Case report

Gerbode’s defect resulting from infective endocarditis

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Abstract

We present a report of a Gerbode’s defect (left ventricular–right atrial communication) resulting from bacterial endocarditis in a 63-year-old man. Also presented is a brief overview of the literature and a possible preoperative echocardiographic diagnostic criterion relating to this unusual condition.

Keywords: Gerbode’s defect

1. Introduction

Gerbode’s defect describes a left ventricular–right atrial fistula most often seen as a congenital defect or in association with aortic valve endocarditis. It is often not identified by echocardiography; however, the presence of an aortic root abscess in association with vegetations on the interatrial septum in the right atrium should arouse suspicion of the defect.

2. Case report

A 63-year-old man was admitted with a 2 week history of fever, shortness of breath and ankle swelling. He was febrile with hepatomegaly and bilateral pitting pedal edema but there were no splinter hemorrhages or other stigmata of endocarditis. He had a collapsing pulse and a grade 3/4 diastolic murmur along the left sternal border. An electrocardiogram revealed complete heart block (CHB). His C-reactive protein (CRP), white cell count (WCC) and erythrocyte sedimentation rate (ESR) were elevated. No organism grew in any of three blood cultures. Echocardiography confirmed severe aortic regurgitation and demonstrated echogenic masses consistent with vegetations on the aortic valve and a large aortic root abscess with preserved ventricular function. Another vegetation 1 cm in diameter was seen on the atrioventricular (AV) septum above the septal leaflet of the tricuspid valve but no obvious fistula could be demonstrated between the left ventricle and either the aorta or the right atrium.

A trans-venous pacing wire was inserted and he was commenced on intravenous Augmentin and Gentamycin. One week later he was afebrile with a normal WCC, CRP and ESR. His echocardiogram however, was unchanged. A pre-operative coronary angiogram showed normal coronary arteries. Pulmonary artery and right atrial pressures and right atrial oxygen saturation at the time of cardiac catheterization were not suggestive of a left to right shunt.

At surgery, there was a large abscess cavity related to the membranous interventricular septum, just below the right coronary cusp. The abscess had destroyed the local conduction tissue and muscle, and extended down to the mitral annulus. The aortic valve was replaced with a 22 mm fresh frozen aortic homograft implanted as a free-standing root. In order to exteriorize the cavity, a tongue of tissue was sutured down as far as the mitral annulus. The right atrium was opened and the vegetation visualized in the region of the AV node. It did not involve the tricuspid valve. On removal of the vegetation, however, a defect was found communicating between the left ventricle and the right atrium. This defect was closed by direct suture. Post-operatively the patient remained in CHB and a permanent pacemaker (DDDR mode) was inserted. Twenty-four hours later he required cardio-version for atrial flutter.

Echocardiography before discharge showed a well-
seated normally functioning aortic homograft, a normally functioning mitral valve and good bi-ventricular function. No organism was grown from the excised tissue and antibiotics were discontinued after 2 weeks. The patient remains well 4 years later with a normally functioning aortic valve on echocardiography.

3. Discussion

Gerbode described a syndrome of congenital left ventricular–right atrial shunt with bradycardia and a rise in systemic blood pressure on intraoperative manual closure of the defect [1].

Riemenschneider and Moss [2] described two varieties based on the insertion of the septal leaflet of the tricuspid valve, which divides the membranous septum into interventricular and atrio-ventricular portions. The more common congenital type originates in the interventricular membranous septum and forms a communication between the left ventricle and the right atrium through a defect in the septal leaflet of the tricuspid valve. In the less common form, usually acquired in association with infective endocarditis, the shunt occurs between the left ventricle (LV) and the right atrium (RA) above the septal leaflet of the tricuspid valve, which remains intact [3–5] (Fig. 1). Often there is extension of the infection into the subannular region with involvement of the high membranous septum. This leads to rupture of the portion of the septum that divides the LV from the RA and results in a LV to RA shunt with an intact tricuspid valve. The causative organism is usually a Staphylococcus aureus. Gerbode’s defect has also been reported in association with trauma, following aortic valve replacement, mitral valve replacement, previous repair of an AV septal defect and ischaemic heart disease [8].

The clinical picture varies with mixed symptoms related either to the LV to RA shunt or the underlying etiology. In small defects, as in our case, the shunt is well tolerated and there may be no characteristic symptoms or clinical signs. In larger defects, the physical findings are similar to a ventricular septal defect with a loud harsh holosystolic murmur at the left sternal margin in the fourth or fifth intercostal space. The wide fixed splitting of the second heart sound heard with a large atrial septal defect is absent [1].

Preoperatively, the diagnosis can be established at the time of cardiac catheterization with raised right atrial pressures and a step up in oxygen saturation at right atrial level. Preoperative diagnosis with color Doppler echocardiography has been reported [6,7]. Trans-oesophageal echocardiography (TOE) has been demonstrated to be superior to trans-thoracic echocardiography (TTE) in the detection of vegetations associated with endocarditis and complications such as abscess and fistula formation [8,9]. However, identification of an actual communication is often extremely difficult. In our patient, echocardiography identified a vegetation on the inter-atrial septum above an intact tricuspid valve, the site of a potential Gerbode’s defect. The high index of suspicion prompted its identification at the time of surgery.

A distinctive feature sometimes seen at the time of surgery is the presence of an enlarged RA with systolic expansion [1]. This is because the shunt largely occurs during systole from the high pressure LV into the low pressure RA closed by the tricuspid valve.

Primary closure is possible with small defects while larger defects often require closure with a patch. The proximity of the conducting system often results in preoperative and postoperative conduction abnormalities, which on occasion require pacemaker implantation.

References

