Coronary artery fistula: a rare cause of a left to right shunt

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CASE REPORT

A 48-year-old woman presented with increasing breathlessness on exertion. A continuous murmur had been noted 6 years earlier and attributed to a patent ductus arteriosus. At referral, examination revealed a systolic murmur (2/6) continuing across the second sound into late diastole (1/4), audible at the left sternal edge. Her electrocardiogram showed sinus rhythm, a normal axis and non-specific ST changes in the inferior leads.

A transthoracic echocardiogram demonstrated impaired left ventricular function, most marked in the anterior wall, and a well circumscribed area of turbulent flow in the region of the interatrial septum but no shunt at this level (using Doppler and colour coding). These findings were suggestive of a vessel running through the interatrial septum which was subsequently confirmed using transoesophageal echocardiography (*Figures 1* and *2*). Running a tortuous course from its origin at the left main coronary artery, through the interatrial septum to drain into the right atrium, the serpiginous appearance of the vessel was typical of a coronary artery fistula. The left ventricular hypokinesia and fistula were confirmed at cardiac catheterization but no additional atherosclerotic disease was demonstrated. A thallium perfusion scan, including adenosine vasodilation, failed to demonstrate any perfusion defect. Nonetheless the left coronary artery to right atrial fistula and left ventricular impairment suggested the patient's symptoms were a function of inadequate myocardial perfusion resulting from coronary artery steal and the patient was referred for surgical ligation. At surgery a 1 cm fistulous dilatation of the left main stem of the left coronary artery was found: the fistula then coursed over the roof of the left atrium and down through the interatrial septum to drain into the right atrium. The fistula was ligated as it crossed over the roof of the left atrium. The patient made an uneventful postoperative recovery. When assessed several months later her symptoms had fully resolved and repeat echocardiography confirmed normal left ventricular function.

We report a case of a middle-aged woman, assumed to have a patent ductus arteriosus on the basis of clinical findings. Subsequent investigations in this patient demonstrated a left coronary artery to right atrial fistula.

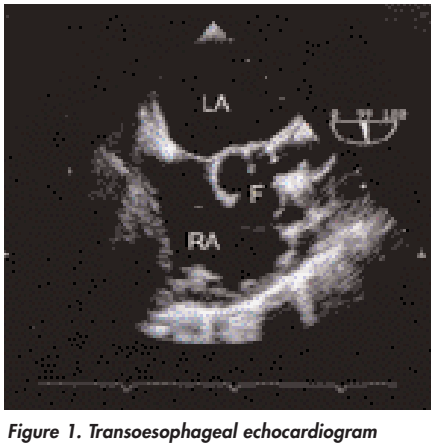

Figure 1. Transoesophageal echocardiogram image in the vertical plane showing the left atrium (LA) and right atrium (RA) with a tortuous left coronary artery fistula (F) draining into the RA.

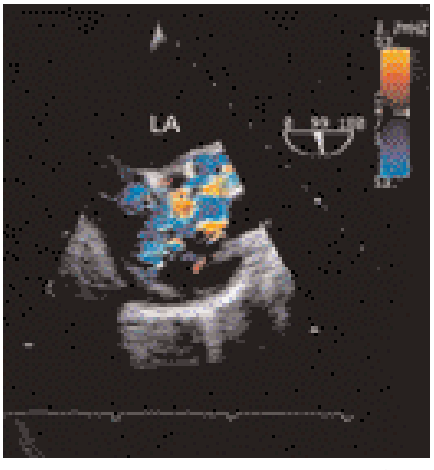

Figure 2. Identical plane as* Figure 1 *showing flow from the fistula into the right atrium.

DISCUSSION

We report the case of a left coronary artery to right atrial fistula which is a less common cause of a continuous murmur and left to right shunting, than the patent ductus arteriosus for which it was mistaken. Although there can be some overlap in clinical signs and symptomatology, as this case illustrates, it is relatively unusual for a patient with a patent ductus arteriosus to remain asymptomatic and undiagnosed until middle age, whereas patients with coronary artery fistulae typically present at this age (Liberthson et al, 1979). Their usual symptoms are of angina and or exertional dyspnoea as a result of coronary steal.

The hypoperfusion of some or all of the left ventricle is exacerbated by exercise and the coexistence of coronary atherosclerosis which is common in the same age group. The murmur associated with a coronary artery fistula is frequently continuous, as in this case, although systolic and diastolic murmurs have both been reported in isolation.

