

CASE REPORT

BEGINNER

CLINICAL CASE

# Infective Endocarditis Complicated by Pseudoaneurysm of the Mitral-Aortic Intervalvular Fibrosa Without Valvular Involvement



Miho Kuroda, MD,<sup>a</sup> Hiroyuki Yamamoto, MD, PhD,<sup>b</sup> Yoshitsugu Nakamura, MD, PhD<sup>a</sup>

## ABSTRACT

A pseudoaneurysm of the mitral-aortic intervalvular fibrosa (p-MAIVF) can be a catastrophic sequela of untreated active infective endocarditis. We describe a case of infective endocarditis complicated by p-MAIVF without valvular involvement. This case highlights the importance of this rare clinical entity and of transesophageal echocardiography in reaching a diagnosis. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:1212-6) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## HISTORY OF PRESENTATION

A 20-year-old man with fever and headache was admitted to Chiba-Nishi General Hospital, Matsudo, Japan. On admission, physical examination results were as follows: blood pressure, 81/49 mm Hg; high-grade fever, 40.0°C; tachycardia, 94 beats/min; and grade 4/6 systolic murmur. Motor aphasia was observed on neurological examination.

## PAST MEDICAL HISTORY

No previous history of infective endocarditis (IE) or cardiac surgery was revealed in the anamnesis.

## LEARNING OBJECTIVES

- To understand the cause of IE.
- To understand the complications and imaging findings associated with p-MAIVF.

## DIFFERENTIAL DIAGNOSIS

Septic embolism, mycotic aneurysm, brain abscess, and meningoencephalitis were considered.

## INVESTIGATIONS

A chest radiograph revealed cardiomegaly with pulmonary congestion. Laboratory test results revealed neutrophilic leukocytosis and elevated C-reactive protein levels. Transthoracic echocardiography (TTE) revealed a left atrial heterogeneous mass with vegetation adjacent to the posterior part of the aortic root. An eccentric supra-annular mitral regurgitation jet (**Figure 1A**, **Video 1**) and a bicuspid aortic valve (BAV) and inhomogeneous mass on the posterior side of the aortic valve (AV) annulus were observed, suggesting AV ring abscess (**Figure 1B**). On the basis of multiple blood cultures, positive findings of methicillin-

From the <sup>a</sup>Department of Cardiovascular Surgery, Chiba-Nishi General Hospital, Matsudo, Japan; and the <sup>b</sup>Department of Cardiovascular Medicine, Narita-Tomisato Tokushukai Hospital, Chiba, Japan. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Case Reports* [author instructions page](#).

Manuscript received March 31, 2020; revised manuscript received April 28, 2020, accepted May 12, 2020.

susceptible *Staphylococcus aureus* were obtained, thereby fulfilling the modified Duke criteria for a diagnosis of IE. Brain computed tomography revealed hemorrhagic infarctions in the right frontal lobe, suggesting systemic embolism (Figure 2).

### MANAGEMENT

Treatment with broad-spectrum antibiotics, including vancomycin and cefazolin, was initiated; however, the patient's clinical condition rapidly deteriorated after 2 days because of heart failure exacerbation. Subsequent transesophageal echocardiography (TEE) demonstrated rapidly growing vegetations in the left atrium (LA), rapid pericardial effusion accumulation (Figure 3A, Video 2), and a pulsatile echo-free space corresponding to expansion at systole and collapse at diastole between the left ventricular outflow tract (LVOT) and the LA (Figures 3B and 3C, Video 3). Color flow imaging revealed a turbulent jet into the LA during systole and reverse flow into the LVOT during diastole. On the basis of this finding, a diagnosis of IE complicated by the rupture of a pseudoaneurysm of the mitral-aortic intervalvular fibrosa (p-MAIVF) into the LA was made. This diagnosis was confirmed using contrast-enhanced multislice computed tomography (Figure 4). Mild aortic regurgitation with an eccentric posterior jet targeting the mitral-aortic intervalvular fibrosa (MAIVF) was noted; however, no obvious

vegetation was observed in the BAV (Figure 3C, Video 3).

