IDIOPATHIC INFANTILE CARDIOMYOPATHY

(A Case Report)

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Introduction

Cardiomyopathies commonly occur in infants and children. They are considered primary when no associated cardio-vascular or systemic disease is present or secondary when the myocardial abnormality is related to an associated disease. Primary cardiomyopathy may be classified as hypertrophic, dilated/congestive or restrictive. They usually present as congestive cardiac failure (CCF), chest pain, arrhythmias or sudden death [1].

Case Report

A 17-day-old male neonate, the only sib of a nonconsanguineous parents and with no relevant antenatal or perinatal history was admitted to the Hospital in April 96 with history of single episode of early morning vomiting followed by severe respiratory distress. He weighed 3.5 kg and clinical examination revealed a listless, lethargic, afebrile baby with a heart rate of 148/min, regular and respiratory rate 84/min. Respiration was laboured with intercostal retractions and mild cyanosis. There were basal crepitations and bilateral wheeze on auscultation of the lungs. Precordium was normal. Apex beat was displaced just outside the mid clavicular line in 5th inter costal space (ICS) and there was no para sternal heave or palpable P2. S1 and S2 were normal, S3 was present and a grade II/VI ejection systolic murmur was heard at the left sternal edge (LSE) in the 4th ICS. There was moderate hepatomegaly. No other congenital anomalies were detected. Based on above clinical profile, he was diagnosed as a case of aspiration pneumonia superimposed upon congenital heart disease left to right (L-R) shunt leading to CCF. He was managed with restricted IV fluids, oxygen, IV Ampicillin/Gentamicin, nebulisation, diuretics and digitalisation. Investigations revealed Hb 14.5 g per cent TLC 16,800/cm with a polymorphonuclear leucocytosis of 79 per cent. Chest radiograph revealed mild cardiomegaly (CT ratio 0.7) and few patchy infiltrates in the lung fields. ECG was normal. With the treatment as outlined above, the baby showed marked improvement within 36 hrs with stabilisation of cardiac and respiratory status. The cardiac murmur disappeared but S3 persisted. After one week of hospitalisation, the baby was suckling well at the breast, there was no evidence of cardiac decompensation and lungs were clear. He was put on decongestive therapy and sent for echocardiography which revealed decreased ejection fraction (36%) and fractional shortening of the left ventricle (LV). All the valves were normal in anatomy and motion. The LV was dilated with global hypokinesia.

Colour doppler studies revealed mild mitral regurgitation (MR) (grade 1) and patent fossa ovalis with small L-R shunt. With these findings he was diagnosed as a case of idiopathic dilated cardiomyopathy (IDC). At present he is stabilised on decongestive therapy. Repeat echocardiography after 3 months continues to show dilated LV with global hypokinesia with no evidence of MR.

Discussion

Sudden onset CCF during neonatal period occurs due to a variety of cardiac conditions which include congenital valvular abnormalities, L to R shunts (VSD, PDA) myocarditis and cardiomyopathies.

IDC is a relatively rare disorder characterised by dilated and poorly contractile ventricles, especially the LV and absence of inflammatory cells in the myocardium [3]. Ventricular dilatation is associated with reduced ejection fraction and increased end systolic volume, as well as, left atrial dilatation [4]. It is difficult to distinguish clinically from endocardial fibroelastosis and myocarditis and at times definitive confirmation of diagnosis is obtained only at necropsy. The etiology of this condition is unknown and is probably multifactorial. Endomyocardial biopsy studies in some patients considered clinically to have IDC have reported a substantial incidence of inflammatory myocarditis and viral

Fig. 1 : 2-D echo showing dilated left ventricle.

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infection has also been implicated in the etiology [5].

In IDC, all age groups are affected and in infancy it can occur as an acute illness in the setting of severe CCF. Usually onset is insidious. Patients present with signs of heart failure with hepatomegaly, pulmonary congestion and a prominent S3. There may be no murmur or only a soft ejection systolic murmur along LSE. A murmur typical of MR may be present at the apex [6]. All the above features were detected in our patient at initial presentation. After stabilisation of cardiac status, the soft systolic murmur at LSE disappeared. ECG does not contribute much to the diagnosis and usually shows nonspecific changes. Chest radiograph demonstrate cardiomegaly of varying proportions.

Two dimensional echocardiography shows increased left and right ventricular and left atrial dimensions [4]. In our case only the left ventricle was involved. It also demonstrates decreased left ventricular fractional shortening as well as regional contraction abnormalities in cases of dilated cardiomyopathy evolving from myocarditis [4]. It is useful in longitudinal evaluation of children with congestive cardiomyopathy and in assessing effects of medical treatment.

It has been difficult to define the clinical course and prognosis for children with IDC. In those who im-

prove and survive, it is difficult to distinguish clinically from conditions like myocarditis and endocardial fibroelastosis. It usually has a poor prognosis in children and adolescents. Conservative management with aggressive decongestive therapy may help in symptomatic improvement but very rarely it will result in normalization of systolic function [5]. The only hope for prolonged survival is cardiac transplantation.

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