Dilated Cardiomyopathy Due to Aortic Coarctation in Childhood

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ABSTRACT

Dilated cardiomyopathy (DCM) is rarely seen secondary to Coarctation of aorta. Twenty-one months old male was referred to our hospital because of tachypnea, coughing and cardiomegaly. Echocardiography revealed most importantly CoA. Because sufficient improvement could not be acquired with interventional balloon angioplasty, he had surgical operation after one month. Coarctation of aorta should be taken into consideration in the differential diagnosis of causes of DCM in chidren. It is also so important that before we decide definitely for any patient presenting with a DCM as an idiopathic cardiomyopathy, we must rule out all possible specific causes of myocardial dysfunction. Because some of specific etiological factors of cardiomyopathies can be completely treatable, just like in our patient.

Key words: *Coarctation of the aorta, dilated cardiomyopathy, child*

Çocukluk Döneminde Aort Koarktasyonu Sonucu Gelişen Dilate Kardiyomiyopati

Aort koarktasyonuna bağlı dilate kardiyomyopati ise nadir görülmektedir. Yirmibir aylık erkek hasta öksürük, takipne ve kardiyomegali mevcuttu. Ekokardiyografide önemli derecede aort koarktasyonunun olduğu izlendi. Balon anjiyoplasti ile yeterli düzelme olmaması üzerine bir ay sonra cerrahi girişim yapıldı. Bu vaka aort koaktasyonu tanısında eş zamanlı brakial ve femoral nabız muayenesinin ne kadar önemli olduğunu ve gecikilmiş vakaların dilate kardiyomiyopati adayı olabileceklerini hatırlatmak amacıyla sunuldu. Çocuklarda dilate kardiyomiyopati nedenlerinin ayırıcı tanısı yapılırken aort koarktasyonunun akılda bulundurulması gerekir. İdiyopatik dilate kardiyomiyopati tanısını kesin olarak koymadan önce miyokard disfonksiyonu yapan muhtemel spesifik nedenlerin araştırılması büyük önem arzeder. Çünkü bizim vakamızda da görüldüğü üzere, kardiyomiyopatinin bazı nedenlerinin tamamen tedavi şansı bulunmaktadır.

Anahtar kelimeler: *Aort koarktasyonu, dilate kardiyomiyopati, çocuk*

INTRODUCTION

Cardiomyopathy is a common disorder resulting from a variety of causes that lead to impaired cardiac function and often result in congestive heart failure. Idiopathic dilated cardiomyopathy (DCM) is defined as presence of left ventricular dilatation and systolic dysfunction in the absence of abnormal loading conditions (valvular, congenital, hypetension, coronary artery anomaly, pulmonary vascular disease etc.) or coronary artery disease sufficient to cause global systolic impairment (1,2). Coarctation of aorta (CoA) is presented with congestive heart failure in infancy. But later, hypertension and heart murmur are the most common signs of this congenital heart defect (3). Up to our knowledge, CoA presented as a DCM, is rarely reported (4-8). Patients diagnosed as DCM should be evaluated for long segment thoraco-abdominal CoA. Early diagnosis and treatment of CoA are important to prevent development of secondary DCM (7).

CASE

Twenty-one months old male was referred to our clinic because of tachypnea, coughing and cardiomegaly. He was also suffering from dyspnea and poor feeding since birth. Physical examination revealed; weight: 8900 g (<3 percentile), height: 77 cm (3-10 percentile), weight for height: 80-90%, respiratory rate: 32/min, heart rate: 132/min, weak femoral pulses according to brachial pulse, right arm blood pressure: 130/90 mmHg, right leg systolic blood pressure: 80 mmHg. Dyspnea and tachypnea were also noted. Cardiac examination was significant for hyperdynamic precordium with a S3 gallop rhythm and a grade 1-2/6 systolic ejection murmur maximally heard on apex and at the left interscapular area. In addition to these findings, hepatomegaly and pectus carinatum deformity were determined. Examination of musculoskeletal system was normal for his age. He had no significant family history and was not on regular medication. Laboratory investigation revealed low hemoglobin level and leukocytosis; 10.3 g/dl and 18.100/mm3 respectively. Serum electrolytes, cardiac enzymes, erythrocyte sedimentation rate, and C-reactive protein were all normal. Blood cultures were sterile. Electrocardiography showed normal QRS axis, sinus tachycardia, T wave abnormality and left ventricular hypertrophy with 'strain' pattern (Figure 1). Increased pulmonary vascularity and also cardiomegaly (cardiothorasic index:0,66) were both determined with telecardiography. Echocardiography of the patient revealed markedly dilated and hypokinetic left ventricle, right deviation of interventricular septum, abnormal systolic function of left ventricle and most importantly CoA in suprasternal view (Figure 2). Other causes (coronary artery anomalies, myocarditis, arrhythmias, valvular etc.) of cardiomyopathy were also ruled out before definite diagnosis.

