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Case Report

Aberrant Coronary Artery in Association With a Quadricuspid Pulmonary Valve

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Summary. A case of quadricuspid pulmonary valve with an accessory coronary artery in an 82 year old woman is reported. This represents persistance of an early embryonic stage of development of the coronary circulation.

Introduction

Abnormalities of the similunar valves are common malformations. They are of varying degrees of severity and have varying physiological effects (see Table 1). The coronary circulation is also prone to developmental anomalies. These are conventionally divided into major and minor types depending upon their effects on the myocardial blood supply (see Table 2).

Abnormalities of the semilunar valves occurring in association with congenital anomalies of the coronary circulation are excessively rare. In an extensive review of the literature we were unable to find another case report.

Table 1. Abnormalities of the semilunar valves

Valve	Abnormality	Incidence	Effect
Aortic	Bicuspid	2% (Roberts, 1970)	Nil or sclerosis
	Quadricuspid	0.008% (Peretz et al., 1969)	Nil or regurg.
Pulmonary	Bicuspid	0.002% (Koletsky, 1941)	Nil
	Quadricuspid	0.1% (Enoch, 1968)	Nil or regurg.

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Table 2. Abnormalities of the coronary circulation

Abnormality	Incidence	Effect
Minor abnormalities		
Dominant left coronary artery	10% (Lurie, 1977)	Nil
Separate origin of the conus branch of the right coronary artery	50% (Lurie, 1977)	Nil
Single coronary artery	Unknown (Pomerance and Davies, 1975)	Possibly increased susceptibility to atheromatous narrowing
Major abnormalities		
Left coronary artery arising from the pulmonary trunk	0.46% of C.H.D. (Gasul et al., 1961)	1. Infantile ischaemia 2. Adult-cardiomyopathy Sudden death Mitral regurgitation
Right coronary artery arising from the pulmonary trunk	Unknown (Lurie, 1977)	Nil
3rd coronary artery arising from the pulmonary trunk	Unknown (Lurie, 1977)	Nil
Stenosis of the coronary ostium	Unknown (Mullins et al., 1972)	Ischaemic heart disease

Clinical History

An 84 year old lady presented with a long history of chronic bronchitis, congestive cardiac failure and with a large, irreducible, ventral hernia. On her last admission to hospital she developed an acute chest infection, her heart failure was uncontrolled and despite adequate treatment she died.

There was no record of any heart murmurs.

Autopsy Findings

Externally she was noted to be obese, to have marked ankle and sacral pad oedema, a large irreducible ventral hernia and numerous pigmented macules over her trunk and the upper parts of her thighs.

Internally, the main findings of interest were the cardiac abnormalities (described below). The bronchi were dilated with thickened walls and showed an acutely inflamed mucosa. There was pulmonary oedema. The liver was enlarged and had a 'nutmeg' appearance, the spleen was slightly enlarged. There were four meningiomata within the skull.

Approximately 1 m from the duodeno-jejunal flexure there was a 7 cm diameter smooth mass, arising from the mesenteric border of the bowel. Histological examination showed this to be a leiomyosarcoma.

There were isolated plaques of atheroma in the ascending aorta, the arch of the aorta and in the carotid vessels. There was complicated atheroma in the descending thoracic aorta and in the abdominal aorta.

Cardiac Findings

The external form of the heart was normal and its weight was 460 g. The circumference of the tricuspid valve was 12 cm, that of the pulmonary valve 8.3 cm, that of the mitral valve 11 cm and that of the aortic valve 7.75 cm. The wall thickness of the right ventricular outflow tract was 0.4 cm and that of the left ventricular inflow tract 1.8 cm.

There was widespread, but non-occlusive, atheroma in all main branches of the normal coronary arteries (the lumen of the left circumflex coronary artery was reduced to 0.2 cm) but there was no ulceration and no thrombosis. The right coronary artery was dominant. There was a 2 cm diameter area of myocardial necrosis in the anterior wall of the heart involving the interventricular septum, it did not extend through the full thickness of the heart wall.

