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ABSTRACT

Dilated cardiomyopathy (DCM) in children is characterized by left ventricular dysfunction in the absence of chronic increased afterload or volume overload. There is no local study describing DCM in children. Diagnosis requires a high index of suspicion as it may easily be missed or diagnosed late which results in high morbidity and mortality. We present 2 cases of female infants admitted with DCM to our paediatric intensive care unit at MP Shah Hospital who did well on medical management. Determining the etiology of DCM remains a challenge in resource limited setting as imaging, viral markers and genetic studies may not be readily be available, hence clinical judgement remains the key in diagnosis and management. Cardiorespiratory therapies and vasoactive medication such as diuretics, inotropes and vasopressors and vasodilators used in management have shown to improve outcomes.

INTRODUCTION

Dilated cardiomyopathy (DCM) in children is characterized by left ventricular dysfunction in the absence of chronic increased afterload or volume overload. Studies done in Africa shows it is more common in children under 5 years. There is no study in Kenya describing the prevalence of DCM. DCM is associated with genetic disorders, malnutrition, endocrine/metabolic disorders to infection, but identifying the etiology is challenging in resource poor settings [1]. Myocarditis is due to inflammation of the walls of the heart and it is the most common cause of heart failure in children. Research shows myocarditis progresses to DCM from distinct viral injury or inflammation by host via immunological mechanisms [2]. These may precipitate cardiogenic shock (defined as decreased cardiac output and evidence of tissue hypoxia with adequate intravascular volume), which is the 3rd most common type of shock in children. The infant presents with tachycardia

as the major compensatory mechanism to improve systemic perfusion [3].

Factors that point towards diagnosis of myocarditis include recent viral illness, including fever and presence of either elevated inflammatory markers or elevated serum troponin or CKMB. A cardiac MRI and endomyocardial biopsy are useful in distinguishing between DCM and myocarditis [3], but this is not available in resource limited settings, hence high index of clinical suspicion. Chest radiography, electrocardiogram and echocardiogram are used for diagnosis.

The initial management goal is to restore peripheral perfusion using diuretics, inotropes and vasodilators and manage other associated symptoms. Cardiac transplant may be needed in some children with DCM and myocarditis.

CASE REPORT

We report 2 cases of female infants diagnosed with DCM.

Case 1

A 10-month previously well infant, who was transferred from a peripheral hospital and admitted to our PICU with 5-day history of fever on/off with mild coryza and 1-day history of worsening respiratory distress, skin rash and poor feeding. There was no history of cough, wheezing, vomiting, diarrhoea and convulsions. There was no history of travel to malaria endemic zone. Was admitted 2 months ago in a private facility and managed for pneumonia. Family history was unremarkable.

On admission she was febrile (40 degrees Celsius), in respiratory distress (80 breaths/minute, lower chest wall in drawing, flaring of the alae nasae and oxygen saturation of 70% at room air) and had tachycardia (190 beats/minute). She had abnormal perfusion (capillary refill time of 6 seconds, cold extremities up to the knee and elbow, weak radial pulse and GCS of 3/15). Blood pressure was 76/61 (68). She had no cyanosis, no lymphadenopathy and no oral thrush. On systemic exam, the air entry was equal bilaterally with no rhonchi or crackles, apex was at the 6th intercostal space and mid clavicular line, both heart sounds were heard with no murmurs heard and had a hepatomegaly of 4cm below costal margin. The skin appeared mottled with generalized macular rash and mild conjunctival injection in both eyes.

Investigations done on admission included venous blood gas (PH 7.013, PCO2 20mmhg, HCO3 5.1, BE - 26 and lactate 15.1). Random blood sugar 1.3mmol/L. Septic screen (WBC 15.4; N 76%; L 16%, Hb 12.5g/dl, platelets 76, CRP 7.17, procalcitonin 9.99 and had a negative blood culture). Cardiac markers were elevated (Troponin T 199 ng/L and CKMB 440 U/L) and kidney function and electrolytes were normal, liver functions were deranged with elevated AST (5504 U/L) and ALT (2727 U/L) and low albumin (27g/dl). Viral markers for HIV, CMV, Hepatitis B and C were negative. Chest radiograph showed cardiomegaly with normal lung fields, electrocardiogram showed sinus tachycardia and echocardiogram shows severe dilated cardiomyopathy with very low ejection fraction 20%.

An initial impression of respiratory tract infection and shock was made at the peripheral hospital and management was initiated. The infant was placed on oxygen, given 10% dextrose bolus at 5mls/kg, 3 boluses intravenous crystalloid fluid at 20mls/kg, packed red cells at 10mls/kg and ceftriaxone antibiotics. In our unit a diagnosis of cardiogenic shock was made and was

given, dopamine was started at 7mcg/kg/minute and later milrinone at 0.5mcg/kg/minute and frusemide 0.15mg/kg/hour in view after laboratory and imaging done. Later in the day developed one episode of supraventricular tachycardia and this corrected with 3 doses of adenosine.

