

CASE REPORT

Uncommon Ultrasound Findings in a Patient with Extensive Bicuspid Aortic Valve Endocarditis

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regurgitation*

ABBREVIATIONS

LV = left ventricle
MAIVF = mitral aortic intervalvular
fibrosa
RA = right atrium
TEE = transesophageal echocardiogram
TTE = transthoracic echocardiogram

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ABSTRACT

We report the case of a 32-year-old man, who presented with dyspnea and diffuse abdominal pain with associated vomiting about two months after surgery for aortic valve replacement due to endocarditis. The echocardiographic examination of the patient revealed uncommon findings, such as pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF), perforation of the anterior mitral leaflet, large aneurysm of the perimembranous inlet ventricular septum, Gerbode flow through the perimembranous aneurysm and finally abnormal position of the tricuspid valve towards the ventricular apex.

INTRODUCTION

A left ventricle-to-right atrium communication is either congenital or acquired as the result of endocarditis, valvular surgery, myocardial infarction or trauma. This type of communication, albeit rare, is anatomically possible, because normally the tricuspid valve is apically displaced relative to the mitral valve. Echocardiography, particularly via the transesophageal approach combined with color flow Doppler, is pivotal in rendering a correct diagnosis. We herein present such a case which probably occurred from ventricular septal perforation during surgery of aortic valve replacement.

CASE REPORT

A 32-year-old man was admitted to the Department of Cardiology with complaints of diffuse abdominal pain and vomiting, as well as dyspnea, over the preceding two days. He complained of malaise and poor appetite for a week. Patient's past medical history was significant for recent hospitalization due to acute pulmonary edema as a complication of severe aortic regurgitation due to bicuspid aortic valve endocarditis. Blood cultures had revealed growth of *Streptococcus constellatus* and *Enterobacter agglomerans*. During that hospitalization the patient underwent aortic valve replacement at the second week of the antibiotic treatment due to persistent heart failure. Postoperatively, the transeophageal echocardiogram revealed severe mitral regurgitation, which was not diagnosed before and remained untreated. He was discharged afebrile,

asymptomatic and hemodynamically stable, on treatment with spironolactone, lisinopril, carvedilol, ivabradine, furosemide and acenocoumarol in view of significant mitral regurgitation.

On physical examination, the patient was pale and afebrile; pulse rate was 71/min and blood pressure 110/60 mm Hg. The auscultation revealed diminished heart sounds, systolic ejection murmur and closing click of the mechanical valve and a great holosystolic murmur that was heard throughout the precordium, best along the left parasternal region. There was also moderate jugular venous distension noted and palpable splenomegaly.

ECG showed sinus rhythm and first degree atrioventricular block, while chest X-ray showed enlarged cardiac silhouette. Laboratory blood tests showed only elevated transaminases, without elevated inflammatory markers. Abdominal ultrasound identified thickening and swelling of the gallbladder wall, amount of fluid in minor pelvis, right pleural effusion, and splenomegaly.

Transthoracic echocardiogram (TTE) rendered a diagnosis of enlargement of the left ventricle. It also showed prosthetic valve in the aortic position with concomitant mild to moderate transvalvular aortic regurgitation (Figure 1). On the parasternal long axis and the apical four chamber view severe mitral regurgitation was visualized with a jet originating from the anterior mitral leaflet that was highly suspicious of a mitral leaflet perforation (Figure 2). In addition, TTE detected a vegetation that was attached to the anterior mitral leaflet. The apical four chamber view showed a large aneurysm at the perimembranous part of ventricular septum (Figure 3a). Furthermore, on the parasternal long axis and the apical five chamber view a pseudoaneurysm of the mitral-aortic intervalvular fibrosa,

with systolic expansion and diastolic collapse, was detected. It is noteworthy that there was downward displacement of all leaflets of the tricuspid valve into the right ventricular cavity (Figure 3a). More specifically, the tricuspid valve was displaced toward the apex by 3.3 cm from the mitral valve insertion (apical four chamber view). However, the tricuspid valve could not be classified as Ebstein's anomaly. Finally, a mild tricuspid valve regurgitation was detected and the transvalvular pressure

