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Case Report

Left Ventricular Non-Compaction Cardiomyopathy Co-Existing with HIV Infection in a Nigerian Child

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Abstract

Background: Left ventricular non compaction cardiomyopathy is uncommon disease in children, its co-existing with HIV infection in children has not been reported in the Nigeria.

Introduction

Left ventricular non-compaction cardiomyopathy (LVNC), also known as non-compaction of the ventricular myocardium (NCVM) is a relatively rare congenital condition and represents an arrest in the normal process of myocardial compaction, resulting in persistence of multiple prominent ventricular trabeculations and deep intertrabecular recesses [1]. When compaction does not occur during intrauterine life, spongy network with deep intratrabecular recesses persist. The reason for non-compaction is not known but believed to be due to pressure overload and myocardial ischaemia [2]. LVNC can occur in isolation or coexist with other cardiac and/or systemic anomalies.

World Health Organization (WHO) and European Society of Cardiology classified LVNC as unclassified cardiomyopathy because of morphological trait shared by many phenotypically distinct cardiomyopathies, while American Heart Association in recognition of the rapid evolution of molecular genetics in cardiology classified it as a primary, genetic cardiomyopathy [3]. LVNC is a genetically heterogeneous disorder, with a sporadic and familial form [4]. Autosomal dominant inheritance appear commoner than X-linked, though autosomal recessive inheritance has also observed, and familial recurrence varies between 18 and 50% [3]. Several genes that cause LVNC have been identified. These genes appear to encode sarcomeric (contractile apparatus) or cytoskeleton proteins, and disturbance of the NOTCH signalling pathway seems part of a final common pathway for this form of disease. Disrupted mitochondrial function and metabolic abnormalities have a causal role too. LVNC is linked to mutations in G4.5 gene, α-dystrobrevin, ZASP genes encoding for the Z-band of the Z-line in skeletal and cardiac muscle, chromosome 1, 5, and 11 though the specific genes has not been identified, mitochondrial, and sarcomeric proteins [5,6]. Non-compaction of the left ventricular myocardium lies within the diverse spectrum of cardiac morphologies triggered by sarcomere protein gene defects and shares molecular etiology of different cardiomyopathic phenotypes. Patients with isolated LVNC, hypertrophic, and dilated cardiomyopathy share common mutations in sarcomere protein genes.

Though HIV infection has been reported in association with LVNC in adult patients, it has not been noted to be the cause of the disease, [6] but rather a known cause of dilated cardiomyopathy which could be from HIV itself, or from drug myotoxicity, or autoimmune process or from secondary infections. There has been controversy over LVNC being acquired. This was from serial echocardiographic studies where LVNC was not diagnosed on initial echocardiogram but was becoming evident in subsequent examinations [7]. Patients with dilated cardiomyopathy, hypertrophic cardiomyopathy, and LVNC share common mutations in sarcomere protein genes. This common genetic background raises the hypothesis whether LVNC can also develop later in life, though developmental changes strongly supports abnormality in the early myocardial morphogenesis or failure of full maturation of the compacted myocardium [8].

LVNC has a wide range of clinical presentations: from asymptomatic patients, to patients who develop ventricular arrhythmias, systemic thromboembolism phenomenon, heart failure, and sudden cardiac death.9 The disease can be associated with LV dilation, LV hypertrophy, systolic or diastolic dysfunction, or both, or with other congenital heart disease (CHD), or other systemic metabolic diseases, mitochondrial, cytoskeleton anomalies or occur in isolation [9]. The first reported case was an autopsy finding in a newborn infant with aortic atresia/coronary-ventricular fistula in 1932 [10]. Three case reports on HIV infected adult patients with cortical blindness, progressive multifocal leucencephalopathy, heart failure, and neurologic dysfunction has been reported [11-13]. LVNC has also been described in a Nigerian male adult with facial dysmorphism [14]. Literature search did not reveal any report of an HIV infected child with LVNC. We report a case of LVNC in a girl with HIV whose initial manifestation was an acute stroke for baseline documentation.

Case Presentation

B.F. is a 10 year old girl, double orphan from HIV infection, and referred from a private health facility. She was first diagnosed of HIV infection at the age of 2 years and has been on highly active antiretroviral therapy (HAART), lamivudine, zidovudine and nevirapine for the past 8 years. She presented to our health facility with a 4 day history of facial asymmetry with deviation of the mouth to the left, a right hemiplegia and expressive aphasia. Prior to onset of above symptoms, she had complained of generalized body weakness and had gone to bed only to suddenly wake up at night with a shout

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followed by aphasic and other presenting symptoms. There was no history of headaches, neck stiffness, diplopia, and photophobia, loss of consciousness or trauma to the head. There was also, no history of easy fatigability, body swellings, orthopnoea or paroxysmal nocturnal dyspnoea. She had a past medical history of recurrent ear discharge and one previous hospital admission but no blood transfusions or surgeries. She is the last of the three children of her late parents who died from complications of HIV. She lives with her paternal aunt in a state different from where she accesses treatment for HIV. She is a primary 2 pupil because of several missed school days from follow up clinic appointments and ill health. Siblings are alive and not HIV infected.