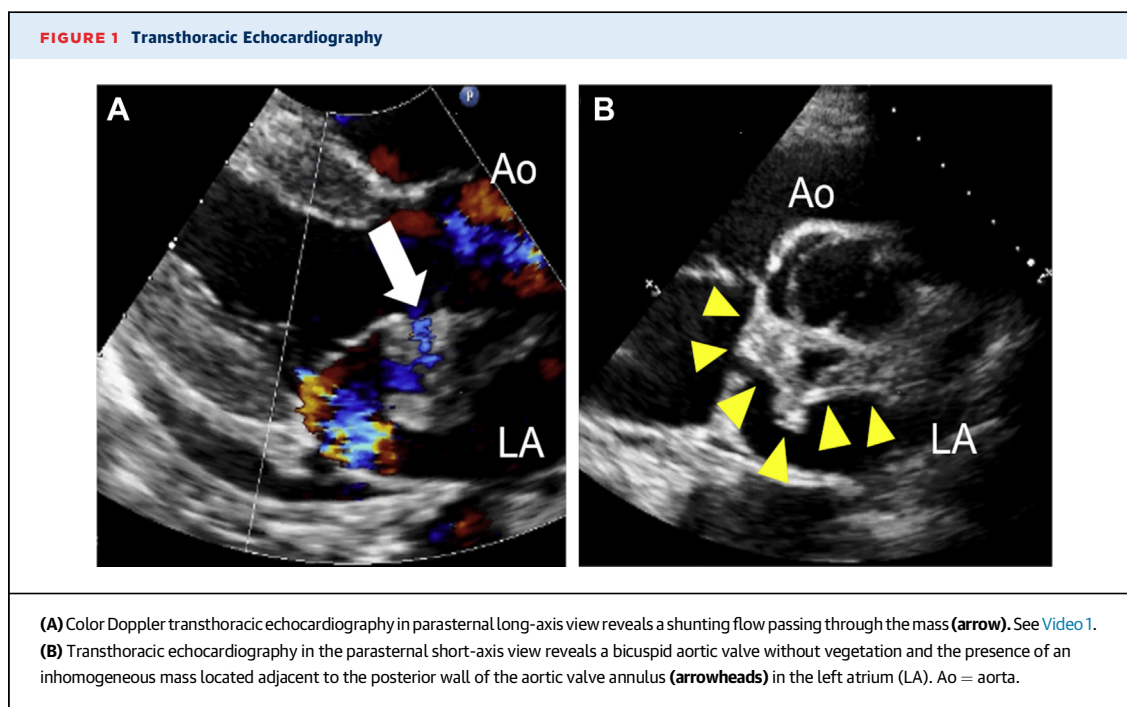
Semiurgent surgery performed by a multi-disciplinary team was planned after confirming no worsening of the patient's neurological deficits on day 3. The procedure consisted of complete abscess debridement with vegetation removal and closure by a Dacron (DuPont, Wilmington, Delaware) patch of p-MAIVF that showed a 15-mm perforation between the LVOT and the LA (Figures 5A and 5B). Surgical drainage was required for approximately 500 ml purulent hemorrhagic pericardial fluid after p-MAIVF rupture into the pericardium. However, no vegetation was observed in any BAV leaflets, the sinus of Valsalva, and mitral valve leaflets. Bacterial infection was confirmed through pathological examination. Long-term antibiotic therapy, including intravenous vancomycin followed by oral linezolid, was administered.

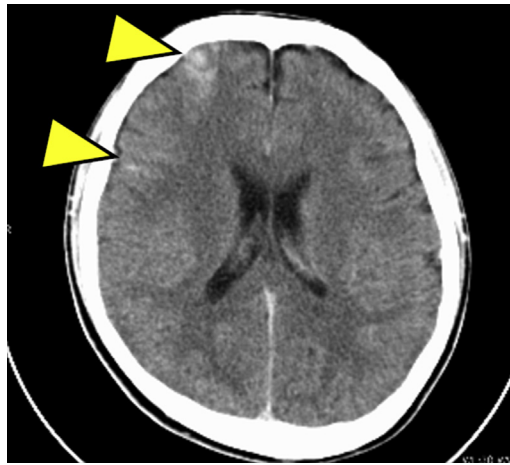
### DISCUSSION

MAIVF, an avascular structure adjacent to the LVOT between the mitral valve and the AV, is necessary for maintaining functional and anatomic integrity (1). Secondary p-MAIVF is a potentially life-threatening sequela of active or previous valve endocarditis and surgical trauma, which causes compression or fistula

