The Cardiology 2(1) : 14-15, 2006

An Incidental Diagnosis of Isolated Interrupted Aortic Arc in a 75 Year Old Female Patient with Acute Myocardial Infarction: The Oldest Case of Isolated Interrupted Aortic Arc

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Abstract: Interrupted aortic arc is defined as a loss of luminal continuity between the ascending and descending aorta. It is a rare congenital malformation and it is classified on the basis of the branches that originate from the proximal aorta. An interrupted aortic arc is usually associated with another congenital intracardiac malformation and patients with isolated interrupted aortic may survive to advanced age. Here we report a very rare case of an isolated interrupted aortic arc in a 75-year-old woman who presented with acute myocardial infarction.

Key words: Isolated interrupted aortic arc, myocardial infarction, adult

INTRODUCTION

Interrupted Aortic Arc (IAA) is a rare congenital malformation that occurs in 3 per million live births[1]. It is defined as a loss of luminal continuity between the ascending and descending aorta[2]. IAA is usually associated with an intracardiac malformation such as ventricular septal defect, patent ductus arteriosus, bicuspid aortic valve, left ventricular outflow tract obstruction or aortapulmonary window. Up to date limited number of cases have been described. In the literature available to us 13 cases of the adult form of an interrupted aortic arc could be established.

CASE REPORT

A 75-year-old woman with a past medical history of hypertension presented to emergency room of our institution with recent onset angina. On physical examination, there was 40 mmHg systolic blood pressure gradient between the arms and legs. Her peripheral extremity pulses were palpable, but lower limb pulses were extremely weaker than the upper limb pulses. Cardiac auscultation revealed a mild systolic murmur at the left sternal border. An electrocardiogram showed a sinus rhythm at rate of 82 beats per minute with ST segment elevation in the derivations D2, D3, aVF, V3R and V4R. Fibrinolytic therapy was started with streptokinase. Left ventricular concentric hypertrophy, segmental wall motion abnormality and minimal mitral regurgitation were detected on transthoracic echocardiography. She was hypertensive during the follow up period and renal artery doppler ultrasonography did not detect any abnormality. The patient was taken into the catheterization laboratory at the seventh hospital day. At cardiac catheterization the diagnosis of IAA was suspected when the aortic arc could not be entered despite repeated attempts. Contrast material injected into the descending aorta did not visualize the aortic arcus (Fig. 1). An ascending aortic angiogram that made through the right brachial artery showed the innominate, left common carotid and left subclavian arteries. There was a complete interruption just distal to the left subclavian artery (Fig. 2). As the associated cardiovascular anomalies such as patent ductus arteriosus or ventricular septal defect were not

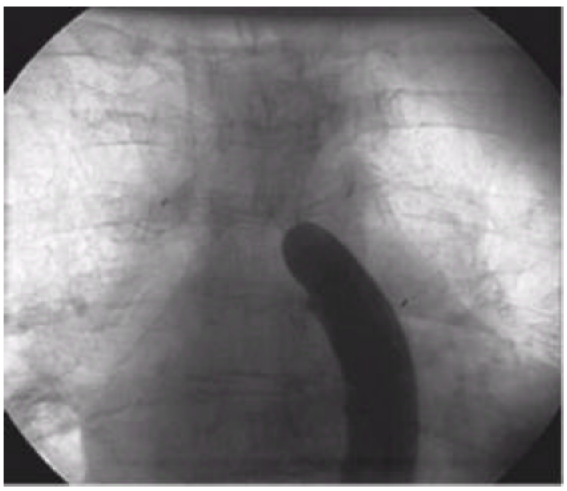

Fig. 1: Selective injection of contrast into the descenden aorta demonstrates the interruption. Aortic arcus is not visualised and no arc vessel arose from the descending aorta

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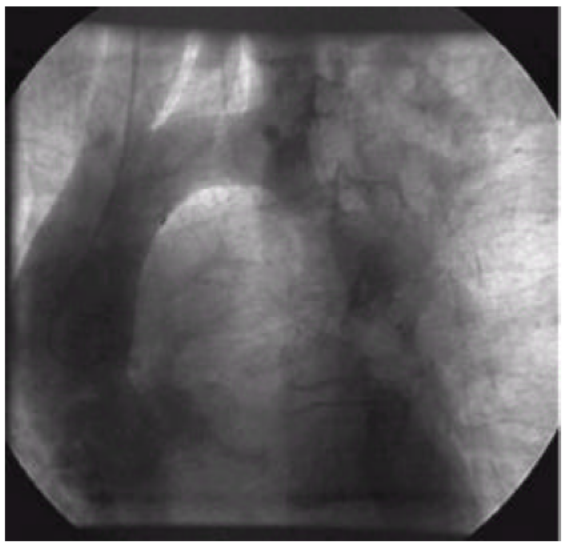

Fig. 2: Aortography through the right brachial artery showed the vessels originating from the arcus and an abrupt interruption distal to the left subclavian artery

found, the diagnosis of isolated IAA Celoria-Patton Type A was made. A coronary angiography disclosed a 70% left anterior descending artery and an 80% right coronary artery lesion. The patient refused to have an operation.

DISCUSSION

Of the various forms of congenital heart disease, aortic arc interrupted, a conotruncal heart defect is relatively uncommon[3]. It is so rare that occurs in 3 million per live births[2]. IAA is defined as abscence of luminal continuity between the ascending and descending aorta and occurs in about 1% of the patients with congenital heart defects[4].

The interruption is classified on the basis of the branches that originate from the proximal aorta and there are 3 types: Type A has the interruption distal to the left subclavian artery. Type B occurs between the left carotid and the left subclavian artery. Type B interruption is the most common type and there is an association with DiGeorge's syndrome. Type C, the interruption occurs between the two carotid arteries[5].

Abnormal involution of the third and fourth aortic arches[6] and decreased antegrade blood flow in the ascending aorta due to the presence of ventricular septal defects and left ventricular outflow obstructions[7] are the suggested pathogenetic mechanisms for IAA.

In infants, its clinical presentation involves severe congestive heart failure[8]. Unless treated surgically, 75 percent of patients with IAA die within the first month of life and 90 percent within the first year[9]. Patients with isolated IAA not accompanied by other cardiovascular anomalies may survive to advanced age[10]. In adults, the presentation ranges from a lack of symptoms to limb swelling with differential blood pressure in all extremities. In the present case, IAA was detected incidentally during the coronary angiography. IAA should be suspected when a blood pressure gradient detected between the upper and lower limbs even in a geriatric asymptomatic patient. To our knowledge, this is the oldest reported case in the literature of IAA.

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