Case report
Intraoperative localisation and management of coronary artery fistula using transesophageal echocardiography

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Abstract
Coronary artery fistula is a rare congenital malformation that can be complicated by intracardiac shunts, endocarditis, myocardial infarction, coronary aneurysm and sudden death. Clinical symptomatology depends upon the underlying anatomy and the size of the fistulous connection between the left or right side of the heart. We report the successful management of a giant right coronary artery with fistulisation into the right atrium. Intraoperative transesophageal echocardiography with colour flow Doppler was used for precise location of the fistulous communication, selective demonstration of vessels feeding the fistula and documentation of abolition of fistulous flow all without the need for cardiopulmonary bypass. Furthermore the effect of shunt occlusion on regional wall motion was documented which facilitated the successful ligation of the fistula. © 1999 Elsevier Science B.V. All rights reserved.

Keywords: Coronary artery fistula; Transesophageal echocardiography; Without cardiopulmonary bypass

1. Case history

A 36-year-old female was referred for assessment following a 6 month history of palpitations associated with shortness of breath on exertion and more recently extreme fatigue and atypical chest pain. Examination demonstrated the presence of a continuous murmur and a hyperdynamic circulation. The chest X-ray and electrocardiogram were unremarkable. Transthoracic two dimensional and colour flow Doppler echocardiography revealed a high velocity shunt into the right atrium which persisted in diastole. There was no evidence of right heart overload. The right sinus of valsalva was enlarged but there was no aortic regurgitation. Magnetic resonance imaging confirmed the presence of a 6 × 4 cm mass indenting the superior aspect of the right atrium. Coronary angiography demonstrated a normal left coronary artery and a giant proximal right coronary artery with a fistulous connection into the right atrium (Fig. 1). The distal bed of the right coronary artery beyond the fistula was visualised and noted to be extremely small.

At operation, the dilated segment of the right coronary artery was identified between the aorta and the superior aspect of the right atrium. The sinus of origin of the right coronary artery as well as the proximal segment of right coronary artery was grossly dilated. This segment was tense with a continuous prominent thrill and transesophageal echocardiography selectively demonstrated the feeding coronary artery, the precise location of the fistulous connection and turbulent flow signals in the right atrium (Fig. 2). Momentary occlusion of the feeding vessel with slings resulted in loss of the continuous thrill and disappearance of the fistulous flow on the echocardiogram. In addition the dilated aneurysmal segment of the feeding vessel became totally collapsed. There was no tricuspid regurgitation or ventricular wall motion abnormalities and furthermore the electrocardiogram remained unchanged. The fistulous vessel was thus doubly ligated in continuity using silk sutures. In our view it was felt that with a single feeding vessel communicating with a low pressure chamber the decompressed aneurysmal segment could be left alone. Moreover this single feeding vessel that controlled the flow into the decompressed aneurysmal segment was well visualised intraoperatively providing further justification that the aneurysmal segment could be left alone. Postoperative course was uneventful and the patient was discharged home on day 5. Six months after the procedure the patient is asymptomatic and has been discharged from surgical follow up.

2. Comments

Intraoperative transesophageal echocardiography has
be shown to have an increasing number of useful applications in recent years. The addition of colour flow and pulsed Doppler to two dimensional echocardiography has greatly facilitated the detection of coronary artery fistulae as the presence of turbulent intracoronary or intracardiac flow can be used to modify standard imaging planes in such a way as to locate precisely their drainage sites [1].

The aim of treatment of patients with coronary artery fistulae is the abolition of the fistulous connection without interference with the native coronary artery [2]. This can be achieved either by transcatheter embolisation or an open surgical approach which may consist of either repair on cardiopulmonary bypass or simple ligation without cardiopulmonary bypass depending on the location of the fistulous connection and its surgical accessibility [2–6]. There still however remains some controversy about the chosen treatment for coronary artery fistulae. Mavroudis et al. in a review of their 28-year experience of 17 patients concluded that early surgical management is safe and effective treatment while transcatheter coil embolisation has been proposed as a reasonable alternative to standard surgical closure in a very small select group of patients with a single, narrow restrictive drainage site into a cardiac chamber or vessel and with the absence of multiple fistulous communication [5].

Transesophageal echocardiography with colour flow Doppler has greatly facilitated the diagnosis and operative management of this condition and it has been suggested that is may be of even sufficient accuracy to preclude coronary angiography [7]. The superior imaging of the proximal coronary arteries form the esophageal window can aid in the location of the origin and course of the abnormal feeding vessel. The effect of temporary ligation of these vessels on fistulous flow can also be demonstrated. In addition, transesophageal echocardiography can be used for the evaluation of the adequacy of fistula ligation. Rittenhouse and colleagues observed that two of eight patients had small residual fistulous flow. Although this may be insignificant in the early post operative period it may have the potential to lead to a larger shunt over time [8]. Continuous transeso-

Fig. 1. A schematic of the coronary angiogram demonstrating a giant right coronary artery with a fistulous connection into the right atrium.

Fig. 2. Color flow demonstrating the fistulous flow between the aorta and the right atrium. (RA, right atrium; LA, left atrium; AO, aorta; CAF, coronary artery fistula).
phageal echocardiography provide the opportunity to observe changes in regional wall motion of the ventricle supplied by the coronary artery and to determine the need for distal coronary artery bypass grafting [2,9].

This case report has shown that intraoperative transesophageal echocardiography can provide precise information during the intraoperative management of coronary artery fistulae. The clear visualisation of the site and extent of the fistulous communication as well as the effect of shunt occlusion on regional wall motion permitted the successful ligation of the abnormal coronary fistula.

References