shift, c-reactive protein of 20 mg/dl, elevated troponin-I (1.5 ng/ml), but normal creatine-phospho-kinase/creatinine phosphokinase-MB, and pyuria. The patient underwent electrocardiographic testing, which revealed sinus tachycardia, left atrial enlargement, left axis deviation, right bundle branch block, and left ventricular hypertrophy with ST-T changes.

Blood and urine cultures grew Methicillin-resistant

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Staphylococcus aureus, and IE was highly suspected. Transthoracic echocardiography showed good left ventricular contractility, dilated left atrium chamber, markedly concentric left ventricular hypertrophy with systolic anterior motion of the MV, and moderate mitral regurgitation. The pressure gradient during systole across the left ventricular outflow tract (LVOT) was 68 mmHg, which was compatible with an obstructive pattern. Subsequent transesophageal echocardiography (TEE) proved the presence of a large vegetation ( $0.95 \times 0.9 \text{ cm}^2$ ) over the left coronary cusp of AV (Figure 1). Despite treatment with daptomycin, his fever and unstable hemodynamic persisted. Thereafter, the patient successfully received a tissue aortic heart valve replacement. During the operation, no abscess formation around the LVOT or the interventricular septum was observed. The patient regained consciousness, but remained with sequelae of the left hemiplegia and aphasia. He was transferred to a rehabilitation hospital after 6 months of Daptomycin treatment and complete resolution of bacteremia.

Unfortunately, the patient returned to our emergency department two weeks later, presenting with disturbances of consciousness, progressive dyspnea, and severe shock. Electrocardiography revealed complete atrioventricular block. Transthoracic echocardiography demonstrated good left ventricular contractility, dilated left and right atrial chambers, and a new, yet severe eccentric tricuspid regurgitation with poor visualization of the prosthetic AV. TEE was repeated and an aortic valve periannular hypodensity ring compatible with ab-

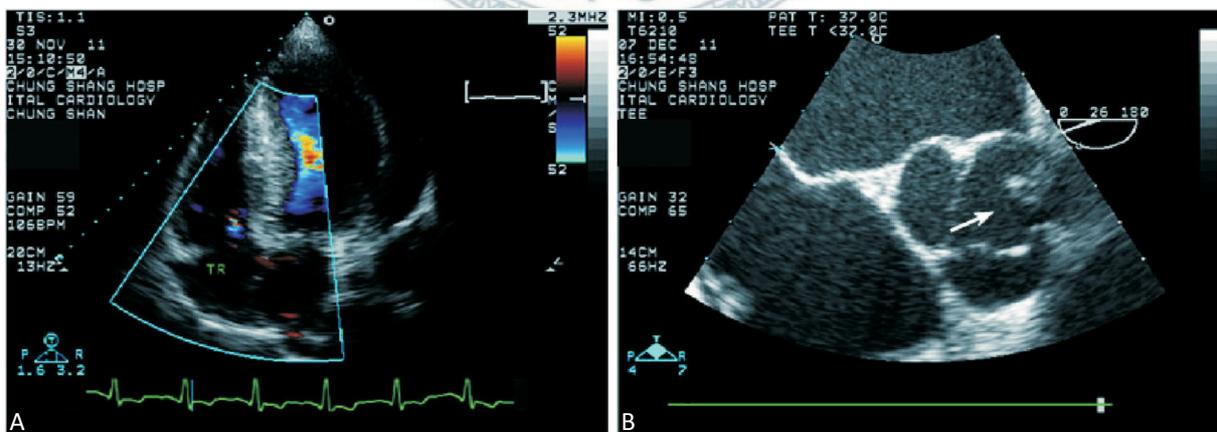
cess formation and leakage, and a subaortic to right atrial fistula mimicking severe tricuspid regurgitation were recorded (Figure 2). Vancomycin and imipenem were administered immediately, but the patient expired two days later. Blood cultures reported Methicillin-resistant Staphylococcus aureus.