Thus, DCM presumably resulting from the isolated coarctation was diagnosed. Digoxin, diuretic and angiotensin-converting enzyme inhibitor medications were administered. The infant was referred to an other center to be performed interventional balloon angioplasty. Coarctation of aorta was also confirmed with angiography and additionally, a markedly dilated and poorly contracting left ventricle and aortic collaterals were also determined (Figure 3). The aortic valve and the origins of the main coronary arteries were normal. Because of insufficient improvement with angioplasty, he had surgical operation with resection and end-to-end anastomosis after one month of diagnosis. Normal blood pressure, and weight and height gain were noted ten months later. In addition, echocardiography revealed approximately normal left ventricular systolic function and smaller dimensions of left ventricle than before. Initial and final echocardiographic findings are presented in Table 1.

DISCUSSION

Dilated cardiomyopathy is an important cause of chronic congestive heart failure in infants and children. Cardiomyopathies are heart muscle diseases, which have been defined by their central hemodynamics and macropathology and divided in five major forms: restrictive, dilated, hypertrophic, arrhythmogenic right ventricular, and unclassified (left ventricular non-compaction) cardiomyopathies (1). Furthermore, the most recent World Health Organization/World Heart Federation's definition also comprises, among the specific cardiomyopathies, inflammatory cardiomyopathy as a distinct entity, defined as myocarditis in association with cardiac dysfunction. Idiopathic, autoimmune, and infectious forms of inflammatory cardiomyopathy were recognized. Viral cardiomyopathy has been defined as viral persistence in a dilated heart (9,10). Although a variety of etiological factors; such as muscular dystrophies, myocarditis, Kawasaki disease, nutritional (thiamine, carnitine, selenium) deficiencies, drugs and tachycardiomyopathy

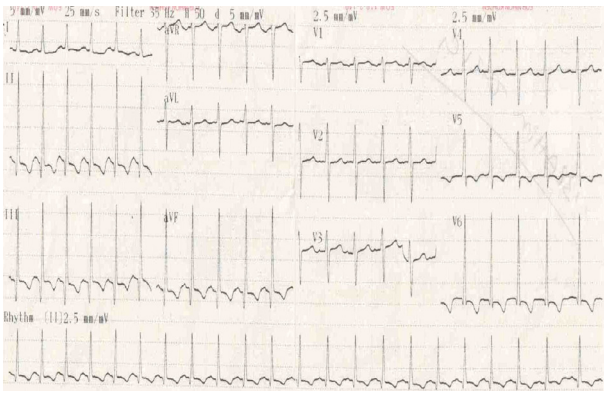

***Figure 1**. Electrocardiography showed normal QRS axis, sinus tachycardia, T wave abnormality and left ventricular hypertrophy with 'strain'*

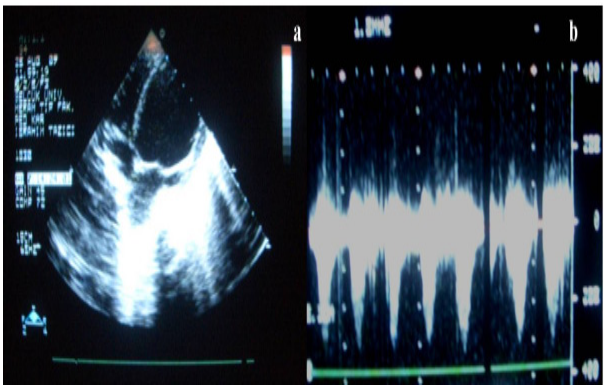

***Figure 2**. a) Markedly dilated left ventricle and right deviation of interventricular septum, b) Velocity of descending aorta (3.5-4 m/sec) and "shoulder sign" are seen*

have been listed, most patients with echocardiographically documented DCM do not possess a demonstrable cause. In the European Society of Cardiology position statement published in 2008, cardiomyopathies were defined as myocardial disorders in which the heart muscle is structurally and functionally abnormal, and in which hypertension, valvular and congenital heart disease, pulmonary vascular disease, myocardial ischemia as a result of coronary artery disease and congenital coronary anomaly (the most common being anomalous origin of left coronary artery from pulmonary artery) are absent or do not sufficiently explain the observed myocardial abnormality (1). This term usually denotes a dismal prognosis in short of cardiac transplantation. However, several organic diseases of the heart can result in right or left ventricular dysfunction resulting in congestive heart failure and prompting the physician to label them as cardiomyopathy; the etiological factor can be overlooked as it produces very subtle features. Therefore, before labelling any child as cardiomyopathic, all possible causes of ventricular dysfunction must be excluded by clinical and investigative means. The causes of "treatable cardiomyopathy" include mechanical factors as critical aortic and pulmonic stenosis, severe CoA in an infant and aorta-arteritis in an older child and some of the persistent arrhythmias (2).