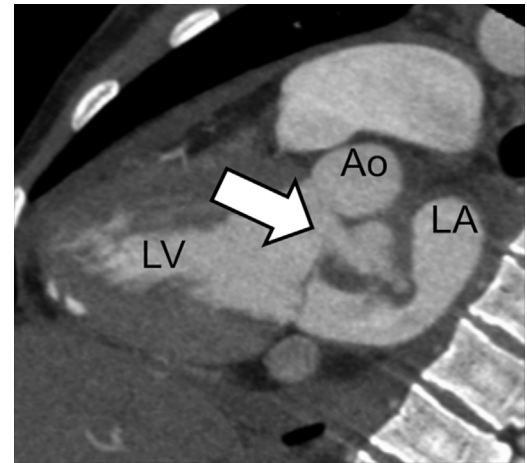
### ABBREVIATIONS AND ACRONYMS

- AV = aortic valve
- BAV = bicuspid aortic valve
- IE = infective endocarditis
- LA = left atrium
- LVOT = left ventricular outflow tract
- MAIVF = mitral-aortic intervalvular fibrosa
- p-MAIVF = pseudoaneurysm of the mitral-aortic intervalvular fibrosa
- TEE = transesophageal echocardiography
- TTE = transthoracic echocardiography



**FIGURE 2** Brain Computed Tomography

Noncontrast brain computed tomography showing multiple areas of hyperdensity (**arrowheads**) in the right frontal lobe.

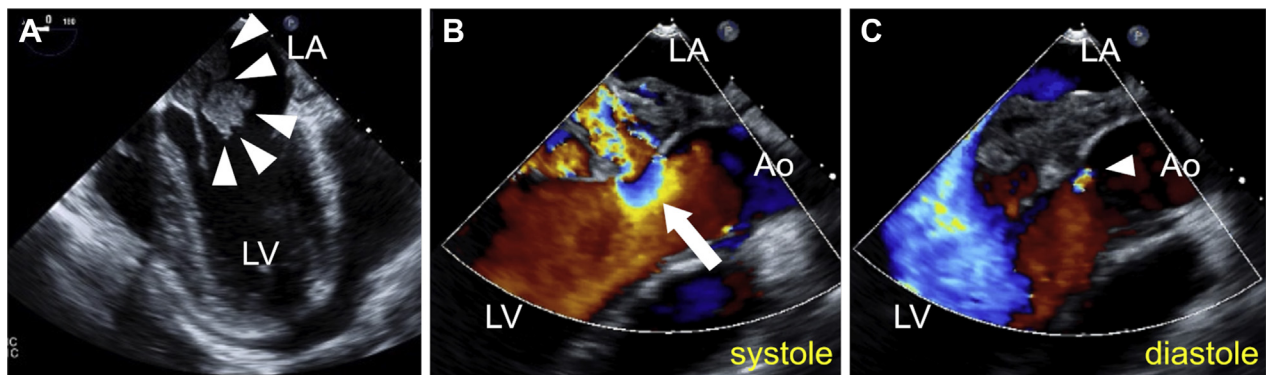
**FIGURE 4** Contrast-Enhanced Multislice Computed Tomography

Contrast-enhanced multislice computed tomography shows a pseudoaneurysm originating from the mitral-aortic intervalvular fibrosa compressing the left atrium (LA) (**arrow**) and the pericardial fluid. Abbreviations as in [Figure 3](#).

formation at adjacent cardiac and vascular structures (2,3). However, the correct diagnosis in the absence of these factors is extremely difficult. Our case may provide several clinical lessons.

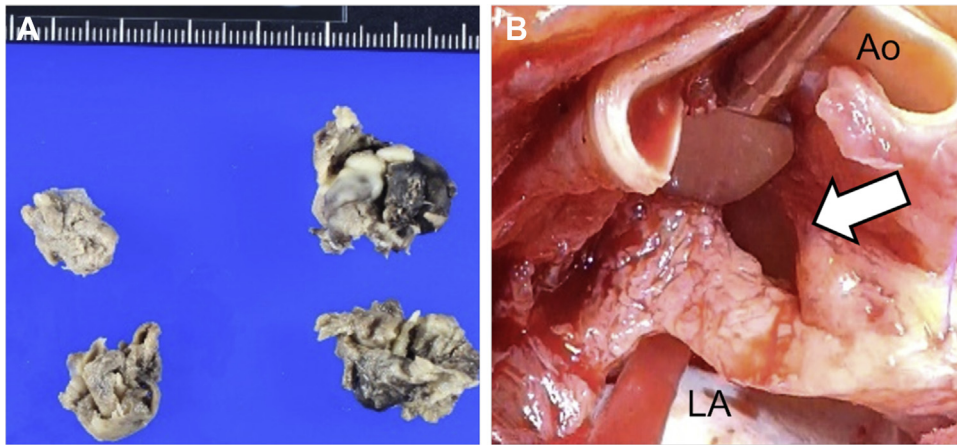
First, we report a case of IE complicated by p-MAIVF without BAV involvement. p-MAIVF frequently manifests with signs of active IE, followed by heart failure (2). AV IE is the most common causative factor in p-MAIVF because infectious insults

may allow direct or indirect damage to the MAIVF. BAV is associated with p-MAIVF in terms of inherent vulnerability (4), and it is also at high risk for IE (5). Thus, patients with BAV IE may be at a higher risk of p-MAIVF. Nevertheless, BAV was not involved in IE in our case, a situation that is very rare. The number of diagnoses of p-MAIVF has been steadily increasing in

