

A Rare Occurrence of Congenital Coronary Ectasia Combined with Ventricular Septal Defect

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Abstract

Idiopathic or congenital coronary artery ectasia (CAE) is an uncommon form of coronary artery disease. In adults, coronary artery ectasia is usually associated with atherosclerotic change and is well recognized clinical entity encountered during cardiac catheterization. Coronary artery dilatation in pediatric is usually associated with the sequelae of Kawasaki disease. Congenital coronary artery ectasia is uncommon and rarely reported in children. We present a case of an infant who have dilated coronary artery, she also had ventricular septal defect (VSD) and heart failure who had received VSD repair at infancy. There was no obstructive coronary artery disease, and no cause for the lesions could be identified. The prognosis and optimal management of such patients remains unknown. Antiplatelet therapy might be necessary for the patient to remain free of myocardial ischemia.

Keywords: Congenital coronary artery ectasia; Coronary fistula; Congenital heart disease

Introduction

Coronary artery ectasia (CAE) is a well-recognized but relatively uncommon finding encountered during diagnostic coronary angiography in adult [1]. In infant, coronary dilatation is mostly due to coronary complications of Kawasaki disease (KD). Also, coronary fistula may cause associated coronary artery dilation. Congenital coronary artery ectasia is an uncommon lesion and frequently unrecognized incidental finding with other congenital heart disease. Chakrabarti et al. had reported three cases of congenital coronary ectasia associated with left heart obstructive lesions [2]. In our patient there is no acquired coronary inflammatory aneurysm or coronary fistula. However, the patient has a perimembranous ventricular septal defect that needed repair in infancy. Ectatic coronary arteries are known to be associated with potential complications, resulting from coronary stenosis and impaired coronary perfusion. In isolated CAE, prognosis is better and anti-platelet drugs are the mainstay of treatment [3].

Case Report

A 20 days old female neonate was brought to emergent department due to dyspnea and poor appetite. Subsequent lab exam showed a large ventricular septal defect and patent ductus arteriosus (PDA) with congestive heart failure. She then received surgical intervention with Dacron Patch VSD repair and PDA ligation. At age 2 months, she suffered an episode of urinary tract infection (UTI) and was treated with antibiotics. However, echocardiogram followed up at age 3 months showed an abnormal left coronary arterial flow. The previous echocardiograms showed a large VSD color jet and coronary artery was not adequately evaluated. Her physical examination revealed a healthy well grown infant with a body height of 65 cm, body weight of 5.2 kg. Cardiovascular examination revealed a surgical scar over anterior chest wall, regular heart beats with no heave, normal maximal

point of impulse over left 4th intercostal space at mid clavicular area. There is a split second heart sound over left upper sternal border due to postsurgical right bundle branch block. Laboratory data showed normal blood cell count and normal troponin I level. Chest x-ray showed no cardiac enlargement. An electrocardiogram showed normal sinus rhythm with incomplete right bundle branch block pattern. Echocardiogram revealed an abnormal mosaic flow over high parasternal area (Figure 1).

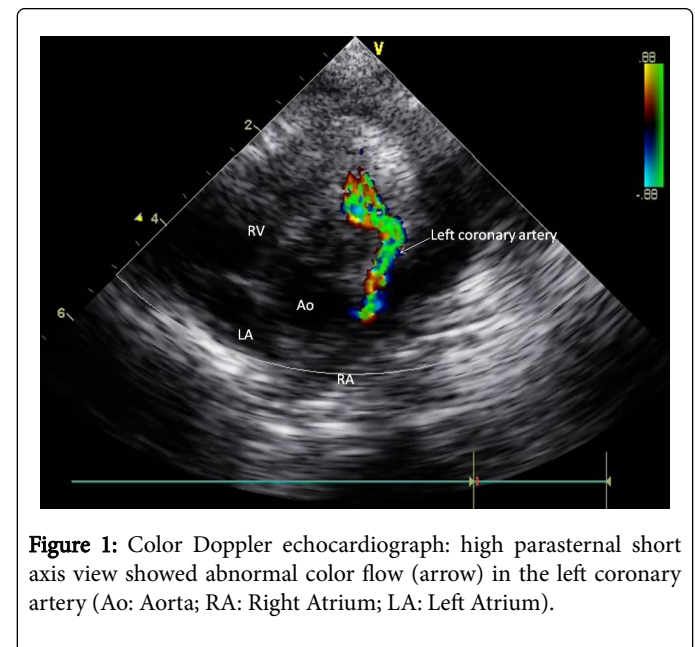


Figure 1: Color Doppler echocardiograph: high parasternal short axis view showed abnormal color flow (arrow) in the left coronary artery (Ao: Aorta; RA: Right Atrium; LA: Left Atrium).