Coarctation of aorta is presented with congestive heart failure in infancy, but later hypertension and heart murmur are the most common signs of this congenital heart

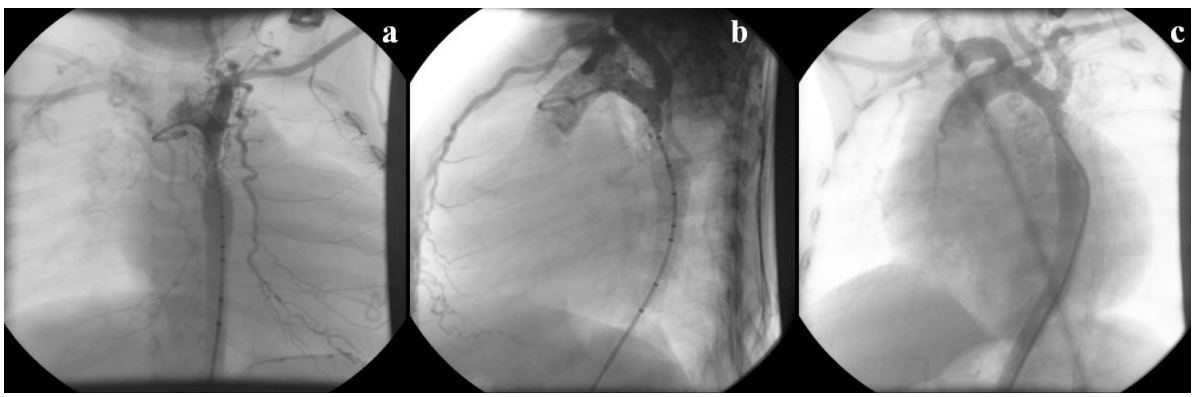

***Figure 3**. Retrograde aortogram reveals the aortic coarctation. Also note the prominent collateral arteries and marked cardiomegaly*

***Table 1**. Initial and final (after ten months from surgery) echocardiographic results.*

	Initial Results	*Final Results*
LVEDD	*55 mm*	*39 mm*
LVESD	*45 mm*	*28 mm*
Ejection Fraction (EF)	*37 %*	*56 %*
Fractional Shortening (FS)	*18%*	*29 %*
Velocity of descending aorta	*3.5-4 m/sec (50-64 mmHg)*	*3 m/sec (36 mmHg)*
Mitral regurgitation	*Grade 1*	*Absent*
Aortic regurgitation	*Mild*	*Absent*

LVEDD: Left ventricular end diastolic dimension, LVESD: Left ventricular end systolic dimension

defect (3). Coarctation of aorta presenting as a DCM has previously been reported in infants and adults (4-8). Coarctation of aorta can cause hypertensive cardiomyopathy at earlier life. After a period of compensatory phase, the relatively long-standing increased systemic afterload might exhaust the myocardium and then myocardial remodeling develops as a consequence of 'afterload mismatch'. Later, several macroscopic events take place; there may be myofibrillar lysis, increased number of lysosomes, distortion of the sarcoplasmic reticulum, reduction in the surface density of the key tubular system, and fibrous replacement of cardiac cells (11). All these changes may lead to DCM and failure of the cardiac pump (11,12). Poor myocardial function in DCM triggers a sequence of compensatory mechanisms, mediated through the renin angiotensin and sympathetic system (neural and humoral) and a number of vasodilatory molecules. These favour myocardial and peripheral vascular remodeling by necrosis, fibrosis and apoptosis which ultimately does more harm than good (13). Its pathogenesis is related to increased afterload and probably similar to valvular (stenosis) or hypertensive DCM (12).

In literature, Hwang et al. and Cetin et al. have reported improved left ventricular functions of the infants and newborns after 8-15 months from the coarctation repair (6,14). The direct cause and effect relationship was supported by our patient's rapid recovery from the DCM after the coarctation repair. Another study noted that 30% of native CoA cases developed myocardial dysfunction because of hypertensive cardiomyopathy. Ultimately, marked improvement of left ventricular function was acquired with balloon angioplasty (15). Another two adult patients with severe DCM were successfully managed with combined orthotopic heart transplantation and CoA repair (16,17). Therefore, early diagnosis and management of the treatable causes of DCM are quite important. Left ventricular function of our case also improved gradually and recovered completely after 10 months postoperatively.

Consequently, the present case is presented to remind that CoA can lead to DCM. The simultaneus palpation of strong brachial and weak femoral pulses should lead the clinician to suspect about CoA in any case. Therefore, routine examination of blood pressure and pulses in both lower and upper extremities should be performed in all patients. Coarctation of aorta should be taken into consideration in the differential diagnosis of causes of DCM in chidren. It is also so important that before we decide definetely for any patient presenting with a DCM as an idiopathic cardiomyopathy, we must rule out all possible specific causes of myocardial dysfunction. Because some of specific etiological factors of cardiomyopathies can be completely treatable, just like in our patient.

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