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가성동맥류를 동반한 *Haemophilus parainfluenzae* 인공판막 심내막염

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A Case of Prosthetic Valve Endocarditis with Pseudoaneurysm Formation Caused by *Haemophilus parainfluenzae*

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Prosthetic valve endocarditis (PVE) caused by *Haemophilus parainfluenzae (H. parainfluenzae)* is very rare. Here, we report a case of *H. parainfluenzae* PVE that developed following the Bentall procedure complicated by a pseudoaneurysm and cerebral emboli. A diagnosis was delayed in this case because of the slow-growing nature of the organism and the unusual clinical presentation. (Korean J Med 2014;87:589-592)

Keywords: Aneurysm, False; Endocarditis; *Haemophilus parainfluenzae*; Heart valve prosthesis

INTRODUCTION

Haemophilus parainfluenzae (H. parainfluenzae), a rare cause of infective endocarditis, is responsible for only about 1% of all causes of endocarditis in adults [1]. Pseudoaneurysm formation associated with infective endocarditis is a relatively infrequent but potentially lethal complication. The most frequent site of pseudoaneurysm formation is the mitral-aortic interventricularfibrosa (MAIVF), and its predisposing factors are infective endocarditis of the aortic valve or aortic valve surgery [2]. Here, we describe a case of pseudoaneurysm formation associated with an infected artificial graft following the Bentall procedure caused by *H. parainfluenzae*.

CASE REPORT

A 77-year-old male was hospitalized with acute chest pain that had started three hours earlier. His medical history was not

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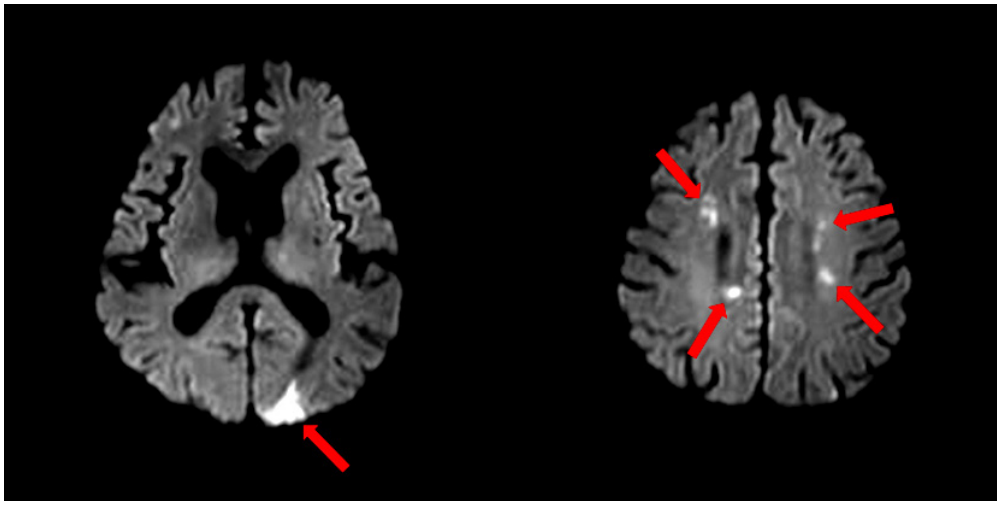

Figure 1. Brain magnetic resonance imaging showing multifocal enhancing lesions (red arrows) in both hemispheres, suggesting embolic infarction.

significant. He was diagnosed with an acute type A aortic dissection and underwent emergency surgery successfully with replacement of both the ascending aorta and aortic valve (the Bentall procedure). Although his chest pain improved after surgery, the patient complained of sustained general weakness and ill-defined malaise. Serial blood cultures and transthoracic echocardiography (TTE) showed no abnormal findings during the postoperative period. Four months after surgery and during hospitalization, the patient became drowsy and developed a high grade fever. His blood pressure dropped to 79/49 mmHg, his body temperature was 39.1ºC, and his white blood cell count was 13,500/mm^3 with 91% neutrophils. The patient's C-reactive protein level was markedly elevated to 17.84 mg/dL. Brain magnetic resonance imaging demonstrated multiple small enhancing lesions in both cerebral hemispheres, suggestive of embolic infarction (Fig. 1). Under strong suspicion of infective endocarditis causing cerebral embolization, TTE and subsequent transesophageal echocardiography (TEE) were performed. The prosthetic aortic valve functioned well and had no visible vegetations. However, a pseudoaneurysm was noted in the region of the MAIVF (Fig. 2). Surgical correction was not considered because of high operative risk and the family's refusal. Blood samples for culture were obtained, and treatment was initiated with vancomycin and cefepime. Six days later, Gram-negative bacteria, which were later identified as *H. parainfluenzae*, were detected in all three sets of blood cultures. The antibiotic regimen was changed to cefotaxime. Despite treatment, progressive multiorgan failure developed and the patient died on day six of TEE.

