

A case of Marfan's syndrome with absent right coronary artery complicated by aortic dissection and right ventricular infarction

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Leitch, A. G. and Caves, P. K. (1975). Thorax, 30, 352-354. A case of Marfan's syndrome with absent right coronary artery complicated by aortic dissection and right ventricular infarction. The first patient to be reported with Marfan's syndrome and an absent right coronary artery is described. Dissection of the ascending aorta was associated with right ventricular infarction which was probably due to the coronary artery anomaly and caused his death.

CASE REPORT

A 19-year-old boy presented with sudden severe central chest pain. Examination showed the features of Marfan's syndrome. He was 1.9 metres (76 inches) tall with frontal bossing, pigeon chest, high arched palate, and arachnodactyly. He was myopic but there was no clinical evidence of ectopia lentis. He had bilateral pes cavus. The blood pressure was 120/70 mmHg, all peripheral pulses were present and equal, and there was no evidence of cardiac failure. Auscultation revealed a soft apical systolic ejection murmur. The chest radiograph was normal. Serial cardiac enzymes showed a moderate elevation in the first three days. The admission electrocardiogram showed second-degree atrioventricular block with ST elevation in leads II, III, and aVF, T-wave inversion in leads V2-3, and ST depression in leads V4-6. Serial ECGs indicated evolving inferior myocardial infarction. A pericardial friction rub developed on the fourth day and persisted. On the sixth day left and right ventricular failure developed with a small right pleural effusion. The cardiac failure did not respond to conventional therapy, and a further deterioration in his condition was noted on the eleventh day associated with a fall in haemoglobin concentration and the development of an aortic early diastolic murmur. Emergency operation was recommended, the diagnosis being dissection of the ascending aorta with myocardial infarction, aortic incompetence, and haemopericardium.

OPERATIVE FINDINGS The pericardial cavity contained 500 ml of blood and blood clot. The right atrium was dilated and tense, and its appendage was occluded by clot. The right ventricle was dilated and tense and did not contract well. The left atrium and ventricle appeared normal. There was an almost circumferential dissection of the ascending aorta which, on cardiopulmonary bypass, was found to extend from just proximal to the innominate artery down into the right and non-coronary sinuses but not into the left coronary sinus. There was no right coronary artery ostium but the left coronary artery ostium was large and normal. The aortic annulus was dilated, and the thin, fine valve cusps were incapable of occluding it.

The aortic valve was replaced with a no. 1 Starr-Edwards composite seat prosthesis. The ascending aorta was resected, leaving only a cuff of aorta around the left coronary ostium. Aortic continuity was restored with a 30 mm crimped woven Dacron tube graft.

After defibrillation the left ventricle contracted vigorously. The right ventricle distended and did not appear to eject blood. Despite maximum use of inotropic agents all attempts to wean the patient from bypass failed because of gross right ventricular failure, and the patient died in the theatre.

PATHOLOGICAL FINDINGS The heart weighed 600 g. Both the dilated right atrium and ventricle

contained organized antemortem thrombus adherent to their walls. There was extensive old and recent infarction of the entire free wall of the right ventricle. The infarcted area also involved the posterior part of the interventricular septum and extended on to the diaphragmatic surface of the left ventricle. A separate strip of more recent infarction was present in the mid-portion of the interventricular septum. The left atrium was normal, but the left ventricle was hypertrophied.

The right coronary artery was absent. The left coronary artery divided into an anterior descending branch, a large vertical branch to the left ventricle, and a circumflex branch. Terminal branches of the circumflex artery supplied the posterior aspect of the heart and the right ventricle. Plaques of atheroma were present in the left main coronary artery at the point where the three major branches arose and in their proximal portions. There was no luminal narrowing, recent thrombosis nor embolus.

The lungs were congested and oedematous.

MICROSCOPY The resected aorta showed areas of cystic medial degeneration and recent haemorrhage in the adventitia with marked absence of elastic tissue on special staining.

Sections of coronary arteries from both ventricles showed recent mixed thrombus occluding the lumen. There was marked thickening of the intima with severe atherosclerotic changes. The media was thinned with patchy deficiency of elastic tissue but no cystic degeneration.

Sections of the right ventricle showed extensive replacement of myocardium by active fibrous tissue, consistent with infarction some weeks before death. There was also recent infarction, some areas showing necrotic muscle without an inflammatory reaction, and others an early acute inflammatory response. This was considered to be consistent with two terminal episodes of acute infarction, one taking place six to 12 hours and the other less than six hours before death. The left ventricle also showed focal areas of active fibrosis similar to those seen in the right ventricle and consistent with infarction some weeks before death. There was hypertrophy of both right and left ventricle myocardium.

DISCUSSION

There are no previous reports of a patient with Marfan's syndrome and a single coronary artery. A case of dissection of the aorta in a 28-year-old man with an absent right coronary artery, high

arched palate, and loss of aortic elastic tissue has been described (Alderson, 1972), but the major stigmata of Marfan's syndrome were absent.

A single coronary artery is a rare finding (Allen and Snider, 1966). Hillestad and Eie (1971) found only 85 cases in the literature and themselves reported three cases in 1000 coronary angiograms. The single left coronary artery in the present case followed the normal course for that artery, the right ventricle and atrium deriving their blood supply from the circumflex branch, a common arrangement in such cases (Smith, 1950; Ogden and Goodyer, 1970).

Isolated right ventricular infarction is also rare (Laurie and Woods, 1963; Hudson, 1965) and has been reported in association with single left (Swann and Fitzpatrick, 1954) and single right (Roberts and Loube, 1947) coronary arteries. It is unclear why the condition is so rare since the right ventricle is most commonly supplied by the right coronary artery (Gross, 1921) and this artery is frequently narrowed or occluded (Schlesinger and Zoll, 1941). Blumgart, Schlesinger, and Davis (1940) suggested that because of its thin wall the right ventricle might derive oxygen from the blood within its chamber, and Wood (1962) was of the opinion that the good collateral circulation of the right ventricle was an adequate explanation. Wade (1959) has drawn attention to the possible importance of right ventricular hypertrophy as a factor in the aetiology of right ventricular infarction, undue demands in this circumstance being made upon the collateral circulation.

The aetiology of the right ventricular infarction in the present case is not clear. Right ventricular hypertrophy was present and, as indicated above (Wade, 1959), may have been a predisposing factor. The infarct was within the distribution of the circumflex branch of the left coronary artery, and yet there was no evidence of embolism, occlusive thrombosis or stenosis of this artery although marked atheroma was present. Diffuse coronary arterial disease of a type known to be found in Marfan's syndrome (James, Frame, and Schatz, 1964) was not present, and there was no evidence of cystic medial necrosis or dissection of the coronary artery (Boschetti and Levine, 1958; McKeown, 1960). At operation and post mortem there was no conclusive evidence of compression of the single coronary artery by the dissected aorta or by any associated haematoma (Hirst, Johns, and Kime, 1958), although there seems little doubt that had flow through this atherosclerotic coronary artery been reduced, the right ventricle would have been predisposed to infarction by virtue of

the absent right coronary artery and consequent lack of collataeral circulation.

In Marfan's syndrome dissection of the thoracic aorta is a fairly common cardiovascular complication (Hirst and Gore, 1973), and the prognosis following acute dissection without operation is extremely poor (Hirst *et al.*, 1958; Anagnostopoulos, Prabhakar, and Kittle, 1972). Although this complication occurred in our patient, he survived long enough for an emergency operation to be completed. Right ventricular infarction, related to the absence of a right coronary artery, was the cause of death.

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