

Left Ventricular Non-compaction Associated With Patent Ductus Arteriosus

M. Esmailzadeh, Moshkani M. Farahani and A.A. Usefi
Department of Echocardiography and General Cardiology, Shahid Rajaei Cardiovascular
Medical center, Iran University of Medical Sciences, Tehran

Abstract: Isolated Left Ventricular Non-Compaction (LVNC) is a rare congenital anomaly which is characterized by numerous prominent ventricular trabeculations and deep intertrabecular recesses. The incidence of associated cardiovascular complications and congenital heart disease is high. It is associated with congestive heart failure, ventricular arrhythmia and embolic events. It is believed to represent an arrest in endomyocardial morphogenesis. We report a 23-year-old female with Patent Ductus Arteriosus (PDA) and incidental finding of PDA and LV non-compaction. As far as we know this is the second report of LV non-compaction associated with PDA.

Key words: Left ventricular non-compaction, patent ductus arteriosus

INTRODUCTION

Noncompaction of the ventricular myocardium also known as left ventricular non compaction (LVNC) is a rare congenital cardiomyopathy and represents an arrest in the process of myocardial compaction, resulting in persistence of numerous prominent ventricular trabeculations and deep intertrabecular recesses^[1]. The disorder has recently been recognized as a distinct form of cardiomyopathy. It was previously termed spongy myocardium, although this term has been abandoned because it underscores the hypothesis that the basic morphogenetic abnormality may be arrest of normal compact of the loose interwoven mesh of myocardial fibers in the embryo^[2,3]. It typically involves the left ventricle; although involvement of the right ventricle has been reported^[4]. Clinical presentations include depressed systolic and diastolic function, systemic embolism, and the development of ventricular arrhythmias^[5]. The most common cause of death in these patients is sudden cardiac collapse. Many patients suffer from ischemic attacks, pulmonary embolism, heart failure, pulmonary edema episodes, cardiogenic shock and sustained ventricular tachycardia^[6-12]. We report a 23-year-old female with incidental finding of patent ductus arteriosus (PDA) and left ventricular noncompaction. As far as we know this is the second report of left ventricular non-compaction associated with PDA.

CASE REPORT

A 23-year-old female presented with nausea, vomiting and loss of consciousness. Her vital signs were as

follows: BP = 100/60 mmHg, PR = 88/min RR = 24/min, T = 37.9. A continuous murmur was heard in cardiac examination. Laboratory tests showed rising of blood urea nitrogen and creatinine and she needed hemodialysis. FANA and ANCA were negative, Anti dsDNA = 50 and blood cultures were negative. Transthoracic and transesophageal echocardiography showed a large (25x30 mm) mobile mass on pulmonic valve with a highly mobile mass in right pulmonary artery originating from patent ductus arteriosus (Fig. 1).

There was continuous shunt flow from descending aorta into the pulmonary artery which suggested the presence of PDA (Fig. 2).

Left ventricular ejection fraction was mildly reduced (LVEF = 45%) and there was multiple prominent trabeculations with deep intertrabecular recesses in posterolateral LV wall with a ratio of noncompacted/compacted myocardium about 2.1/1 (Fig. 3).

Renal sonography showed increased kidney size and renal biopsy revealed class IV lupus nephritis.

The patient developed severe pulmonary insufficiency on the follow up echocardiographic examination at two weeks without any change in the size of pulmonary valve and artery masses. Because of high grade lupus nephritis surgery is postponed after clinical and medical stabilization.

