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RESEARCH ARTICLE

A CASE OF LEFT VENTRICULAR NON COMPACTION CARDIOMYOPATHY WITH ATRIAL SEPTAL DEFECT

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Abstract

Left ventricular non-compaction (LVNC) is a rare form of cardiomyopathy characterized by abnormal myocardial trabeculations and deep recesses within the left ventricle. It is often associated with other congenital heart defects, such as atrial septal defects (ASD). This report discusses the case of a 5-year-old female with both LVNC and ASD, highlighting the clinical presentation, diagnostic challenges, and management strategies. The case emphasizes the importance of early detection and a multidisciplinary approach to optimize patient outcomes. Recent literature is reviewed to provide updated insights into the pathophysiology, diagnosis, and treatment of this rare condition.

Introduction:-

Case report:

-We illustrate a clinical presentation and imaging of a 5 year old girl who underwent a routine echocardiography after her family physician found a cardiac murmur. It showed prominent left ventricular trabeculae and deep intratrabecular recesses communicating with the ventricular cavity. It also showed a 15 mm ostium secundum atrial septal defect (ASD) causing a left to right shunt across it and relatively enlarged right heart cavities with no significant elevation of pulmonary arterial pressures. ASD percutaneous closure was indicated after the transesophageal echocardiography showed sufficient rims. Further evaluation by a cardiac MRI was indicated and confirmed the LVNC cardiomyopathy diagnosis

Discussion:-

LVNC is recognized by the American Heart Association (AHA) as a primary cardiomyopathy with a genetic origin [1]. However, the European Society of Cardiology (ESC) categorizes LVNC differently, classifying it as an "unclassified" cardiomyopathy [2]. Moreover, the recent ESC guidelines on the management of cardiomyopathies take a distinctive stance by not labeling LVNC as a cardiomyopathy per se. Instead, they describe it as a phenotypic trait that can appear either in isolation or in association with other conditions, including left ventricular hypertrophy, dilatation, systolic dysfunction, or developmental anomalies [3].

The incidence of LVNC in pediatric populations has been estimated at 0.12 per 100,000 children under ten years old and 0.81 per 100,000 infants under one year old [4]. Its increased recognition in pediatric cardiology is largely due to advancements in imaging techniques, which have enhanced the identification of LVNC's distinct morphological characteristics. LVNC can present as an isolated condition or, as in the current case, in conjunction with other congenital heart defects such as atrial septal defect (ASD) [5].

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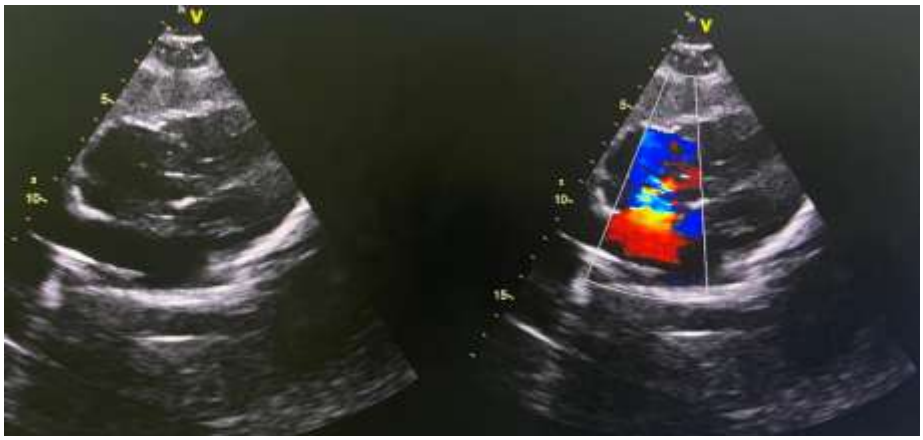
The prevailing theory regarding LVNC's development is the "myocardial non-compaction" hypothesis. During the fourth week of gestation, before the epicardium and coronary artery system have fully developed, the cardiac cells rely on a large exchange surface for nutrient and oxygen diffusion from the blood across the endocardium [6]. This is facilitated by the formation of a network of myocardial trabeculae and deep intertrabecular recesses, which increase the surface area available for exchange [7]. The myocardium is then differentiated into two layers: a compact subepicardial layer and a thicker, non-compacted layer [8]. As the coronary circulation develops and nourishment transitions from passive diffusion to active circulation, compaction of the myocardium begins, progressing from the base to the apex and from the epicardium to the endocardium, thereby reducing the non-compaction to compaction (NC/C) ratio [9].

