Multiple Coronary Arterioventricular Fistulae

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SUMMARY A 62-year-old man who died suddenly was found at autopsy to have multiple coronary arteriosystemic fistulae involving all three major coronary arteries and both ventricles. This is the second such case reported and the first with biventricular involvement and autopsy findings.

A congenital bicuspid valve was also present. Numerous small blood vessels opened onto the endocardial surface of both ventricles and there were microscopic foci of persistent embryonic spongy myocardium.

A CORONARY ARTERY communicating with a cardiac chamber or the pulmonary trunk may cause a continuous murmur. Most patients with this condition are symptomless, but the fistula may lead to congestive heart failure, infective endocarditis or myocardial ischemia. The condition is usually congenital, but has been reported after myocardial infarction.1 The congenital form is not rare and in most instances a single fistula is present.

However, congenital coronary fistulae communicating with the left ventricle are very rare, only nine cases have been reported in the world literature. Multiple coronary arteriosystemic fistulae involving all three major coronary arteries have been reported only once.2 This report describes the second such case and for the first time describes autopsy details.

Our patient is different from that of Reddy et al.2 in that he had fistulae draining into both ventricles and he had a congenital bicuspid valve.

Case Report

A 62-year-old male manual laborer collapsed and died at work. He had no history of heart disease or chest pain. No details of past medical examinations were available. At necropsy the pertinent findings were confined to the heart, which weighed 340 g. The epicardial coronary arteries and their branches showed an extreme degree of dilatation and tortuosity (figs. 1 and 2). Some distal coronary arteries attained a diameter of up to 1 cm. The elongated vessels had formed multiple mobile loops on the surface of the heart. The luminal dilatation involved all the epicardial coronary arteries in a diffuse fashion. No atherosclerotic plaques were present. The coronary arteries arose from separate aortic ostia. A large conus artery originated next to the right coronary artery. The coronary veins appeared normal. A congenital bicuspid aortic valve was present.

The intra-myocardial arteries in all portions of the ventricles were enlarged and many were visible to the naked eye. On multiple sections several could be traced right down to the endocardial surface (fig. 2). Histologically, numerous large blood vessels were seen within the myocardium of both ventricles (figs. 3 and 4). Most of these vessels had a stout muscular wall with a distinct, sometimes reduplicated, internal elastic lamina. Some vessels showed intimal sclerosis. Dilated sinusoidal vessels lined by flattened endothelium, with an appearance similar to the embryonic trabecular pattern called spongy myocardium,9 were also present (fig. 5). Large, thick-walled muscular arteries were present close to the endocardium and were seen to communicate with the more delicate sinusoidal-type vessels. A few areas of confluent sub-endocardial fibrosis were present (fig. 6). The left ventricle had a few myofibers showing myofibrillar degeneration. Multiple sections of the conduction system revealed no abnormal feature.

Discussion

Several authors have comprehensively reviewed the literature of anomalous communications of coronary arteries.4–8 Fistulae involve the right coronary artery more frequently than the left. McNamara and Gross7 reviewed 172 cases of coronary fistulae, and four of those cases involved both coronary arteries. Six others involved a single coronary artery. Over 90% of coronary fistulae empty into the right ventricle, right
atrium or pulmonary artery. Only 23 fistulae were found to drain into the left side of the heart (17 to the left atrium, and six into the left ventricle). A recent study\(^9\) indicated that patients with this lesion develop symptoms at a young age only in the presence of a large shunt. All the other patients are asymptomatic until the third decade of life, when they may slowly develop congestive heart failure.

Myocardial ischemia and infarction are rare complications, usually seen in the elderly, and it has been suggested\(^8\) that the uninvolved coronary artery contributes to the adequacy of myocardial perfusion.

Although he was elderly, the subendocardial fibrosis in our patient may be due to the absence of such an uninvolved coronary artery. If our patient had developed symptoms from the fistula during life, surgery would not have been feasible due to the numerous fistulous connections present. We do not know whether the fistulae played any role in the patient's sudden death.

Vlodaver et al.\(^8\) found reports of five cases of communication of a coronary artery with the left ventricle and added one of their own, making a total of six cases. In five out of the six cases the right coronary artery was involved, and the sixth case involved the anterior descending artery. Okuda et al.\(^10\) described a right coronary artery-to-left ventricle fistula, which they report as the sixth case in the literature. I have been able to find nine reported cases of congenital coronary fistulae involving the left ventricle (table 1). Our patient is thus the 10th reported case of coronary fistulae involving the left ventricle.
fistula to the left ventricle and the first with multiple fistulae of both coronary arteries to both ventricles. We do not know of any other reported instance of the association of coronary fistula with a congenital bicuspid aortic valve.

References

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Abbreviations: RCA = right coronary artery; LCA = left coronary artery; LAD = left anterior descending coronary artery; CFX = circumflex artery; LV = left ventricle; RV = right ventricle.

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