Anomalous Origin of Left Coronary Artery with Left Anterior Descending Artery Connected to Left Pulmonary Artery from Single Right Coronary Artery

S Williams-Phillips

ABSTRACT

Single right coronary artery, so defined because all three coronary arteries arise from the single right aortic sinus of Valsalva, is an extremely rare entity occurring in less than 0.03% of the population. This case identifies a single coronary artery with the left anterior descending coronary artery distally connected to the left pulmonary artery with development of pulmonary hypertension. To the author’s knowledge, this has never been documented in the English literature.

Keywords: Bland-White-Garland syndrome, left anterior descending coronary artery, pulmonary hypertension, single right coronary artery

INTRODUCTION

Hyrtl, in 1814, documented the first case of a single coronary artery (1). The rarity of a single coronary artery occurs in 0.03% of the population and constitutes less than 3% of all major coronary artery abnormalities. Forty-two percent of single coronary arteries originate from the right coronary sinus. These coronary artery abnormalities are usually detected at autopsy or as an incidental finding on coronary angiography (1–8). The use of modern technology: trans-thoracic and transoesophageal echocardiogram, non-invasive computed tomography coronary angiography (CTCA) and cardiac magnetic resonance imaging has made the diagnosis of this coronary artery anomaly easier (9–15).

Smith has stratified single right coronary arteries into three groups which correlate with morbidity, mortality and surgical significance. Type I has a normal course of left and right coronary arteries and was thought to have no clinical significance. Type II has one main initial vessel which then subdivides into branches analogous to the three main coronary arteries. In Type III, the distribution of coronary arteries has no correlation with the normal course (3). The index case does not fall into any fixed category described by Smith. The circumflex and right coronary artery distribution is normal...
but the distal left anterior descending artery is connected to the left pulmonary artery with a left to right shunt.

A clinically more significant classification was done by Ogden and Goodyer which not only looked at the origin but the course of the coronary arteries, around the trunk of the great vessels, the aorta and pulmonary artery (3). Anomalous coronary arteries are the second most common cause of increased mortality and sudden death when the coronary artery, especially the left anterior descending artery, courses between the trunk of the aortic and pulmonary artery. It is estimated that up to 20% of coronary artery abnormalities can result in syncope, chest pain, seizure, arrhythmia, angina, dilated cardiomyopathy with mitral regurgitation, congestive cardiac failure, myocardial infarction and may be the presenting symptoms in adolescents and adults with extensive development of collaterals between the coronary arteries (15–24).

Bland, White and Garland in 1993 described the clinical syndrome with the left coronary artery originating from the main pulmonary artery in an infant (24). The theory in this case was a retrograde flow from the pulmonary artery to cardiac myocardium, leading to myocardial infarction, which was substantiated by Sabiston et al in 1959. Myocardial infarction and death in infancy in 80% was the main method of presentation noted in the literature (5–7, 25). Augustsson et al (1961) and Rudolph et al (1962) demonstrated a left to right shunt into the pulmonary artery in Bland-White-Garland syndrome (5, 26–28).

This index case had a normal origin single ostium from the right coronary sinus, and a main branch with normal course of right coronary artery and circumflex. The left anterior descending artery, after its initial course and distribution of septal and diagonal, then curves distally and laterally to left pulmonary artery anteriorly, with a left to right shunt as noted in the Figure, with subsequent development of pulmonary hypertension. This has never been documented before as far as the author is aware.

The focus in this index case is on the unusual anatomy of the coronary arteries and their clinical presentation.

**CASE REPORT**

A 49-year old patient who is a known controlled idiopathic hypertensive patient for two years was symptomatic with shortness of breath for two months on mild exertion, functioning at New York Heart Association (NYHA) Classification level II. There was intermittent dependent pedal oedema with intermittent palpitations for two weeks, associated with orthopnea. She also had large leiomyoma (fibroids) causing bleeding post menopause. Total abdominal hysterectomy was planned. She was admitted for coronary angiogram.

On examination, she had pink mucous membranes, no cyanosis, no oedema, and distended superficial neck veins. There was no elevation of jugular venous pressure and no clubbing or loss of angle at the nail bed. Blood pressure was 100/80 mmHg, pulse rate 68/minute and respiratory rate 20/minute. Saturation was normal. Cardiomegaly was evident by displaced apex beat. First and second heart sounds were normal with no clinical evidence of cardiac murmur or signs of pulmonary hypertension. There were no clinical signs of left or right heart failure. A 30/40 large abdominal mass secondary to fibroids was noted. The liver was not palpable.

For medications, she was on Brinderin one tablet once daily, iron sulphate supplementation and acetylsalicylic acid 81 mg once daily.

---

Figure: Anomalous left anterior descending artery connected to left pulmonary artery from single coronary artery.

RCA – right coronary artery, LAD – left anterior descending, LPA – left pulmonary artery, MPA – main pulmonary artery
Blood investigations showed: haemoglobin of 13.2 g/dL and packed cell volume of 0.42. Urea and electrolytes, creatine phosphokinase (CPK) levels and liver function tests were all normal. There were no haematological markers of myocardial infarction. Electrocardiogram showed sinus rhythm with heart rate 92/minute, 1st degree heart block, left axis deviation and left ventricular hypertrophy. No ST-T segment changes or significant Q waves were noted. Chest X-ray confirmed cardiomegaly with an increase in the cardiothoracic ratio and there was normal vascularity of lung fields.