Due to the persistent abnormal left coronary flow, the family agreed for catheterization to rule out a coronary fistula. Angiographic study revealed a large left anterior descending coronary ectasia (Figures 2A and 2B) with pulsatile flow pattern. Traced back her history, there are no abnormal skin rashes except diaper rashes, conjunctivitis or prolonged fever. There was no history suggestive of a possible previous

Kawasaki disease of this patient. The coronary angiography is compatible with localized dilatation of coronary artery lumen exceeding the largest diameter of an adjacent normal vessel more than 1.5 fold. Hence, congenital coronary ectasia instead of coronary aneurysm due to Kawasaki disease or coronary fistula was diagnosed. Subsequent echocardiogram showed the same mosaic flow over left coronary artery. She received clinical follow up with antiplatelet agent prescribed.

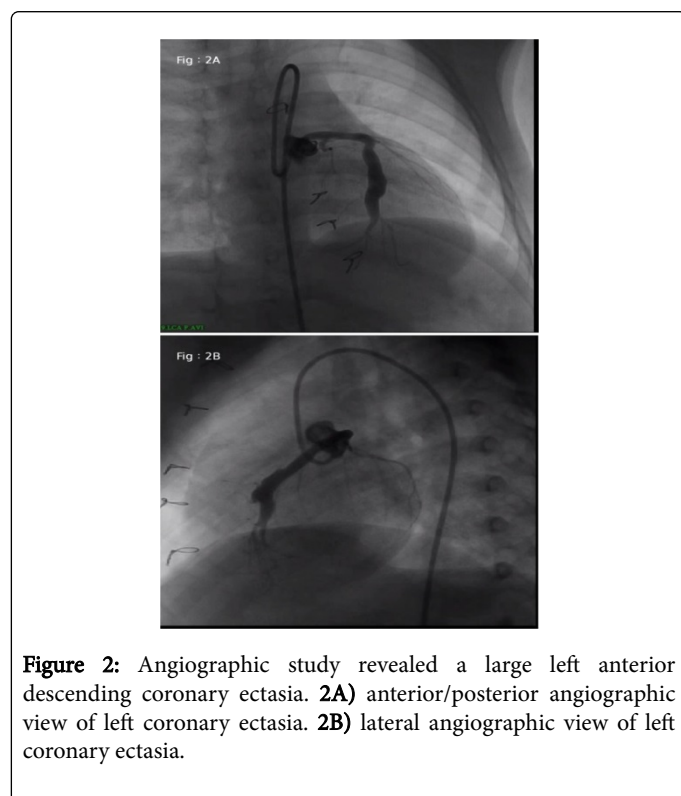


Figure 2: Angiographic study revealed a large left anterior descending coronary ectasia. **2A)** anterior/posterior angiographic view of left coronary ectasia. **2B)** lateral angiographic view of left coronary ectasia.

Discussion

CAE has been defined as localized or diffuse non-obstructive lesions of one epicardial coronary artery with a luminal dilatation exceeding the 150% of a normal adjacent vessel diameter [4]. It is characterized by an abnormal dilatation of coronary artery, represents a form of atherosclerotic coronary artery disease, seen in 3-8% of patients undergoing coronary angiography [1]. Abnormal dilatation of a vessel could be long segmental involvement of partial aneurysm formation. Usually, coronary ectasia refers to involvement of the whole vessel while partial involvement is better called aneurysmal dilatation [5]. Beside the association with Kawasaki disease and coronary fistulae, the aetiology of coronary dilatation in the paediatric age group is not very clear. In a small percentage of patients CAE can be congenital in origin [6]. The differentiation between congenital and acquired coronary dilatation may often be difficult, despite the exclusion of other associated diseases.

The etiopathogenesis of this condition is poorly understood. Based on its association with aortic aneurysm, coronary ectasia is considered to be caused by genetic abnormalities [7]. It is interesting that our patient, who received surgical correction of VSD, also have CAE not

associated with other aortic vessel anomaly. Nevertheless, a scrutinized review of the clinical history only revealed she had a UTI problem at age 2 month which responded to antibiotics treatment. There is no history of prolonged fever, skin rashes or any skin desquamation. Also the long segmental dilatation of left coronary artery is not usually seen in KD. The dilated long segmental coronary artery is compatible with the definition of ecstatic artery (the diameter is more than 50% of length) and due to no other etiology, is preferred a congenital lesion .

Dilated segments are thought to cause the coronary flow disturbances in the affected vessel. The lesions cause decrease coronary flow velocity may then predispose to myocardial ischemia, positive exercise stress test, or acute coronary syndromes [8]. In contrary to atherosclerotic coronary artery disease, the medical management of patients with CAE has not been adequately addressed. Previous studies based on the significant flow disturbances within the ectatic segments suggested chronic anticoagulation as main therapy [9]. However, the natural history of this lesion in such a young child is not fully known. It is very likely that these patients have an increased risk of ischemic coronary disease in later adult life.

Our patient had CAE and ventricular septal defect that received VSD repaired. The coronary dilatation only involved one left anterior descending coronary artery. In a previous report about isolated CAE, the author concluded the prognosis is better and suggested anti-platelet drugs are the mainstay of treatment [10].

Long term follow up is needed to see if coronary arterial ectasia will remain involving single coronary branch or will extend slowly to involve all coronary arteries. And the possibility for the disease to involve the aorta itself and end in aortic aneurysms or dissection needed close monitoring.

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