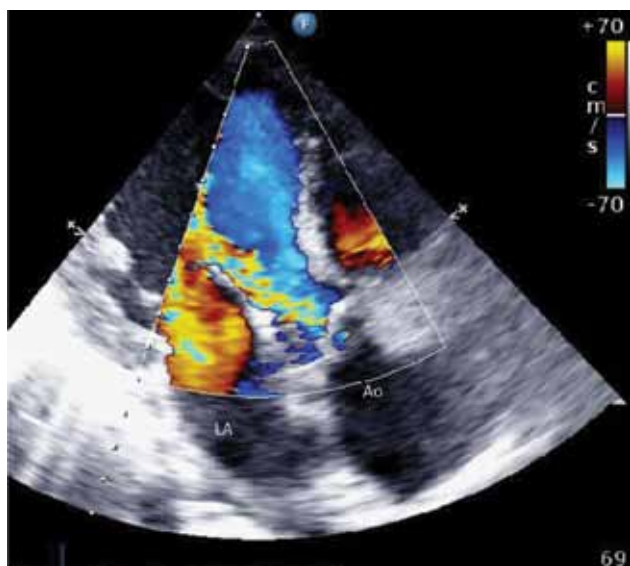


FIGURE 1. Transthoracic echocardiogram illustrates the long axis view of aortic regurgitation through the prosthetic valve.



FIGURE 2. A modified left ventricular outflow view with color-flow Doppler showing the jet (arrow) of mitral regurgitation via the anterior mitral leaflet. Note the prominent pisa.



FIGURE 3A. Transthoracic echocardiogram: four chamber view illustrating the aneurysm of the perimembranous ventricular septum (yellow arrow) and the downward displacement of all leaflets of the tricuspid valve.

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gradient was 35-40 mmHg (modified view). A transesophageal echocardiogram (TEE) confirmed the presence of a large aneurysm of the perimembranous ventricular septum, protruding with each systole, into the right atrium (Figures 3b, 4, 5). The



FIGURE 3B. Transesophageal echocardiogram: four chamber view illustrates the large aneurysm (yellow arrow) at the atrio-ventricular part of the perimembranous inlet ventricular septum and the downward displacement of all leaflets of the tricuspid valve into the right ventricular cavity. Note also the perforation of the anterior mitral leaflet (red arrow).



FIGURE 4. Transesophageal echocardiogram: four chamber view allows the visualization of the aneurysm (yellow arrow) at the atrioventricular part of the perimembranous inlet ventricular septum, the vegetation on the anterior mitral leaflet (red arrow) and the downward displacement of all leaflets of the tricuspid valve into the right ventricular cavity.

color Doppler revealed a color-flow jet from the left ventricle to the right atrium through the aneurysmal perimembranous septum, consistent with a Gerbode defect (Figure 6). On a modified view, two-dimensional and color flow Doppler TEE images also verified a defect on the anterior mitral leaflet with concomitant severe mitral valve regurgitation through this defect (Figures 3b, 6). Moreover, TEE confirmed the pseudoaneurysm of the mitral-aortic intervalvular fibrosa, which protruded into the left atrium (Figure 7).

Since the patient's condition deteriorated progressively



FIGURE 5. Transesophageal echocardiographic image at 94° showing the mitral aortic intervalvular fibrosa (MAIVF) (red arrow). Note also the aneurysm of the perimembranous ventricular septum (yellow arrow) below the anterior mitral leaflet.

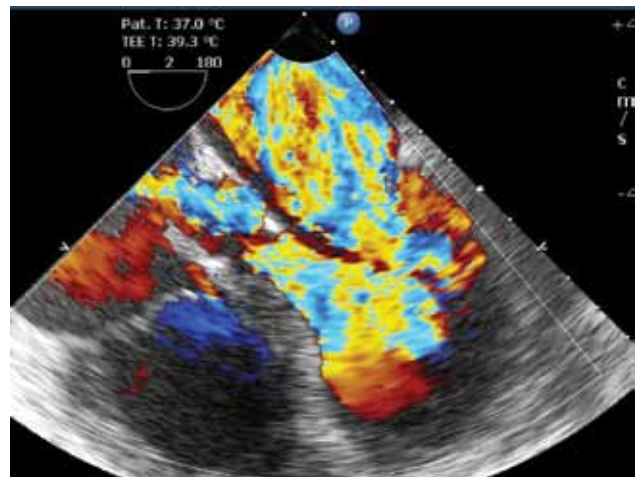


FIGURE 6. Transesophageal echocardiogram: four chamber view at 0° showing the left-to-right shunt through the aneurysm of the perimembranous inlet septum (white arrow) and the severe mitral regurgitation (red arrow).