The aortic valve cusps were normal. The two main coronary arteries were

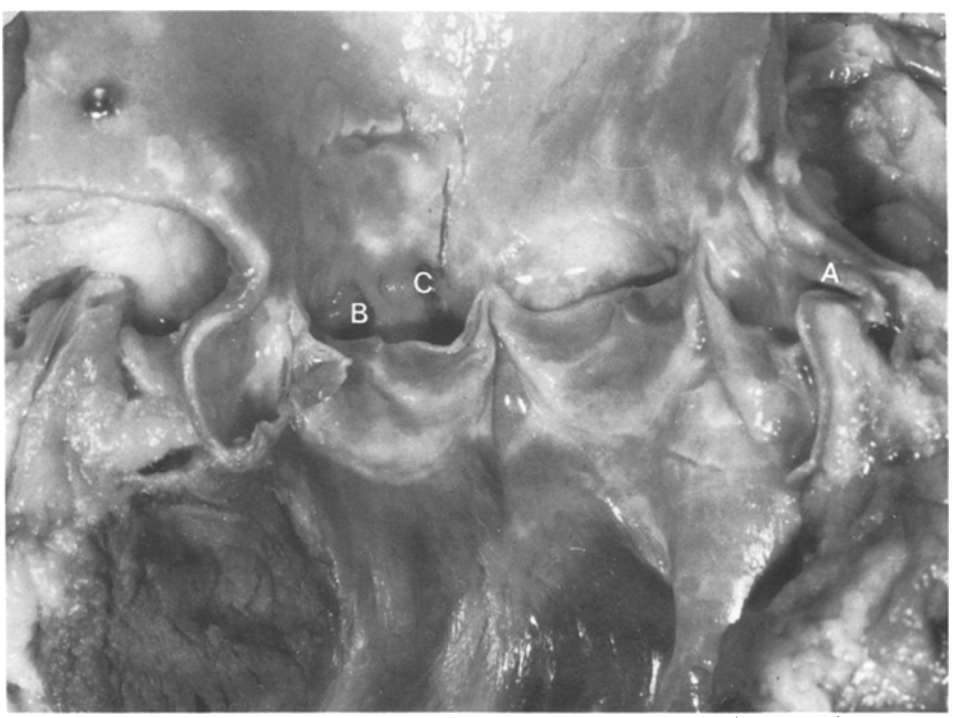

Fig. 1. Aortic valve showing three cusps and the origins of the coronary arteries, including the conus branch of the right coronary artery. *A* Left coronary artery; *B* Right coronary artery (main branch); *C* Conus branch of right coronary artery

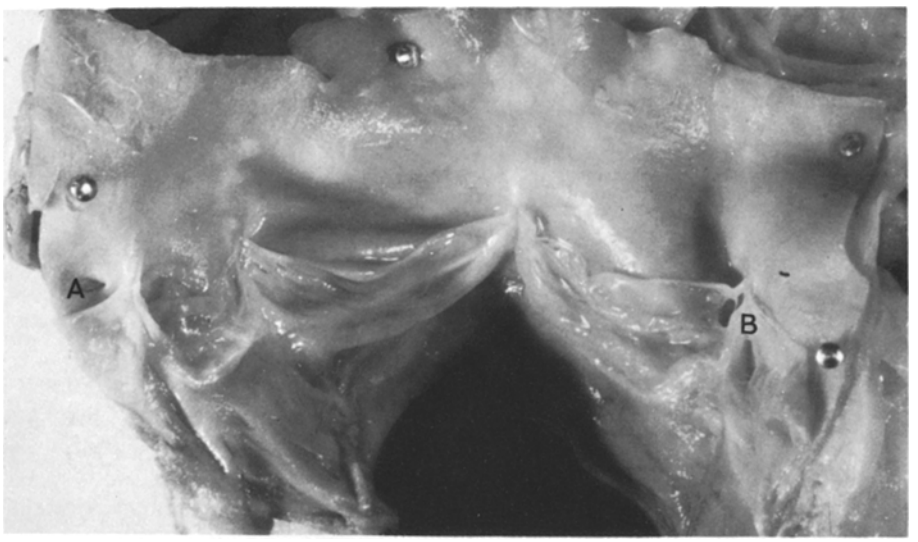

Fig. 2. Pulmonary valve showing four cusps. One cusp has been cut in two. *A* Origin of aberrant coronary artery; *B* Fenestrations in two of the valve cusps

