Case Report

Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery
(The Bland-White-Garland Syndrome)

By

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Summary: Anomalous origin of the left coronary artery from the pulmonary artery (The Bland-White-Garland syndrome) is a rare congenital anomaly of the cardiovascular system. Almost all patients with this condition die during the first year of life. A one-year, 4-month-old boy was autopsied at Nagasaki University Hospital. In the heart, the small ventricular septal defect with aneurysmal and myocardial infarction as observed.

Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital anomaly of the cardiovascular system. The anomalous origin of the left coronary artery from the pulmonary artery was first reported by Abrikosoff in 1911. Bland, White and Garland (1933) first recorded an electrocardiogram in an infant with this condition and described the clinical and pathologic features. Only slightly more than 200 such cases have been reported in the literature (Wesselhoeft et al., 1968; Wilson et al., 1977; Askenazi and Nadas 1975). Anomalous origin of the left coronary artery from the pulmonary artery occurs in 1 of 300,000 children and constitutes approximately 0.24% of all congenital cardiac anomalies (Keith 1978). Despite its rarity, anomalous origin of the left coronary artery from the pulmonary artery is an important type of congenital heart disease since the mortality from this lesion has been estimated to be as high as 93% in the first year of life (Wesselhoeft et al., 1968). Moreover, many investigators recommend surgical correction of the lesion early in infancy (Grace et al., 1977; Levitsky et al., 1980; Neches et al., 1974; Pinsky et al., 1973, 1976). We are reporting on a case of the anomalous origin of the left coronary artery from the pulmonary artery.

Findings

A one-year, 4-month-old Japanese boy, 58 cm in height, 5 kg in weight was autops-
sied at Nagasaki University Hospital. At autopsy a cardiomegaly was found, and his heart weighed 82 g. The left coronary artery originated from the pulmonary artery just above the left posterior pulmonary valve (Fig. 1). It had an usual anterior descending and circumflex branch distribution pattern. The small ventricular septal defect, measuring 7 x 8 mm, with aneurysmal protrusion of membranous septum into the right ventricle was found. At gross lesion, multiple infarctions of papillary muscles and apical lesion of the left ventricle were observed as a whitish color (Fig. 2). At histological findings, these lesions were confirmed infarction by elastic fibers and collagen fibers staining (Senba 1982, 1983b, 1984a, 1984b) and by reticulum fibers staining (Senba 1983a, 1984c). There were chronic epicarditis and chronic endocarditis in the part of these region. The right lung weighed 70 g, and the left lung 30 g. The cut surfaces in both lungs showed forcal fresh bleeding and edematous change. Microscopically, diffuse hyperplasia of lymphoid tissue was seen in the subpleural region and interlobular septa of both lungs and the gastrointestinal tract. The liver weighed 288 g; cut surfaces showed slight congestion, and moderate fatty metamorphosis was seen under microscope. The pancreas weighed 23 g and was unremarkable. The spleen weighed 21 g, and cut surfaces showed slight congestion. The adrenal glands were normal. Both kidneys weighed 20 g, and cut surfaces showed slight congestion. There were petechiae in the epicardium. Similar petechiae were found in the peritoneum, bronchus, esophagus and intestinal mucosa.

Discussion

The condition of anomalous origin of the left coronary artery from the pulmonary artery should be suspected in an infantile type having marked left ventricular enlargement with or without ventricular aneurysm associated with electrocardiographic evidence of the left ventricular infarction. The seriousness of this anomaly is emphasized by the fact that in untreated cases, death secondary to myocardial infarction and cardiac failure occurs within the first 5 months. The reports have indicated that anomalous coronary arteries originating from the pulmonary artery may act as a channel for left-to-right shunt (Case et al., 1958; Edwards 1958; Sabiston et al., 1960). Therefore, a large portion of the coronary capillary bed, pass through arterial anastomoses, and enter directly into the pulmonary artery by retrograde flow (Edwards 1958; Sabiston et al., 1960), through the anomalous coronary with resultant myocardial ischemia. In infants, this mechanism is suspected. This concept supports that the artery which arises from the pulmonary artery does not carry pulmonary arterial blood into the myocardium. Rather, through collateral vessels between the left and right coronary arteries, blood is shunted from that artery which arises normally from the aorta into the anomalous artery and finally into the pulmonary artery. The myocardial ischemia present in such a system should be related to the escape of coronary arterial blood through the arteriovenous fistula.

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References


Explanation of Figures

Plate I

Fig. 1. The left coronary artery arising from the pulmonary artery.

Fig. 2. Cross-section of heart at a level 3 cm from apex showing myocardial fibrosis.