FIGURE 7. Transesophageal echocardiographic image at 0° demonstrates the presence of a pseudoaneurysm of the mitral-aortic intervalvular fibrosa (arrow) protruding into the left atrium.

despite the optimal medical treatment, mitral valve replacement and repair of the Gerbode defect were performed.

DISCUSSION

A Gerbode-type defect, left ventricle (LV)-to-right atrium (RA) communication, due to perforation of the ventricular septum after aortic valve replacement, has rarely been reported in the literature. Gerbode-type defects are rare intracardiac defects that can be either congenital or acquired.¹ Acquired LV-RA communications arise from ventricular septal perforation as a result of excision of the aortic valve due to aortic valve replacement, but they may also be encountered after infective endocarditis, thoracic trauma, repaired atrioventricular septal defect and as mechanical complications of myocardial infarction.²

Normally, the tricuspid annulus has a more apical location than the mitral annulus, with the septal leaflet of the tricuspid valve resting 5-10 mm more apically than the anterior leaflet of the mitral valve.³ This anatomic arrangement allows the septal leaflet of the tricuspid valve to divide the membranous ventricular septum into two portions: a supraventricular (or atrioventricular) portion and an infravalvular (or interventricular) portion. This anatomic arrangement results in a part of the inlet interventricular septum that faces the left ventricle from one side and the right atrium from the other (Figure 8). Our patient's aneurysm was on that part of the interventricular septum, which was surprisingly longer than normal due to the more inferior attachment of the septal tricuspid leaflet. Furthermore, it was remarkable that not only the septal tricuspid leaflet was displaced towards the apex but all the three

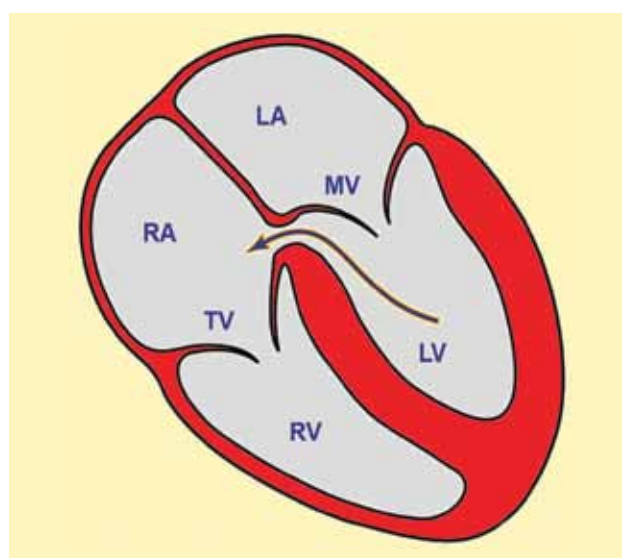


FIGURE 8. Gerbode defect scheme. Abbreviations: LA-left atrium, LV-left ventricle, RA-right atrium, RV-right ventricle, Ao-aorta, AML-anterior mitral leaflet, LAA-left atrial appendage, TV-tricuspid valve, MV-mitral valve, LVOT-left ventricular outflow tract, MAIVF- mitral aortic intervalvular fibrosa.

tricuspid leaflets, a fact that differentiates it from Ebstein's anomaly. Obviously, this aneurysm is congenital in origin and had developed during intrauterine life via a number of mechanisms.³ To our knowledge, an aneurysm of such location and type has never been previously reported. This congenital aneurysm could have been perforated secondarily, probably as a result of the aortic valve excision. Early after aortic valve replacement, a Gerbode defect can be related both to the extension of the valvular debridement and the erosion of the membranous septum by the rigid prosthetic ring.⁴

Mitral valve perforation is a common complication of left-sided native infective endocarditis. Compared to aortic perforation higher prevalence of mitral perforation was observed with mitral valve endocarditis, frequently accompanied by aneurysms of the mitral valve. Mitral valve aneurysms and perforations are usually caused by tissue friability of the anterior mitral leaflet due to inflammatory change in infective endocarditis.³ In our case, perforation might have been the consequence of aortic valve endocarditis, which led to a jet lesion and endocarditis of the anterior mitral valve leaflet, or an iatrogenic complication.