DISCUSSION

Noncompaction of the ventricular myocardium is an embryonic cardiomyopathy that is increasingly being

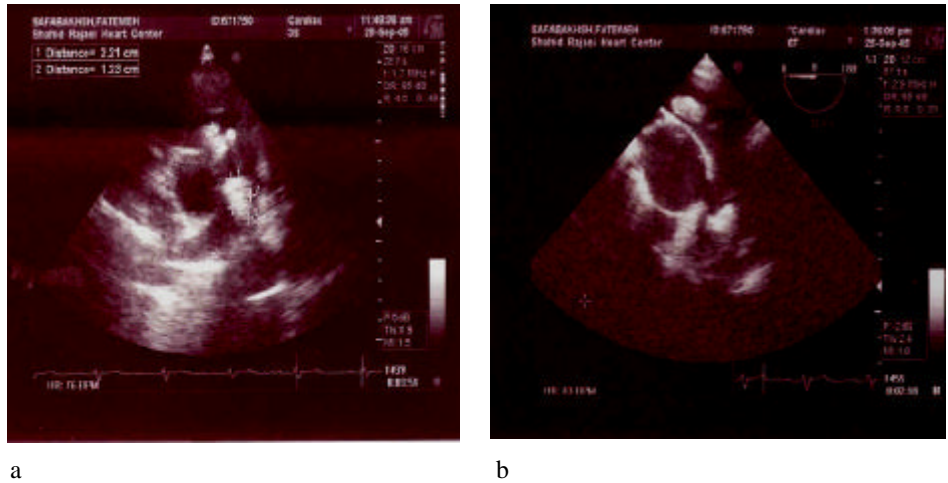


Fig. 1: Transthoracic (a) and transesophageal (b) views showed large vegetations in pulmonary valve and in right pulmonary artery

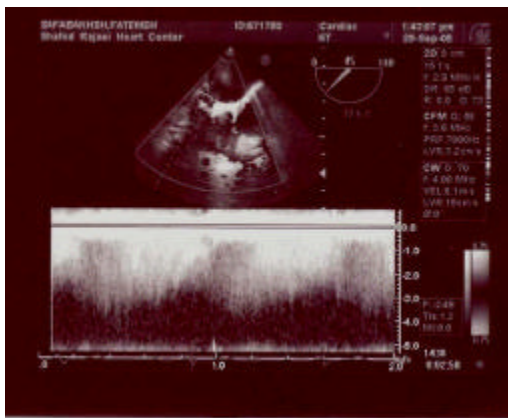


Fig. 2: Transesophageal view in 45° upper esophageal showed large PDA with continuous shunt flow

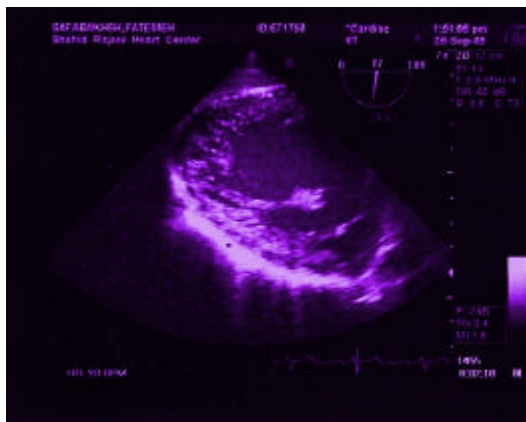


Fig. 3: Transgastric transesophageal view showed multiple prominent trabeculations with deep intertrabecular recesses in posterior apical region

recognized^[13]. LVNC has been categorized as an unclassified cardiomyopathy by the World Health Organization in the recently published report on definition and classification of cardiomyopathies^[1]. It is a result of an arrest in myocardial morphogenesis and is characterized by excessively prominent trabeculations in a ventricular wall segments and deep intertrabecular recesses.

Three types of clinical presentations have been described in patients with LVNC: 1) Systolic and diastolic dysfunction 2) Ventricular arrhythmias and 3) Systemic embolization^[14]. The cause of left ventricular dysfunction is not clear. Epicardial coronary arteries were intact in almost all of reported cases, but intramural hypoperfusion was suspected^[15]. The diagnosis is made by echocardiography, but magnetic resonance imaging (MRI) is increasingly used and provided better delineation of the extent of the abnormal trabeculations in patients with noncompaction of the left myocardium. It was particularly useful when the myocardial involvement was subtle^[16]. Review of literature showed only one report of LVNC associated with PDA^[15]. Our case had some unique feature such as PDA and LVNC with large vegetations on pulmonic valve, pulmonary trunk and ductus arteriosus and high grade lupus nephritis.