At presentation, she was afebrile with a temperature of 36.60C, but later became febrile with a temperature of 38.80C, conscious but restless with left facial nerve palsy, no signs of meningeal irritation, right hemiplegia, decreased tone, and reflexes and 0/5 power on the right upper and lower. Tone, power and reflexes were normal in the left upper and lower limbs. Pain, touch and temperature sensations were all intact in all the limbs. She had a pulse rate of 88 beats/min, blood pressure of 100/50 mmHg, and normal precordium. Her apex beat was displaced at the 5th LICS anterior axillary line with no thrill or heave, and heart sounds 1 & 11 with no murmurs. B.F. was dyspnoeic and tacchypnoeic with RR of 44 breaths /min, flaring alaenasi and intercostal /subcostal recessions. Breath sounds were vesicular with no added sound. She had bilateral, creamy, non-foul smelling ear discharge with an intact tympanic membrane. Her digestive and urogenital system examinations were essentially normal.

CT scan done showed a hypodense, non-enhancing lesion in the left basal ganglia and mild compression of the ipsilateral horn of the lateral ventricle suggesting an acute intracerebral infarct from an ischemic stroke. CXR showed a globular heart with pulmonary congestion and plethora suggesting heart failure and pulmonary oedema. 2D echocardiography (ECG) findings showed: No coexisting of cardiac abnormalities, two–layered myocardial structure with a compacted thin epicardial band and a thicker non-compacted endocardial, trabecular meshwork with deep endomyocardial spaces measuring a maximal NC/C layer ratio > 2, non-compaction predominantly in the mid-lateral, mid-inferior, and apical segments and located mainly towards the apex, colour doppler evidence of perfused intratrabecular recesses. ECG also showed severe left ventricular dysfunction and borderline right ventricular function. There were however, no intracardiac masses or vegetation seen. MP was positive (+). Other results were normal: CD4 count (1,192 cells/ul), HBsAg, HCV antibodies, blood culture, S/E/U/Cr, CBC, urinalysis and lipid profile. PT was 14 seconds (Control 11 secs), INR was 1.5 (Normal 2 – 3).

A diagnosis of cardiovascular accident (CVA) secondary to thromboembolism from LVNC in an HIV–infected patient in WHO clinical stage 2 was made. While still being investigated, the patient had a repeat stroke the day after admission. She was subsequently commenced on warfarin, anti-failure medications (spironolactone, enalapril, digoxin and frusemide), antimalarials, ceftriaxone and physiotherapy. Her ARVs were continued. She was discharged after 17 days of hospitalization. At discharge, she was no longer aphasic and the power in her right upper and lower limbs had improved to 2/5. Her INR is being monitored weekly while being followed up in the cardiology; neurology, physiotherapy, and special treatment clinic for HIV infected or affected children.

Discussion

LVNC is a condition where papillary muscles of the heart are not well-developed; the non-compacted internal myocardial layers consist of more than 50% of the ventricular wall thickness, with some microcirculation disorder. The reported prevalence of LVNC varies considerably and the true prevalence is not known. 9.2% was reported from Australian children with childhood cardiomyopathies, [15] 11.3% from Turkey, [16] and 0.014 to 2%from other studies [2,9,10,17]. There has been no reported case of this disease in children in this country. The non-reporting could be its rare nature in this part of the world or as a result of non-availability of ECG machine/ pediatric cardiologists in many centers in the country.

Diagnosis is made by ECG studies with the first diagnostic criteria described by Jenni et al. in 2001 [18]. Diagnostic criteria by Jenni et al include: At least four distinct trabeculation and intratrabecular recesses, primary involvement of the apical, the mid-inferior and the mid-lateral regions, direct contact of the recesses with the ventricular cavity as seen by color Doppler ECG, and the ratio of the broad non-compacted structure to thin compact structure of >2 on parasternal short-axis image [18]. Our patient met all of the above mentioned criteria in addition to absence of any other heart abnormality in the ECG findings. LVNC can occur in isolation as in this reported case or in association with other CHD. If LVNC does not coexist with other cardiovascular pathologies it is called an 'isolated' LVNC. A new echocardiographic technique, such as tissue Doppler imaging, strain and strain rate, and speckle tracking, may help in the evaluation of functional impact of an abnormal myocardium for the clinician to distinguish between normal trabeculated myocardium from LVNC. Left ventricular twist was determined by speckle tracking ECG [19]. Rotation was clockwise at the base and counterclockwise at the apex in all controls and patients with dilated cardiomyopathy, while the LV base and apex rotated in the same direction in all non-compaction patients. Thus the rotation/twist of the LV may be a new more objective diagnostic criterion for evaluation of non-compaction [19] from other forms cardiomyopathies.

Our patient had remained asymptomatic of LVNC disease, her first presentation being at her 10th year of his life when she suddenly developed facial asymmetry, right hemiplegia and expressive aphasia with no prior history of any other illness or trauma. This was attributed to thromboembolism from LVNC when no other obvious causes after detailed clinical evaluation and laboratory investigations. Thromboembolism is not a common clinical manifestation of LVNC in children as reported by several other authors [16,20,21]. Ali et al [22] however reported only 2 cases in the study. Reason (s) for uncommon presentation of thromboembolic phenomena in children with LCNV as compared to adult is not clear. Late presentation has also been reported in other studies as in this case report

Limitations

We could not perform a cerebral angiogram to further determine the cause of the stroke.

Conclusion

To our knowledge, this is the first reported case of LVNC in HIV –infected pediatric patient whose initial manifestation was a stroke. This report highlight the need for routine screening of children especially those with HIV infection for heart diseases, as the initial manifestation can be quite detrimental, if not fatal.

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