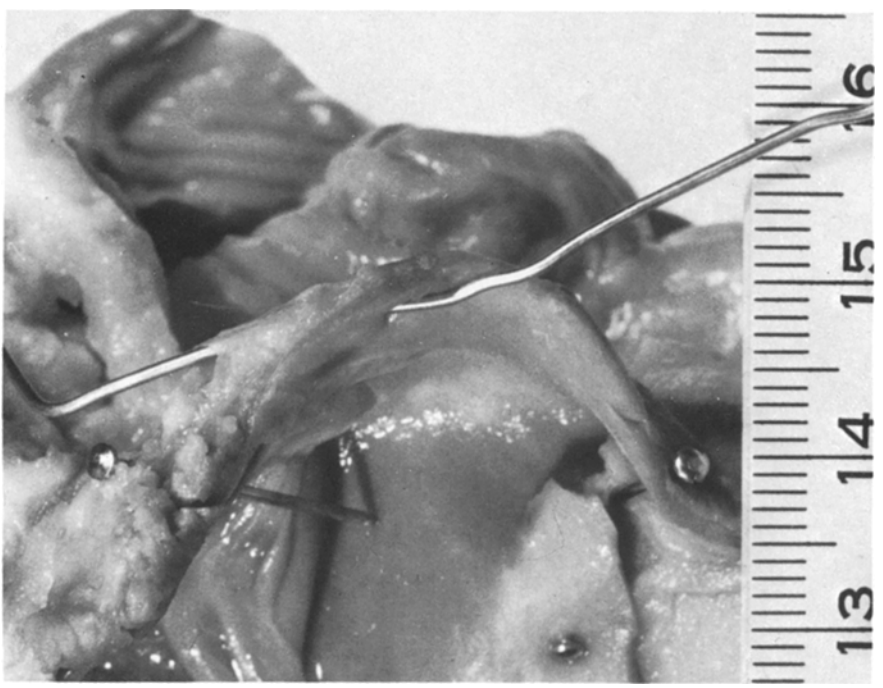

Fig. 3. Pulmonary valve with a probe in the aberrant coronary artery. This picture demonstrates part of the course of the aberrant artery