DISCUSSION

H. parainfluenzae, which is part of the normal flora of the nasopharynx, has been implicated as a rare cause of infective endocarditis. *H. parainfluenzae* is a slow- growing organism, and blood cultures do not usually become positive within 7 days of sample collection [3]. Similarly, the onset of *Haemophilus* endocarditis is most often subacute, the symptoms often develop insidiously over several weeks, and the time to diagnosis varies widely [1]. Systemic embolization is another characteristic of *Haemophilus* endocarditis. It has been reported that arterial emboli occur frequently in up to 60% of patients with *Haemophilus* endocarditis. Although embolic infarction and the deposition of immune complexes can occur everywhere, including the spleen, kidneys, coronary vessels, and cerebrovascular system, embolization to the central nervous system is a major cause of morbidity and mortality in patients with *H. parainfluenzae* endocarditis [1,4,5]. Some of these characteristics were seen in our case: (1)

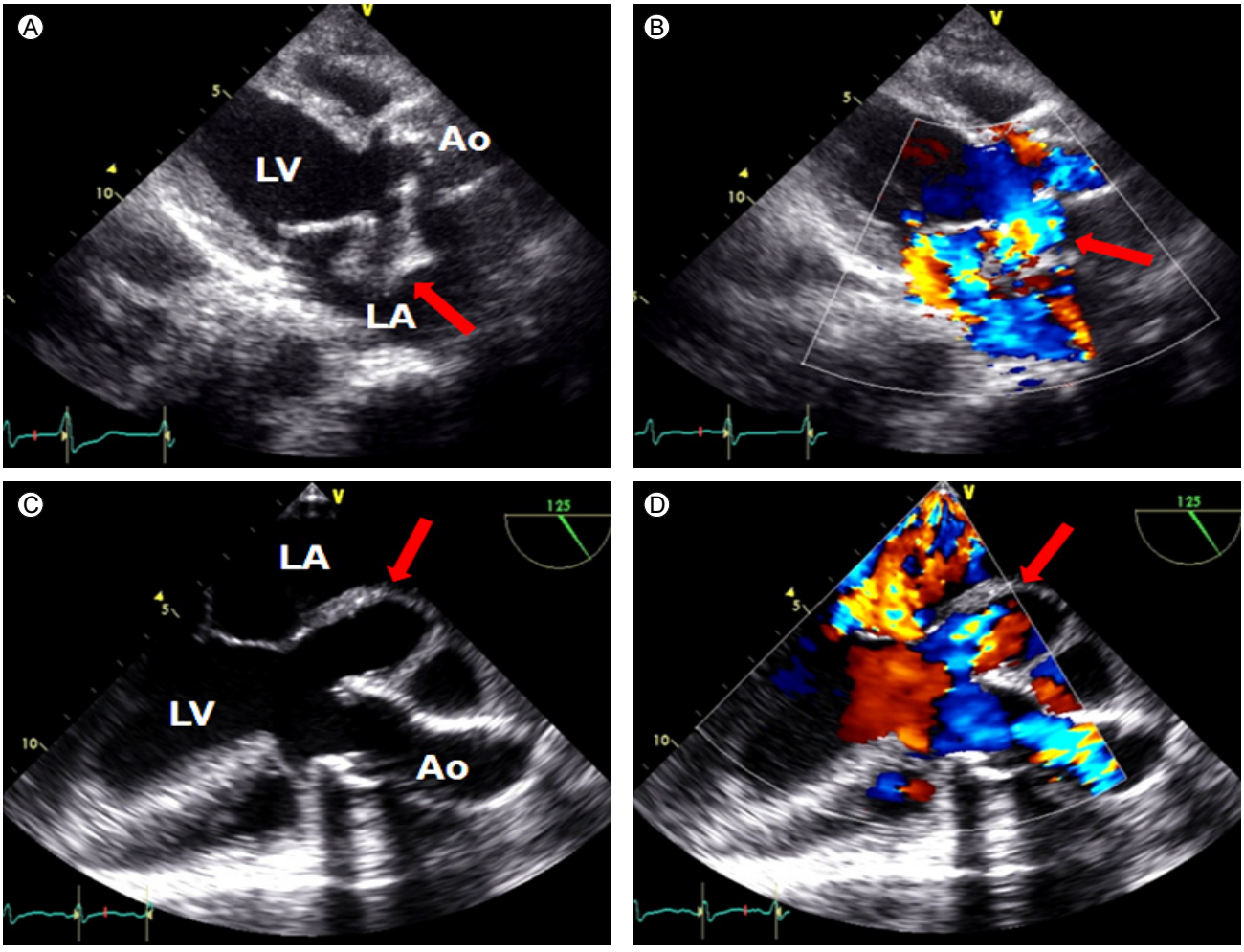

Figure 2. Echocardiography revealed a pseudoaneurysm (red arrows) with a flow-communicating pulsatile cavity in the region of the mitral-aortic interventricularfibrosa. Transthoracic echocardiograms (parasternal long axis view) without (A) and with color Doppler (B), and transesophageal echocardiograms without (C) and with (D) color Doppler. LV, left ventricle; Ao, aorta; LA, left atrium.

the initial clinical presentation was indolent with symptoms of general weakness and malaise extending over several months, (2) the pathogen was identified after 6 days in blood cultures, and (3) *Haemophilus* endocarditis was complicated by cerebral embolic infarction. Given this unusual clinical presentation, the diagnosis of *Haemophilus* endocarditis was difficult.

Haemophilus species are a rare cause of prosthetic valve endocarditis (PVE). There are few case reports on PVE caused by *H. parainfluenzae.* Liang et al. [6] reported the case of a 51-year-old male with *H. parainfluenzae* PVE complicated by septic emboli in the brain. He had a prosthetic aortic valve, and a large vegetation was detected in his mitral valve. He complained of a headache and right-side weakness due to embolic infarction to the brain. He recovered successfully after aortic and mitral valve replacement. Blair and Weiner [7] reported the case of a 14-year-old female who underwent prosthetic mitral valve replacement due to congenital heart disease. Her prosthetic valve was infected with *H. parainfluenzae* and complicated by emboli to the femoral artery and congestive heart failure. A complete cure was achieved after prosthetic valve replacement. Choi et al. [8] reported the case of a 42-year-old male intravenous drug abuser with *H. parainfluenzae* mitral PVE. He was cured with antibiotic therapy without valve replacement. Although the clinical presentation in our case is not