Echocardiogram showed right ventricular enlargement, dilated origin of right origin coronary artery, septal hypokinesia and borderline left ventricular hypertrophy. Origin of left coronary artery was not noted.

Coronary angiogram showed no left coronary artery. A single right coronary artery was found, which formed the circumflex and left anterior descending. The large left main artery emptied and was connected to the left pulmonary artery. Aortic pressures were 125/82, mean 101 mmHg. Right ventricular pressures were 85/0 and the main pulmonary artery pressures were 76/24, mean 46 mmHg. Right ventricular pressures were 68% systemic pressures and pulmonary artery pressures were 60% systemic pressures.

The index case was prescribed the following medications: slow releasing nifedipine at 20 mg twice daily, sildenafil 50 mg three times daily, aspirin 81 mg once daily, furosemide 40 mg once daily and spironolactone 25 mg once daily.

Total abdominal hysterectomy was done shortly after the coronary angiogram, with plans for continued treatment of pulmonary hypertension and correction of anomalous left coronary artery.

Eight months later, she developed gastroenteritis with upper gastrointestinal bleeding, due to oesophageal varices. Her haemoglobin was 17.4 g/dL, packed cell volume 0.53 and saturation was 91%. The index case also had severe right heart failure, liver failure, with subsequent renal failure. Development of pulmonary embolism led to the patient’s demise.

DISCUSSION
A single right coronary artery with normal course of the coronary arteries is of no clinical significance and is usually an incidental finding at post mortem or on coronary angiography. When the course of the coronary artery vessels occurs at an acute angle after origination from coronary sinus or is between the aortic trunk and pulmonary infundibulum, this can lead to sudden death, especially during exercise or on extreme exertion amongst athletes. It is one of the statistically significant causes of sudden death amongst athletes (1–8). Berbarie et al (2006) and Xu et al (2011) consider non-invasive CTCA to be the gold standard for diagnosis and delineation of coronary artery course and anatomy (13, 14).

The index case had no clinically significant problem from the left to right shunt for 49 years secondary to the left anterior descending artery’s connection to the left pulmonary artery. The development of pulmonary hypertension with the left anterior coronary artery having a left to right shunt is rare (5, 25–28).

The Bland-White-Garland syndrome occurring concurrently with a single right coronary artery is a possibility but the left anterior descending coronary artery would be expected to taper distally to the connection to the main pulmonary artery (5, 6, 8, 25–28). In the index case, the largest diameter of the anomalous left anterior descending artery was adjacent or at the origin to the right coronary sinus, with a left to right shunt. Hence, the index case is not considered to have Bland-White-Garland syndrome, which classically has right to left shunt with the flow from pulmonary trunk to coronary vessel.

The left to right shunt from the left anterior descending artery’s connection to the left pulmonary artery eventually led to pulmonary hypertension. The significant increase in haemoglobin and packed cell volume in eight months is haematologically indicative of the progression of her pulmonary hypertension. The clinical indicator signs of same were absence of clubbing, no continuous or systolic murmur, saturation of 90% with further impairment of exercise function (NYHA III) and the development of severe right heart failure leading to development of oesophageal varices. These are all markers haematologically and clinically of poor prognosis, in patients with development of Eisenmenger syndrome.

It is tantalizing to speculate in the index case that the haemorrhage from the fibroids delayed the development of polycythaemia, secondary to pulmonary hypertension, but this was suggested by the haematologic and clinical indices seven months post hysterectomy.

Surgical correction entertained would involve ligation of the connection to the pulmonary trunk with revascularization of the left coronary system with the use of a vein graft or left internal mammary graft to left anterior descending system. The novel approach of transcatheter occlusion of left anterior descending to the pulmonary artery could have been entertained because of the specific anatomy of the index case, who had extensive coronary collaterals, with a left to right shunt. Re-implantation of the left coronary artery system, which would lead to development of an acute angle from main right single coronary vessel, following disconnection from pulmonary artery, was not an option because of the short length of the vessel, and it was unable to reach the left coronary sinus of Valsalva. Wu and Xu have used pulmonary artery tube and Sarioglu et al the pericardial tube for translocation of an anomalous remote origin of coronary arteries when extension of coronary artery translocation is required and considered (21, 28–31).

The dilemma in the index case’s initial management post diagnosis is in the use of pulmonary hypertensive reducing agent sildenafil, which would reduce pulmonary arterio-
lar resistance, thus leading to an increase in the left to right shunt, facilitating further progression of pulmonary hypertension and “coronary steal”. The avoidance of antplatelet agents is usually recommended to avoid exacerbation of haemorrhage which is usually via haemoptysis secondary to necrotizing arteritis or pleuropneumonic lesions in the lungs developed from progressive pulmonary hypertension (6).

This case, which, to the author’s knowledge, has not been documented before, challenges the classification by Smith, and Ogden and Goodyer of single right coronary artery, with a coronary artery anatomy (3). This is an extremely rare case of anomalous left coronary artery connected to the left pulmonary artery, leading to the development of pulmonary hypertension, and should be considered as a differential diagnosis of pulmonary hypertension.

REFERENCES