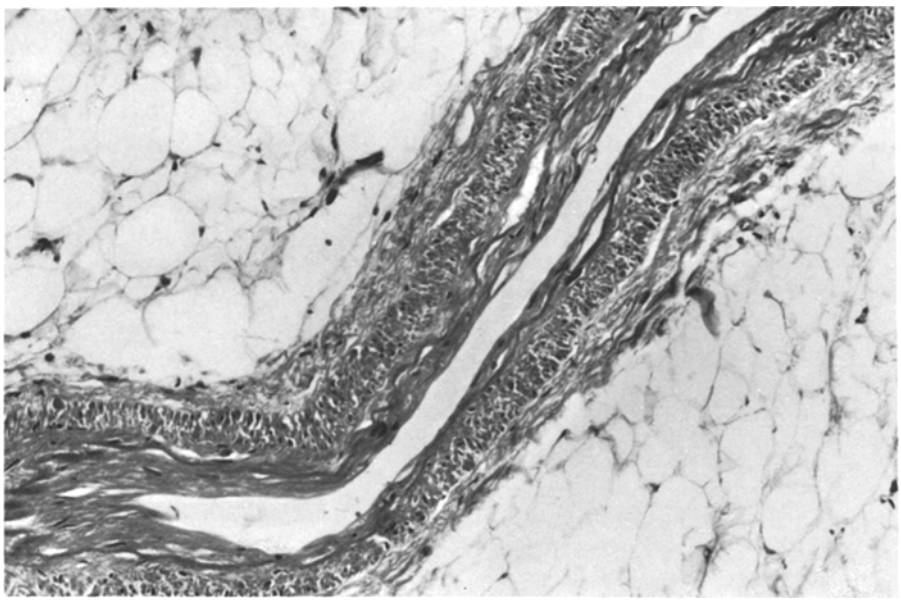

Fig. 4. Longitudinal section of the aberrant coronary artery. This section demonstrates that the artery is thin walled. H & E, ×120

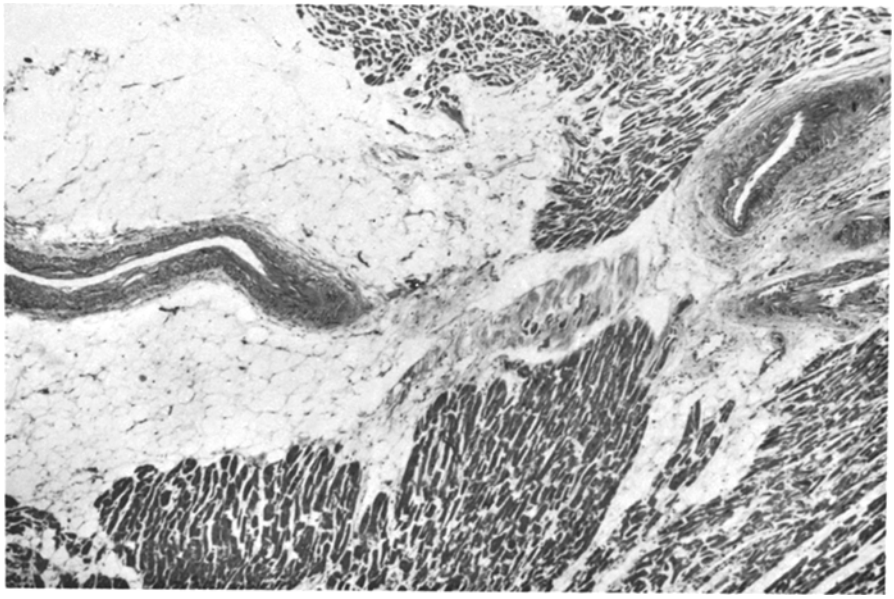

Fig. 5. Section showing the termination of the aberrant coronary artery in the wall of the right ventricle. H & E, ×30

normal, but there was a separate origin of the conus branch of the right coronary artery (see Fig. 1).

The pulmonary valve had four cusps. There were two larger and two smaller cusps and two of the cusps were fenestrated (see Fig. 2). A third coronary artery arose from above one of the smaller cusps, it had a short course and ended in the wall of the right ventricle (see Figs. 2 and 3).

After photography, the vessel was removed and examined histologically. It proved to be a thin walled artery and sections demonstrated its termination in the wall of the right ventricle (see Figs. 4 and 5).

Discussion

Two large and two small valve cusps is one of the rarer patterns of quadricuspid pulmonary valve reported by Kissin (1936) who also described a case in which a supernumerary pulmonary valve cusp was associated with symptoms. Although our case had congestive cardiac failure, there were no documented clinical signs of pulmonary valve disease. This case, therefore, resembles the five cases described by Enoch (1968) which were asymptomatic.

The histology of the aberrant coronary artery is interesting, the thin walled structure presumably reflecting the lower pressures in the pulmonary circulation (Berry, 1978).

This case report illustrates persistance of a stage in the normal development of the coronary arteries. In the seven week embryo buds appear at the base of the pulmonary trunk and the aorta. The buds appearing at the base of the aorta will form the normal coronary arteries, those appearing at the base of the pulmonary trunk usually involute (Hackensellner, 1956). Aberrant coronary arteries arising from the pulmonary trunk probably represent persistence and development of these pulmonary buds.

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