Contrary to earlier theories suggesting that the compact layer emerges from the compaction of pre-existing trabeculations, more recent observations indicate continuous growth in both the trabecular and compact myocardial layers. This suggests that the hypertrabecular phenotype associated with LVNC arises from the thickening of the non-compacted layer rather than a failure in the compaction process. Consequently, it is now recommended to refer to this condition as left ventricular hypertrabeculation rather than non-compaction [10].

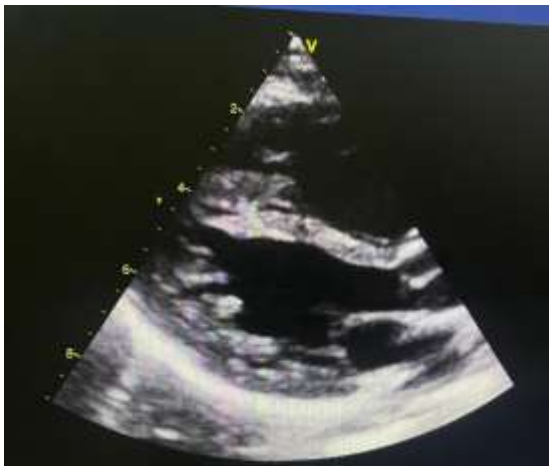
-CHD also arises during the heart development, studies have been conducted to see if there might be possible associations between CHD and LVNC.

-recent studies have found that ventricular septal defects VSD was the CHD with the greater prevalence of LVNC and that the association with ASD is a rare occurrence.(11)
-they also showed that the association of LVNC and CHD increased the risk of congestive heart failure, arrhythmia and surgical risk.(11)
-The presence of both LVNC and ASD in a pediatric patient can significantly impact the clinical presentation and course of the disease. Patients with this combination often present with symptoms such as exercise intolerance, recurrent respiratory infections, and in severe cases, signs of heart failure. The ASD may initially be well tolerated, but as the child grows and the right ventricle's ability to handle increased preload symptoms may become more pronounced (12).
-From a diagnostic standpoint, the presence of an ASD in a patient with LVNC requires careful evaluation. While echocardiography remains the first-line imaging modality, the assessment of LVNC can be challenging due to the overlap with normal trabeculation patterns, especially in children. Cardiac MRI offers superior resolution and can provide a more definitive assessment of myocardial non-compaction, as well as detailed anatomical information about the ASD (13).

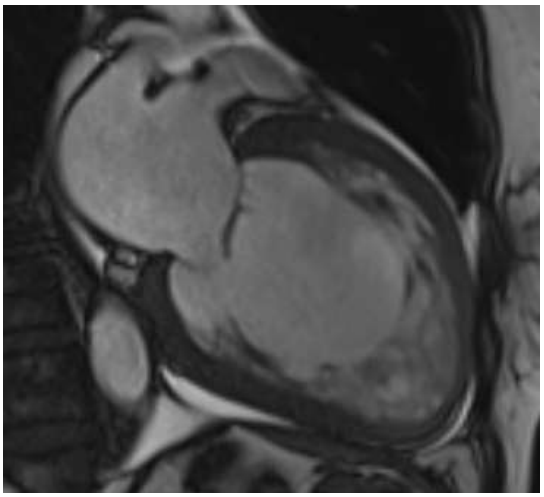
-Management of patients with both LVNC and ASD requires a multidisciplinary approach. The primary goals are to optimize left ventricular function, prevent complications such as arrhythmias and thromboembolism, and address the hemodynamic burden imposed by the ASD. Medical therapy, including beta-blockers and ACE inhibitors, is commonly used to manage LVNC, while the decision to percutaneously or surgically close the ASD depends on factors such as the size of the defect, the presence of symptoms, and the degree of right ventricular volume overload (14).



Subcostal view showing the OS septal defect with a left to right shunting



PSLA view showing the prominently trabeculated left ventricle and the enlarged pulmonary infundibulum, an indicative of right heart chambers enlargement



Long axis 2 cavities cine cardiac MRI sequence showing a double layered myocardium, the inner non compacted and outer compacted with aa non compacted to compacted myocardium ratio greater than 2.3 rendering the diagnosis of LVNC.

Conclusion:-

-The association between LVNC and ASD in pediatric patients presents unique challenges in diagnosis, management, and prognosis. Early recognition of this association is critical to prevent complications and optimize outcomes. A multidisciplinary approach that includes medical therapy, surgical intervention, and ongoing surveillance is essential for managing these complex patients. Continued research into the genetic and molecular mechanisms underlying LVNC and ASD may provide further insights into their pathogenesis and lead to more targeted therapies in the future.

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