We report a previously healthy 21-year-old man who developed disseminated varicella zoster infection complicated with encephalitis, acute renal insufficiency, liver dysfunction, and an apparent pustular skin superinfection with Staphylococcus aureus. He later developed an extensively destructive endocarditis affecting a congenital bicuspid aortic valve, accompanied with leaflet perforation, complete atrio-ventricular (AV) block, and invasion of vegetation to both left and right atrium; the endocarditis was attributed to the same skin pathogen, S. aureus. He underwent radical debridement of the aortic valve, membranous ventricular septum, and mitral anterior fibrous trigone, followed by reconstruction of intracardiac defects with 2 autologous pericardial patches and aortic valve replacement. After a permanent pacemaker implantation and 4 weeks of antibiotic treatment, he was discharged after an uneventful postoperative course.

Keywords: endocarditis, bicuspid aortic valve, varicella zoster

Introduction

Varicella zoster infection is generally a benign, self-limited disease in children; however, serious secondary complications do occur. There are few anecdotal reports of endocarditis occurring after varicella infection, but all have reported pediatric cases.\(^1,2\) Here we report the case of a 21-year-old man who developed disseminated varicella zoster infection complicated with extensively destructive endocarditis affecting the congenital bicuspid aortic cusps, leaflet perforation, complete atrio-ventricular (AV) block, and invasion of vegetation to both the left and right atrium.

Case Report

A previously fit and healthy 21-year-old man developed fever and skin rash accompanied with headache, myalgia, and anorexia for 3 days before admission. The skin rash, which developed initially around his mouth and face and then progressed to his limbs and trunk, was compatible with the clinical presentation of a typical varicella infection. Direct fluorescent antigen testing of skin lesions corroborated the presence of varicella. Erythematous macules were initially clear vesicles that later got superinfected with Staphylococcus aureus, forming pustules. Laboratory data revealed acute renal insufficiency [creatinine level: 1.42 mg/dL, blood urea nitrogen (BUN) level: 29.9 mg/dL], liver dysfunction [total bilirubin level: 5.1 mg/dL, aspartate aminotransferase (AST) level: 164 U/L, alanine transaminase (ALT) level: 105 U/L], leukocytosis [white blood cells (WBC): 27800/µL], and thrombocytopenia (platelet count: 17000/µL). Brain
magnetic resonance imaging (MRI) was performed as the patient was drowsy; it revealed diffuse brain swelling with a right frontal hypodense lesion, compatible with the diagnosis of encephalitis (Fig. 1A). High fever with a body temperature of 40°C and symptoms of heart failure deteriorated despite intravenous acyclovir and vancomycin treatment for 1 week. Positive blood cultures of *S. aureus*, electrocardiographic finding of complete AV block, and the findings of transesophageal echocardiography indicated extensively destructive endocarditis affecting the congenital bicuspid aortic valve complicated with leaflet perforation and paravalvular abscess invasion to both left and right atrium (Fig. 1B).

After an angiogram that excluded intra-cranial mycotic aneurysm, he underwent an urgent operation because of uncontrolled sepsis, frank heart failure, paravalvular abscess, and the new onset of complete AV block. The sinus of Valsalva was intact, and the aortic root was normal; however, abnormalities in other structures were identified: the congenital bicuspid aortic valve with cusp perforation and 2 vegetations under the valve, 1 invading the mitral anterior fibrous trigone with a fistula to the left atrium (Fig. 2A) and the other penetrating the membranous septum to Koch’s triangle of the right atrium (acquired Gerbode defect) (Fig. 2B). The infected tissues, including aortic valve, mitral anterior fibrous trigone, membranous ventricular septum, and Koch’s triangle, were radically resected. Separate defects from the left ventricular outflow tract to the left atrium and to the right atrium were reconstructed with 2 autologous pericardia, followed by implantation of a mechanical valve, which was used considering his young age. He received permanent pacemaker implantation 1 week after surgery, completed 4 weeks of antibiotics, and was discharged after an uneventful postoperative course. Postoperative echocardiography showed neither prosthetic valve endocarditis nor residual intra-cardiac shunt.

**Discussion**

Infection with varicella zoster virus, which causes chicken pox, is usually a benign self-limiting disease; however, severe life-threatening cardiac complications do occur. Most of these are pediatric cases, and complications are limited to pericarditis or myocarditis, but in some extreme cases, the infection may progress to endocarditis. The exact mechanism by which varicella causes secondary bacterial endocarditis remains unclear; superinfection of skin pustules with a more toxic strain of *S. aureus*, and spread of infection to the vulnerable congenital bicuspid aortic valve could be a possible mechanism and can be the cause of the worsening of the condition in our patient.

Compared to a normal tricuspid aortic valve, a congenital bicuspid aortic valve is more susceptible to infective endocarditis and has a strikingly higher rate of AV
block and ventricular septal defects with subvalvular complications.\textsuperscript{5,6} Although antibiotic treatment of chicken-pox-induced endocarditis has been reported to be effective,\textsuperscript{7} surgical intervention should not be delayed if a high degree AV block, paravalvular abscess, or congestive heart failure caused by valve insufficiency is encountered, especially in cases of endocarditis caused by \textit{S. aureus} infection of the valves in the left side of the heart.\textsuperscript{8}

Surgery for active infective endocarditis with paravalvular abscess continues to be a challenge and is associated with high operative mortality and morbidity. Recognition of the extent of infection and radial resection of the abscess are essential and keys to the prevention of recurrence after operation. In our case, autologous pericardial patches obliterated both intracardiac defects and made aortic valve replacement feasible as was reported previously.\textsuperscript{8} Although a homograft is ideally used for extensive endocarditis, we chose mechanical valve replacement because of the patient’s young age.

To the best of our knowledge, this is the only documented case of an adult patient with bicuspid aortic valve endocarditis attributable to primary varicella zoster infection superinfected with \textit{S. aureus}, which resulted in a complete AV block and intracardiac fistula. Protracted fever or dyspnea, which is unexpected in typical cases of varicella clinically, must be aggressively pursued, especially if signs of acute cardiac decompensation develop.

\textbf{References}