Cryptogenic stroke due to Left Ventricular Non-compaction

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ABSTRACT:
Left ventricular non-compaction (LVNC) is a rare form of cardiomyopathy which may be asymptomatic or present as congestive cardiac failure (CCF), arrhythmia or cardio-embolic stroke. This condition is increasingly being reported with improvement in cardiac imaging techniques. We present 2 children with LVNC who presented with CCF and stroke.

Case 1:
A 9-year girl was admitted with headache, fever, vomiting and chest pain for 3 days and facial deviation & weakness of left upper and lower limb for 4 hours. (Acute onset of left hemiplegia). This was preceded by a period of excessive exertion. An episode of congestive cardiac failure at 9 months had been attributed to hypertension of undiagnosed etiology and she was lost to follow up but was reportedly well and remained asymptomatic and off medication until this current episode. On admission, her heart rate was 110 and her BP was 110/50. Her left radial and right ulnar pulses were not palpable. She was aphasic and had a left sided hemiparesis. The differential diagnosis at this time was vasculitis or cardiomyopathy with embolic phenomena. Magnetic resonance Imaging showed a large infarct in right fronto-parietal region. On Magnetic Resonance Angiography there was non-visualisation of right Internal Carotid Artery (ICA), left cervical, vertebral & narrowing of left cavernous ICA. Her Anti Nuclear antibody, ESR and C Reactive Protein were within normal limits. Her ECHO was suggestive of dilated cardiomyopathy. ECHO repeated on an updated machine with sonicated albumin contrast showed trabeculations and deep recesses in the myocardium diagnostic of left ventricular non-compaction. She was started on Carvedilol and Warfarin. Her hemiparesis gradually improved. However there was no improvement in cardiac function. She had episodes of transient weakness while in hospital, all associated with excessive exertion. This brought in the consideration of a possible hemodynamic stroke. Currently she is on warfarin and has had no more strokes and has no cardiac failure.

Figure 1: MRI Brain in 9 year old girl with LVNC and stroke showing right middle cerebral artery territory infarct

Figure 2: CT angiography of 9 year old girl with stroke showing bilateral carotid block with collateral and intracranial vessel reformation.

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Case 2:
A 3 year old boy was admitted with easy fatiguability and breathlessness at rest since 4 days. His mother had noticed that he had palpitation and hyperactive precordium since the past month. He had a history of repair of anorectal malformation. On admission he was hypertensive (BP 170/100) and had features of congestive cardiac failure (bilateral basal crepitations, hyperactive precordium with gallop rhythm and hepatomegaly).

He did not have any focal neurological deficit. His 2D echo showed non compaction of left ventricular myocardium and left ventricular ejection fraction 10-15%. He was treated for hypertensive emergency and cardiac failure (sodium nitroprusside infusion, furosemide, amlodipine and atenolol). Work up for hypertension revealed that he had left sided dysplastic kidney with left renal artery stenosis.

On the 8th hospital day, he had sudden onset right sided hemiparesis with facial involvement. He was normotensive at this time. His MRI brain showed multiple infarcts in left posterior central gyrus, left occipital cortex and left posterior frontal lobe and right parietal deep matter. He was started on low molecular weight heparin. His hemiparesis gradually improved. He reamins on long term anti coagulation.

Left Ventricular Non-compaction (LVNC) is a form of cardiomyopathy with prominent trabeculations, deep endomyocardial recesses and and bilayered myocardium with spongy and compact layers. The embryo has spongy myocardium with deep recesses communicating with the ventricular cavity. Between 12 to 18 weeks of gestation, remodeling occurs and the myocardium becomes more compact, with vascularization and disappearance of the recesses. Arrest in this process of compaction is thought to lead to LVNC although LVNC can occur at any age and recent human and animal studies have challenged this theory.

It may occur in isolation or in association with congenital heart disease or neuromuscular disease. There is genetic heterogeneity. X linked, autosomal dominant and mitochondrial inheritance have been described although autosomal dominant is the most common. Mutations in sarcomere protein genes have been identified. 20% of cases may be familial It is important to screen all first-degree relatives. The major clinical manifestations are heart failure, arrhythmias, embolic events such as stroke and sudden death (3,4). Embolic complications occur in 21 to 38% of cases (5-8) and are related to development of thrombi in the extensively trabeculated ventricle, depressed systolic function or atrial fibrillation. Diastolic dysfunction occurs due to abnormal relaxation and restrictive filling by the numerous prominent trabeculae, and systolic dysfunction due to subendocardial hypoperfusion.

The differential diagnosis includes apical form of hypertrophic cardiomyopathy, hypertensive cardiomyopathy, and endocardial fibroelastosis. ECG changes vary from LVH, repolarization changes, inverted T, ST changes, axis shifts, to intra-ventricular and atrio-ventricular conduction abnormalities. Diagnosis is made on the basis of specific morphologic criteria on ECHO such as thickened myocardium wall with ratio of non compacted to compacted myocardium >2 on short axis view, and deep inter trabecular recesses communicating with the ventricular cavity and multiple trabeculations in end systole (8-10). Cardiac MRI and newer modalities such as 3D ECHO, speckle tracking and tissue Doppler have improved diagnostic yield. There is also the possibility that the increased trabeculation seen in athletes and certain racial groups may be misdiagnosed as LVNC.

Management depends on the ejection fraction and clinical manifestations and may include carvedilol, implantable cardioverter-defibrillator, heart transplant and long term anticoagulation. Implantable cardioverter-defibrillator is indicated in patients with syncope and severely impaired systolic function.

Studies indicate a high incidence of heart failure, arrhythmias and sudden death (7, 9,11). 155 children with LVNC were studied and 5 year outcome was analyzed according to their cardiomyopathy phenotype (12). 44 children (28%) died or needed cardiac transplantation, with worse outcome with the dilated (45%), indeterminate (42%) and hypertrophic (25%) phenotype compared to isolated LVNC phenotype (6%, p<0.035).

Current literature suggests that LVNC may be a phenotypic expression of differing underlying diseases(7). Management is currently based on the clinical presentation and severity of cardiac dysfunction. Our patients improved with anti-
coagulation and treatment of congestive cardiac failure.

Fig 3: Left ventricle of the human embryo at different stages showing normal trabecular compaction. (Adapted from Varnava AM, Heart. 2001; 86:599-600)

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