The first description of a coronary artery fistula was made in 1865 by Krause. The majority are congenital, arising from incomplete obliteration of primitive myocardial trabecular sinusoids (Lowson et al, 1996). More rarely, they are iatrogenic following

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interventions such as percutaneous transluminal coronary angioplasty or endomyocardial biopsy. Coronary artery fistulae most commonly arise from the right rather than the left coronary artery (Levin et al, 1978). Drainage is usually into the right atrium or ventricle, but drainage into the pulmonary artery, coronary sinus, superior vena cava and left heart have all been reported (Lin et al, 1995).

The diagnosis may, as here, be suspected on the basis of a transthoracic echocardiogram but the usually highly tortuous vessel is best demonstrated using transoesophageal echocardiography (*Figures 1* and *2*). This technique allows the origin, course and drainage of the vessel to be defined, along with other cardiac malformations which are reported in 20% of cases (Kugelmass et al, 1992). Transoesophageal echocardiography is comparable or superior to angiography in the diagnosis of coronary artery fistulae (Samdarshi et al, 1991; Lin et al, 1995), although coronary angiography is usually required to diagnose coexistent coronary disease.

Untreated coronary artery fistulae may result in left ventricular dysfunction, congestive cardiac failure with left to right shunts accounting for up to 50% of the cardiac output, thromboembolism, endocarditis and sudden cardiac death (Carrel et al, 1996; Levin et al, 1978). Standard treatment of symptomatic patients is surgical, with the aim of eliminating the fistulous vessel while maintaining normal coronary flow, although fistula obliteration has been achieved using either umbrella devices or embolization (Carrel et al, 1996).

Despite the inability of the thallium to demonstrate any perfusion defect, the most likely cause for the patient's symptoms was coronary artery steal. The dramatic improvement of her symptoms and restoration of normal left ventricular function following ligation of the fistula support this diagnosis. **HM**

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In the Public's View...

Politicians never learn: don't believe the hype?

Politicians never learn. They are so afraid of uncertainty. It is a mantra of the Ministry of Health that mammograms prevent breast cancer. Every other day or week it is breast cancer awareness time — never mind that we are already more aware of breast cancer, with its pink ribbons, than any other adult cancer. (Aggrieved men are trying to do the same for prostate cancer, but who wants to be aware of prostates?) And then the *Lancet* publishes a paper claiming that mammograms might detect cancer, but they don't save lives.

There is nothing wrong in being wrong. People were wrong when they thought the sun went round the Earth. They weren't fools. They had no better evidence than the sun rising, crossing the sky, going below the horizon, and rising again the next morning where it had risen the day before. But looking more carefully, there were anomalies.

At first, anomalies to cherished theory are explained away with modifications to the theory. As the anomalies accrue, the theory begins to creak at the edges. In the days of debating whether the sun did indeed go round the Earth, a few severed heads were used to prevent the creaking. Because truth will out, there was eventually no alternative to accepting a new theory that the Earth goes round the sun, and the old theory and its anomalies were consigned to the waste bin.

It's a perfectly reasonable theory that mammograms stop women dying from breast cancer. What the *Lancet* paper has done is discover anomalies. The anomalies do not yet mean that the theory is wrong; they mean the theory, and the anomalies, must be looked at carefully. It helps nobody that the argument about mammograms immediately polarized. Peter Götzsche, an author of the paper, said the only properly compared trials showed no advantage and screening is not justifiable. Yvette Cooper, Minister of Health, said thousands of women's lives are saved by screening. One thing is certain: they cannot both be right.

Except, of course, that they can. Perhaps some types of breast cancer, in some women, are worth looking for, and others are not. The theory is creaking, but the creak may be outside and need not concern us. What the politicians must not do is rush unthinkingly to bolster their cherished theories, or they will appear even more foolish and untrustworthy if eventually mammography has to be abandoned. They must also ensure that women are not frightened into having mammography, but are encouraged to undergo the procedure in the full knowledge of what it might or might not do.

At one time, mobile X-ray vans toured the streets, no longer needed for TB but hoping to detect lung cancer. When it was realized that X-ray screening was pointless, the vans were withdrawn. If mammograms are eventually withdrawn, it need not reflect badly on the Ministry of Health — unless they prefer political blinkeredness to scientific enquiry. They could start their rehabilitation by taking notice of Karol Sikora's letter in the new year issue of the *BMJ* and humbly admitting that it helps no one — especially in the middle of a flu epidemic — to demand that all patients suspected of having any form of cancer be seen within 2 weeks, a policy for which there is no rational support at all. **HM**

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