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EDITORIAL COMMENT

Mitral-Aortic Intervalvular Fibrosa

A Hidden Region Associated With Infective Endocarditis Complications*

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In this issue of *JACC: Case Reports*, Kuroda et al. (1) report a complex case of a young man with a bicuspid aortic valve (BAV), who presented with infective endocarditis (IE) and a pseudoaneurysm of the mitral-aortic intervalvular fibrosa (p-MAIVF) without valvular involvement. His clinical picture was noteworthy for fever, motor aphasia, and rapidly progressive heart failure. The remarkable findings on transthoracic echocardiography (TTE) were a left atrium (LA) heterogeneous mass with vegetation adjacent to the aortic root, an eccentric supra-annular mitral regurgitation jet, and a mass on the aortic valve (AV) annulus suggesting AV ring abscess. Subsequent transesophageal echocardiography (TEE) showed findings suggesting rupture of a p-MAIVF into the LA and rapid pericardial effusion accumulation.

This interesting case presents an opportunity to briefly make some comments on challenges related to mural endocarditis, myocardial abscesses, and p-MAIVF. First described in 1924, mural endocarditis involving nonvalvular endocardium is rare, especially in the absence of predisposing factors (2). Clinically, it is very similar to valvular endocarditis and sophisticated cardiovascular imaging techniques are required to differentiate these 2 conditions. Although right and left ventricular free walls and apices are the most frequently involved sites, mural vegetation can develop in any cardiac chamber (2–4). More commonly, nonvalvular IE occurs secondary to infected mural thrombus, intracardiac devices or prostheses, cardiac tumors, structural abnormalities including congenital defects, or valvular IE (2–5). Less commonly, it may be primary (3) or results from extension of infection from underlying myocardial abscesses (6). Potential complications of mural endocarditis include systemic embolization, abscess and fistula formation, papillary muscle or chordae damage, and cardiac perforation (3).

Myocardial abscesses develop either after bacteremia or fungemia or from direct extension from valvular or mural IE. Patients at greatest risk from hematogenous seeding comprise those with prolonged hospitalizations, prolonged use of antibiotics, or with indwelling venous catheters. The abscess can rupture resulting in cardiac tamponade, hemopericardium, hemorrhagic or purulent pericarditis; fistulize into the heart chambers; cause arrhythmias and conduction defects; and extend into the endocardium causing IE or extend throughout the myocardium causing diffuse myocarditis (6).

p-MAIVF is a rare complication of endocarditis and surgical trauma in the MAIVF area (7); however, BAV has also been associated with this condition (7,8). This entity is usually asymptomatic in the absence of complications. The most common forms of presentation include signs of infection, heart failure, and cerebrovascular events.

In this case, it is interesting to speculate where the infectious process started and how it progressed. The patient presented on TEE with mild aortic regurgitation with an eccentric posterior jet targeting the MAIVF. The direct impact of high-velocity regurgitant jet can cause endothelial disruption at its point of impact, resulting in platelet and fibrin deposition that can serve as a

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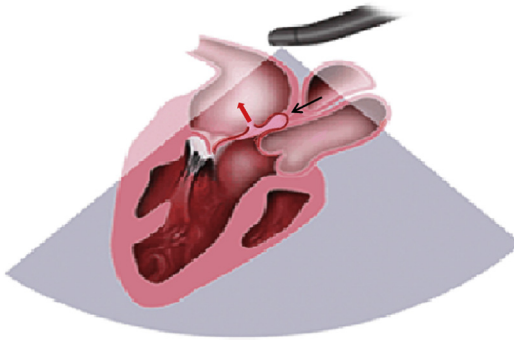
FIGURE 1 Long-Axis Transesophageal Echocardiography View Showing Mitral-Aortic Intervalvular Fibrosa Pseudoaneurysm With Fistula to the Left Atrium



Fistula between the left ventricular outflow tract and left atrium, with blood flow coming in the left atrium (**red arrow** = blood flow). The pseudoaneurysm is shown by the **black arrow**.

nidus for bacterial infection (9,10). Thus, the mild aortic regurgitation presented by the patient could be the source of the cardiac infection starting with mural IE of the MAIVF on the site of the jet impact, which complicated with MAIVF abscess that drained into the LA through a fistulous path, leading to a p-MAIVF, which in turn ruptured into the pericardium. The fact that the vegetation was located in the LA points against this possibility. Alternatively, as emphasized in the case reported (1), the MAIVF abscess could have been the primary site of the cardiac infection, which subsequently drained into the LA. The lack of predisposing factor for myocardial abscesses argues against this possibility. Finally and much less likely, the p-MAIVF could be associated with the BAV, as described by some authors (7,8), and have become infected during bacteremia, thus originating the entire cardiac infectious condition. As the diagnosis of the cardiac infectious process of the patient was defined late in its course, it is difficult to establish the sequence of events in this case.

It is important to emphasize the crucial role of echocardiography for reaching a rapid diagnosis in cases of suspected IE (11). TTE remains a first-line imaging modality for clinically suspected IE. As highlighted in this case, TTE revealed vegetation adjacent to the aortic root, which allowed the diagnosis. TTE also provides value in the assessment of severity of valve lesions and ventricular function (11). Although in the present case AV ring abscess was suggested by TTE, TEE is indicated when abscess or other IE complications are suspected.

Pseudoaneurysm is a rare well-known complication of IE, which consists of a perivalvular cavity that communicates with the cardiovascular lumen (7). It is commonly secondary to ring abscess in the region of the MAIVF, a relatively avascular area that is prone to infection and injury resulting in pseudoaneurysm formation (**Figure 1**). The periaortic spread of the infection is a dynamic process in which the inflammation of the deep tissue causes, in a first stage, a MAIVF thickening, which eventually progresses with the formation of an abscess, and subsequently, a pseudoaneurysm. There has been an increasing recognition of this entity as TEE has come into routine use (10). Complications of p-MAIVF include rupture into the left atrium, aorta, or pericardial space leading to hemopericardium, tamponade, and death (7). As reported in the present case, TEE is crucial to accurate p-MAIVF diagnosis by demonstrating systolic expansion and diastolic collapse of the pseudoaneurysm. Color flow mapping can yield up the site of rupture or fistulization of the p-MAIVF, which is essential to guide surgical management. More recently, other imaging modalities are emerging to help the diagnosis in cases in which TEE is nondiagnostic or to provide additional information on management of IE (12).

Well-established guidelines for treatment of valvular endocarditis recommend early surgical intervention when clinically appropriate (13). However, it is not known whether this approach should also be used in mural endocarditis. The paucity of data on mural endocarditis limits the recommendation of therapeutic strategies (3,6). Surgery is currently the recommended treatment for myocardium abscess and for p-MAIVF (6–8).

This rare case illustrates the challenge that IE can pose, the key role of TEE in the diagnosis of its special forms and complications, and the importance of close cooperation between medical and surgical disciplines to achieve greater therapeutic success. Early recognition of these life-threatening complications is of crucial importance, as urgent surgical approach is necessary.

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