significantly different from that in previous reports, our report is unique for several reasons. First, it demonstrates a local complication of PVE by *H. parainfluenzae*. Second, pseudoaneurysm formation in the prosthesis was noted after the Bentall procedure, given the good quality of the echocardiographic images.

The MAIVF is a thin fibrous structure between the aortic and mitral valves where the anterior mitral leaflet connects with the non-coronary cusp of the aortic valve [2]. This subaortic structure can be infected as a complication of aortic valve endocarditis. The MAIVF is an avascular structure and prone to infection. The direct extension of inflammation from the aortic valve to the MAIVF is common. Less frequently, an infected aortic regurgitating jet flow striking the MAIVF causes damage to the area and makes it vulnerable to infection [9]. For these reasons, involvement of the MAIVF has been reported in 44% of patients with aortic valve endocarditis [9]. Pseudoaneurysm formation in the MAIVF is a rare complication that has been reported to be a sequela of infective endocarditis of the aortic valve, aortic valve surgery, or chest trauma. This complication can be fatal because of the risk of rupture, especially when the pseudoaneurysm is large, and surgical correction is usually warranted as soon as the diagnosis is made [2]. In our case, however, surgery was not performed because the surgical risk was high and because the patient's family refused surgery.

The diagnostic value of TTE is limited in the detection of vegetations and complications associated with infective endocarditis. It has been reported that the recognition of a pseudoaneurysm by TTE is possible only in about half of all patients [2,9]. TEE is superior to TTE in the visualization of vegetations, abscesses, fistulas, and pseudoanuerysms associated with infective endocarditis. Its sensitivity and specificity are 100 and 83%, respectively, in identifying complications of infective endocarditis [10]. In our case, the image produced by TTE was good, and we were able to identify the pseudoaneurysm easily and to confirm it by TEE.

To the best of our knowledge, our patient is the first case of *H. parainfluenzae* PVE following the Bentall procedure that was complicated by a pseudoaneurysm and cerebral emboli. *H. parainfluenzae* PVE is rare, and the slow-growing nature of the microorganism may delay an accurate diagnosis. Thus, there may be life-threatening complications such as rupture of the pseudoaneurysm and cerebral emboli. Therefore, attention should be paid to *Haemophilus*-mediated infective endocarditis, especially in patients with prosthetic valves who present with an unusual clinical course or metastatic infection.

중심 단어: 가성동맥류; 심내막염; *Haemophilus parainfluenzae*; 인공판막

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