REFERENCES

1. Richardson, P., W. McKenna and M. Bristow, *et al.*, 1996. Report of the 1995 World Health Organization/International Society and Federation of Cardiology Task Force on the definition and classification of cardiomyopathies. *Circulation*, 93: 841-842.

2. Dusek, J., B. Ostadal and M. Duskova, 1975. Postnatal persistence of spongy myocardium with embryonic blood supply. *Arch Pathol.*, 99: 312-317.
3. Jenni, N. R., Goebel and R. Tartini, *et al.*, 1986. Persisting myocardial sinusoids of both ventricles as an isolated anomaly : Echocardiographic, angiographic and pathologic anatomical findings. *Cardiovasc Intervent Radiol.*, 9: 127-131.
4. Towbin, J.A. and S.E. Lipshultz, 1999. Genetics of neonatal cardiomyopathy. *Curr. Opin. Cardiol.*, 14: 250-262.
5. Neudorf, U.E., A. Hussein and E. Trowitzsch, *et al.*, 2001. Clinical features of isolated noncompaction of the myocardium in children. *Cardiol. Young*, 11: 439-442.
6. Robida, A. and H.A. Hajar, 1996. Ventricular conduction defect in isolated Noncompaction of the ventricular myocardium. *Pediatr. Cardiol.*, 17: 189-191.
7. Hutchins, G.D., M. Schwaiger and K.C. Rosenspire, 1990. Non-invasive quantification of regional myocardial blood flow in the human heart using N-13 ammonia and dynamic positron emission tomography imaging. *J. Am. Coll. Cardiol.*, 15: 1032-42.
8. Yoshbayashi, M., N. Tamaki and K. Nishioka, 1992. Ischemic myocardial injury evaluated using positron emission tomography in children with coronary artery disease: Comparison with thallium-201 SPECT. *J. Cardiol.*, 22: 21-26.
9. Bleyl, S.B., B.R. Mumford and M.C. Brown-Harrison, 1997. Xq28-linked noncompaction of the left ventricular myocardium: prenatal diagnosis and pathologic analysis of affected individuals. *Am. J. Med. Genet.*, 72: 257-265.
10. Stoberger, C., J. Finsterer and K. Blazek, 1996. Left ventricular noncompaction in a patient with Becker muscular dystrophy. *Heart*, 78: 380.
11. Engberding, R. and F. Bender, 1984. Identification of a rare congenital anomaly of the myocardium by 2-dimensional echocardiography: Persistence of isolated myocardial sinusoids. *Am. J. Cardiol.*, 53: 1733-1734.
12. Conces, D.J., T. Ryan and R.D. Traver, 1991. Noncompaction of ventricular myocardium: CT appearance. *Am. J. Roentgenol.* 156: 717-718.
13. Khalid, S., A. Omran, H. Najm and M. Godman, 2004. Noncompaction of the ventricular myocardium associated with mitral regurgitation and preserved ventricular systolic function. *Am. Soc. of Echocardiogr.*, 17: 87-90.
14. Oechslin, E.N., C.H. Attenhofer Jost, J.R. Rojas, P.A. Kaufmann and R. Jenni, 2000. Long term follow up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. *Am. Coll. Cardiol.*, 36: 493-500.
15. Hsiao, S.H., T.Y. Lee, G.Y. Mar and N.J. Peng, *et al.*, 2004. Left ventricular noncompaction associated with patent ductus arteriosus. *Acta Clinical Sin.*, 20: 251-255.
16. Alhabshan, F., J.F. Smallhorn, F. Golding, N. Musewe, R.M. Freedom and S.J. Yoo, 1998. Extent of myocardial noncompaction: comparison between MRI and echocardiographic evaluation. *Pediatric. Radiology*, pp: 1432.