**FIGURE 3** Transesophageal Echocardiography

(A) Transesophageal echocardiography in the midesophageal 4-chamber view reveals multiple vegetations (**arrowheads**) and a large pericardial effusion. See [Video 2](#). (B and C) Color Doppler transesophageal echocardiography in the midesophageal 3-chamber view shows a unique pulsatile echo-free space (**arrow**) corresponding to expansion with antegrade flow at systole (B) and collapse with retrograde flow at diastole (C), thus indicating the mitral-aortic intervalvular fibrosa fistula between the left ventricular outflow tract and the left atrium (LA). Note the mild posterior aortic regurgitation jet directly hitting the mitral-aortic intervalvular fibrosa (**arrowhead**). See [Video 3](#). Ao = aorta; LV = left ventricle.

**FIGURE 5** Intraoperative Findings



**(A)** The surgically removed vegetations. **(B)** Intraoperative view through the opened left atrium (LA) and aortotomy demonstrates a perforation between the mitral-aortic intervalvular fibrosa and the left atrium (**arrow**). Ao = aorta.

response to increasing awareness of the disease and the use of various imaging modalities, thus suggesting that this condition is no longer rare (2). This case highlights the clinical importance of considering p-MAIVF as a source of IE, even in the absence of valvular endocarditis.

Second, our case of p-MAIVF revealed infections extending through the pericardium as well as the LA. p-MAIVF can have a variable presentation, from asymptomatic to fatal complications. The most frequent complications of p-MAIVF are fistula formation and coronary artery compression. Although it is extremely rare, rupture into the pericardium is among the most feared and catastrophic complications (2,6). Thus, prompt diagnosis and treatment of this complication are required. However, because of its similar clinical signs, it is difficult to distinguish from pericardial effusion associated with heart failure, and the appropriate time for surgery is often missed. As in our case, rapid pericardial effusion accumulation may be the key to identifying this serious complication.

Third, TEE was useful for correct diagnosis and appropriate surgical planning. TEE is of exceptional diagnostic value in detecting p-MAIVF compared with TTE (sensitivity, 90% and 43%, respectively) (7). In our patient, the unique prominent pulsation comprising systolic expansion with antegrade flow and diastolic collapse with retrograde flow observed on TEE allowed differentiation from an AV ring abscess and enabled the correct diagnosis. Development of p-MAIVF may occur even if active valve endocarditis is not apparent. Therefore, cardiac evaluation

using TEE should be recommended for patients with heart failure who have eccentric mitral regurgitation jets on TTE. TEE can also provide more detailed information about the location, mechanism, and complications of p-MAIVF (7). This proved vital for our young patient with potentially fatal complications. A less invasive surgical approach was preferred in this patient to avoid the long-term warfarin treatment necessary for valve replacement. Pre-operative TEE was helpful in performing valve-sparing surgery.

#### FOLLOW-UP

The post-operative course was uneventful, and the patient has remained symptom-free during follow-up.

#### CONCLUSIONS

We describe a case of p-MAIVF related to nonvalvular IE. P-MAIVF may be an underdiagnosed yet potentially life-threatening condition that is curable if treated properly at an early stage. TEE is useful for early diagnostic testing and evaluation of appropriate surgical treatment strategies. Therefore, clinicians should recognize the clinical importance of this rare subset of IE and should not hesitate to perform TEE.

**ADDRESS FOR CORRESPONDENCE:** Dr. Hiroyuki Yamamoto, Department of Cardiovascular Medicine, Narita-Tomisato Tokushukai Hospital, 1-1-1 Hiyoshidai, Tomisato, Chiba 286-0201, Japan. E-mail: [hyamamoto19700908@gmail.com](mailto:hyamamoto19700908@gmail.com).

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**KEY WORDS** acute heart failure, bicuspid aortic valve, endocarditis, mitral-aortic intervalvular fibrosa, pseudoaneurysm, transesophageal echocardiography

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**APPENDIX** For supplemental videos, please see the online version of this paper.