Pseudoaneurysm in the mitral aortic intervalvular fibrosa (MAIVF) is a rare condition that has been reported as a complication of endocarditis and aortic valve replacement.⁵ MAIVF is a relatively avascular structure and, as such, it offers little resistance to infection.⁶ Fistula formation and coronary artery compression were the most common complications. Other reported complications included rupture into the peri-

cardial space and pulmonary artery compression.⁵ Fistulas into the left atrium and aorta were both relatively common findings.

Pseudoaneurysm is an evolution of an evacuated abscess and is identified as a pulsatile, echocardiographic-free sac along the ring. Visualization by TTE of the echo-free space with systolic expansion and diastolic collapse of the pseudoaneurysm is important in making the diagnosis. TEE is a more sensitive technique to evaluate this condition, since the details are likely to be seen well.⁶ Pseudoaneurysm in the MAIVF in our patient is probably a consequence of perivalvular extension of bacterial endocarditis, because it was reported on the echocardiogram before the aortic valve replacement.

We described severe complications of an extensive and complex infective endocarditis. Although *Staphylococcus aureus* is usually the causative organism, due to its pathogenic aggressiveness and capability to cause local complications, *Streptococcus constellatus* and *Enterobacter agglomerans* were the causative bacteria of endocarditis in our patient.⁴ It is worth mentioning that both of these microorganisms rarely cause such an extensive infection.⁷ That is why we hypothesize that in our case, iatrogenic complications combined with the congenital malformations or variations, such as the bicuspid aortic valve and the aneurysm of the perimembranous inlet ventricular septum, might have contributed to the development of this detrimental condition.⁸ We believe that the usefulness of the echocardiography and especially of TEE is of paramount importance in detecting lesions that are congenital, infectious and iatrogenic.

REFERENCES

1. Wasserman SM, Fann JI, Atwood JE, Burdon TA, Fadel BM. Acquired left ventricular-right atrial communication: Gerbode-type defect. *Echocardiography* 2002;19:67-72.
2. Ota T, Yamaguchi R, Tanigawa T, et al. Left ventricular-right atrial communication by perforation of the atrioventricular portion of the membranous septum and severe aortic valve regurgitation caused by infective Endocarditis. *J Echocardiogr* 2011; 9:30-32.
3. Toprak C, Kahveci G, Akpınar S, Tabakci MM, Guler Y. Concomitant Gerbode-like defect and anterior mitral leaflet perforation after aortic valve replacement for endocarditis. *Echocardiography* 2013;30:E231-235.
4. Dores H, Abecasis J, Ribeiros R, Neves JP, Mendes M. Uncommon acquired Gerbode defect following extensive bicuspid aortic valve endocarditis. *Cardiovasc Ultrasound* 2012;10:7.
5. Sudhakar S, Sewani A, Agrawal M, Uretsky BF. Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF): a comprehensive review. *J Am Soc Echocardiogr* 2010;23:1009-1018.
6. Xie M, Li Y, Cheng TO, Wang X, Lu Q, He L, Fu M. Pseudoaneurysm of the mitral-aortic intervalvular fibrosa. *Int J Cardiol* 2013;166:2-7.
7. Bachour K, Zmily H, Kizilbash M, et al. Valvular perforation in left-sided native valve infective endocarditis. *Clin Cardiol* 2009; 32, 12:E55-E62.
8. Yilmaz AT, Ozal E, Arslan M, Tatar H, Ozturk OY. Aneurysm of the membranous septum in adult patients with perimembranous ventricular septal defect. *Eur J Cardiothorac Surg* 1997; 11:307-311.