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SHORT CASES IN CARDIOLOGY

Single coronary artery with infundibular pulmonary stenosis

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Single coronary artery is a rare congenital anomaly.¹ Infundibular pulmonary stenosis (IPS) without ventricular septal defect or tetralogy of Fallot (isolated IPS) is also very rare.² We report a case of single coronary artery with isolated IPS; to our knowledge, this is the first report of such a case.

Case report

A 57 year old asymptomatic man was referred to our hospital because of chest radiography and ECG abnormalities. Physical examination revealed a grade 4 harsh systolic ejection murmur heard best in the fourth intercostal space at the left sternal border. Chest radiography showed right atrial and ventricular enlargement with a cardiothoracic ratio of 0.55. Pulmonary vascularity was normal. ECG showed right ventricular hypertrophy.

Echocardiography revealed a dilated right ventricular body and a protruding lesion at the proximal portion of the infundibulum. Cardiac catheterisation disclosed a peak systolic pressure gradient of 68 mm Hg between the right ventricular body and the infundibulum. The right atrial and the pulmonary arterial pressures were normal. Oxygen sampling revealed no evidence of shunting. Right ventricular angiography revealed a prominent muscle band at the junction of the right ventricular body and the infundibulum (fig 1). Both the pulmonic valve and the ventricular septum were intact. Coronary artery angiography revealed a solitary left sided coronary ostium. An anomalous right coronary artery ran from the left anterior descending artery anteriorly to the root of the main pulmonary artery without stenosis (fig 2). It arose from the middle portion very close to the first septal perforating branch of the left anterior descending artery. These findings are compatible with isolated IPS associated with single coronary artery.

The patient underwent surgical resection of the anomalous muscle band and no vestige of ventricular septal defect was found at operation.

Discussion

In most cases of single left coronary artery, the right coronary artery arises from the main trunk, the proximal portion of the left anterior descending artery, or the left circumflex artery.³ Few reports have described cases of anomalous right coronary artery arising from the middle portion of the left anterior descending artery without other congenital anomalies.^{1,3} Single coronary artery is usually combined with various cardiac anomalies such as improper division of truncus arteriosus, transposition of great vessels, and tetralogy of Fallot. The present case was associated with isolated IPS, itself a rare anomaly. Most patients with IPS are diagnosed and surgically

Figure 1 Right anterior oblique view of right ventricular angiography revealing infundibular pulmonary stenosis (arrows). Both the pulmonic valve and the ventricular septum were intact.

Figure 2 Right anterior oblique (upper panel) and left anterior oblique (lower panel) views of coronary artery angiography revealing an anomalous right coronary artery (arrows), which arose from the left anterior descending artery.

treated in early infancy because other combined anomalies, such as ventricular septal defect or tetralogy of Fallot, produce a shunt. In this case, the patient had been asymptomatic and was first diagnosed in his late 50s because no shunt was present and the stenosis was mild. No case of single coronary artery with isolated IPS has been reported.

From an embryological standpoint, a failure in sprouting of the primitive aortic sinus wall during the fifth to sixth intrauterine week results in single coronary artery.⁴ Insufficient absorption of the mesenchymal tissue in the right ventricle leads to anomalous muscle partition between the right ventricular body and the infundibulum. This absorption occurs at the same embryonic period as the sprouting of coronary arteries from the aortic sinus.⁵ This close timing between the development of coronary arteries and the right ventricle may explain the rare combination of single coronary artery and IPS in the present case.

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