The infant continued to improve, and repeat echo done after one week showed recovery phase of dilated cardiomyopathy and started on digoxin, frusemide, enalapril and spironolactone. She is currently doing well, growing well and medications have been tapered down.

Case 2

A 2-month-old infant admitted with cough, inability to feed and fever for 1 day as a readmission. A week ago, was admitted with fever, difficulty in feeding and diarrhoea (adenovirus positive) for four days. On this admission she had no diarrhoea, passed urine well and noted to have normal milestones. On initial examination was found to be febrile (38.5 degrees Celsius), in respiratory distress (had lower chest wall indrawing, flaring of ala nasae, respiratory rate of 50/min, grunting, cyanosis and oxygen saturation of 80% at room air). She had normal blood pressure and capillary refill time of 2 seconds. On auscultation had bilateral basal crackles and widespread wheeze. Apex was at 5 intercostal space and mid clavicular line with a systolic murmur at the left lower sternal border (grade 3). There was no hepatomegaly and other systems were essentially normal.

An initial diagnosis of bronchiolitis/severe pneumonia was made, and the baby was started on oxygen, maintenance fluids, crystalline penicillin, ceftazidime and hypertonic saline. Initial investigations showed normal haemogram (haemoglobin of 11.7g/dl, white cell count of 9.33 and platelets of 167), blood slide for malaria negative, normal procalcitonin of 0.086 and normal kidney and liver function tests. Respiratory distress worsened while on oxygen and chest radiograph done the same day showed enlarged cardiac shadow with normal lung fields and ECHO done showed dilated cardiomyopathy with ejection fraction of 33%. No cardiac markers were done then. A diagnosis of cardiogenic shock secondary to DCM was made. The infant was then transferred to PICU with respiratory distress, poor perfusion, hypotension and was intubated and started on adrenaline 0.1mcg/kg/min and milrinone 0.5mcg/kg/min infusion. The infant improved and inotropes were stopped after 3 days. She was started on oral digoxin, Aldactone, enalapril and Lasix and repeat ECHO after 5 days showed improvement in ejection fraction to 46%. The infant was off oxygen after a week, feeding well and was discharged to be followed up in the cardiology clinic where she has continued to improve on the oral medication.

DISCUSSION

Diagnosis of Cardiogenic shock, DCM and myocarditis is a challenge in resource limited settings where laboratory and imaging may not be readily available which may delay or result in misdiagnosis, hence clinical diagnosis requires a high index of suspicion as the diagnosis overlaps other causes of shock as seen in our case. The diagnosis was made in our case after ruling out lung pathology/hypovolemic/septic shock and a chest radiograph that showed cardiomegaly raised the suspicion of cardiac pathology. A study done by Yamini et al in 2008 showed respiratory infection was the most common 1st diagnosis in 25% patients presenting with a diagnosis of myocarditis. These children had presented with shortness of breath, fever,

poor feeding, hepatomegaly and tachycardia [4]. Hence when a child does not respond to supportive care and respiratory distress worsens with abnormal perfusion a chest radiograph should be done and there after ECHO (if available) to promptly recognize DCM in resource limited setting to reduce morbidity and early transfer to tertiary hospital.

Viral etiology has been implicated as a cause of majority of the cases of cardiomyopathy, which causes myocarditis resulting in DCM [1,5]. Identifying the underlying etiology is a challenge in our set up. In our case we tested for some of the viruses which were negative for the first case and adenovirus was positive in the second case. Yamini et al described parainfluenza was the commonest virus isolated in children with myocarditis [4].

Cardiorespiratory therapies and vasoactive medication such as diuretics, inotropes and vasopressors and vasodilators are used in the management of shock and have shown to improve the outcome as seen from our case. Advanced cardiorespiratory support such as ECMO and Ventricular assisted device (VAD) are not available in our region [6]. However, the outcomes of paediatric cardiogenic shock are highly variable and depend on the etiology, extent and nature of myocardial inflammation, treatment given and overall reversibility of the affected myocardium [3]. Both our infants showed improvement in a few days of starting the medical management as evidenced by clinical improvement and Echocardiography.

CONCLUSION

There are no diagnostic and management protocols of DCM in the sub-Saharan set up and it is hoped this case report can be a start for further research. There is need for more sensitization on DCM, myocarditis and cardiogenic shock in Kenya. It being misdiagnosed or diagnosed late is associated with high morbidity and mortality. More research is needed to classify the etiology in sub-Saharan region as the causes may be different from those in other regions.

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