## DISCUSSION

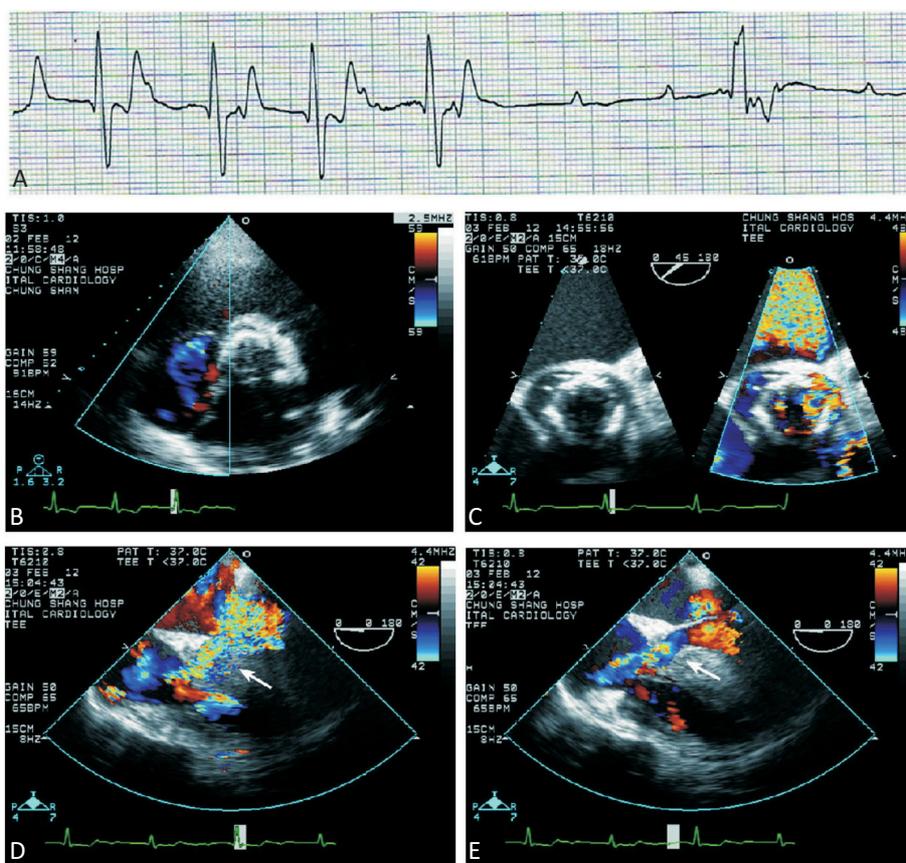
Infective endocarditis rarely complicates the typically benign course of adult HCM. The estimated incidence of IE in overall HCM cases is 1.4 per 1000 patient-years, with a higher incidence (3.8 per 1000 person-years) in patients with obstructive LVOT. The cumulative probability of endocarditis is 4.3% at 10 years in HOCM patients.<sup>1</sup>

The high flow velocity and the turbulence created by the narrowed LVOT results in mitral-septal contact, systolic anterior motion, and secondary mitral regurgitation. These environments cause recurrent micro-traumas to the MV, AV and septal endocardium, seeding microorganisms. The known risk factors associated with IE in HOCM are invasive procedures with blood exposure, LVOT obstruction, and a dilated left atrium.<sup>1</sup> Spirito et al. reported an increased incidence of IE (9.2 per 1000 person-years) in patients with both LVOT obstruction and a significantly dilated left atrium ( $> 5 \text{ cm}$ ).

Acquired communication between the LVOT and right atrium may develop after MV or AV replacement, endocarditis, periannular abscess, or chest trauma. The



**Figure 1.** Preoperative echocardiography at first admission. (A) Apical four-chamber view demonstrates only mild tricuspid regurgitation. A markedly thickened interventricular septum and left ventricular posterior wall (estimated 2.3 cm and 2.1 cm, respectively) are present. (B) Transesophageal echocardiography shows a large floating vegetation ( $0.95 \times 0.9 \text{ cm}^2$ ) located on the left coronary cusp of aortic valve.



**Figure 2.** Electrocardiography and echocardiography on following admission. (A) Complete atrioventricular block with variable conduction associated with shock status. (B) Transthoracic echocardiography shows a moderate to severe eccentric tricuspid regurgitation not seen preoperatively. (C) Transesophageal echocardiography demonstrates color flow Doppler surrounding the prosthetic aortic valve, compatible with periannular abscess and leakage. (D, E) A subaortic to right atrial fistula with to-and-fro flow during systole and diastole, mimicking tricuspid regurgitation.

fistulous communication is due to a defect in the interventricular septum, commonly inferior to the crista supraventricularis.<sup>3-5</sup> Patients may remain asymptomatic with small fistulas, but usually present with hemodynamic instability, prosthetic valve dysfunction, or periprosthetic leaks. A number of clinical parameters have been proposed to be associated with periannular and fistulous involvement; Blumberg et al. suggested that the presence of new atrioventricular blocks to be the most reliable parameter.<sup>6</sup> TEE is the modality of choice in assessing prosthetic valves and intracardiac pathology, such as periannular abscesses and fistulas. High sensitivity and specificity and good predictive values have been reported where TEE is used to evaluate these complications.<sup>7-8</sup> Prompt surgical repair is generally required to relieve the symptoms and decrease the mortality rate.

We present a case with *Staphylococcus aureus* endocarditis in HCM, complicated with a periannular abscess and a subaortic to right atrial fistula. Management of this condition can be complex and challenging. First, the basal and the dynamic pressure gradient of HOCM make the diagnosis and subsequent follow up for IE and prosthetic valve dysfunction based on auscultation very difficult, as the murmurs are quite complicated. Second, the dynamic LVOT pressure gradient may be induced by volume resuscitation and sympathomimetic agents, decreasing cardiac output and deteriorating hemodynamic dysfunction. Third, early TEE examination and aggressive surgical intervention are highly recommended in patients with recent valve replacement and unstable conditions. Lastly, antibiotic prophylaxis for IE is recommended for high risk valvular heart patients both by the American Heart Association and the European Society

of Cardiology, especially those with HOCM. Yet, in real clinical scenarios, antibiotic prophylaxis is seldom implemented.

## CONCLUSION

The virulent and unpredictable nature of *Staphylococcus aureus* infections and the associated risk factors and complications predict a poor outcome in cases of hypertrophic cardiomyopathy.

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