Left Ventricular Noncompaction

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Left ventricular noncompaction (LVNC) is a rare form of cardiomyopathy of emerging importance. We report a case of a 3- months- old boy who presented with congestive heart failure due to LVNC.

Key words: *Cardiomyopathy, Noncompaction, Congestive heart failure.*

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Left ventricular noncompaction (LVNC) is a reportedly rare disorder of endomyocardial morphogenesis [1]. The prevalence of LVNC among patients referred to echocardiography laboratories is reported to be between 0.014 and 1.3% [2,3] and accounts for 9.5% of all primary cardiomyopathies [4]. It is commonly misdiagnosed as hypertrophic or dilated cardiomyopathy. There is only one case report of LVNC in children from India [5].

CASE REPORT

A 3-months-old boy presented with feeding difficulty and sweating over forehead since 15 days and breathlessness since 2 days. His parents were unrelated and he was their first child. On examination, he had congestive heart failure (heart rate -100 /min, weak pulses, capillary refill time of 5 seconds, S3 gallop, grade 3/6 pansystolic murmur at apex, hepatomegaly, tachypnea and bilateral basal crepitations). Other systemic examination was normal. He had no dysmorphic features.

The *X*-ray chest showed cardiomegaly. The ECG revealed left ventricular hypertrophy. The echocardiography demonstrated grossly enlarged left ventricle. A two layer structure was seen, with a compacted thin epicardial band and a thick noncompacted endocardial layer with coarse trabeculations having deep intertrabecular recesses at the apical and lateral wall segments of the left ventricle (***Fig.* 1**). The end systolic ratio of noncompacted to compacted layers was 3.2. The color Doppler imaging showed blood filling the recesses from the ventricular cavity. The origin of both the coronary arteries was normal. The M-mode showed left ventricular end- diastolic diameter (LVEDD) – 3.72 cm (Z score = 7.52), left ventricular end- systolic diameter (LVESD) – 3.39 cm (Z score = 12.12), the left ventricle ejection fraction (LVEF) -30.2% and shortening fraction (SF) - 9%. There was mitral regurgitation with velocity of 279 cm/s. The right ventricle was normal. These findings were consistent with isolated left ventricular noncompaction.

He was treated with decongestive therapy and inotropes for 3 days, and subsequently discharged on oral enalapril, frusemide and ecosprin. The echocardiography of parents was normal.

DISCUSSION

Left ventricular non-compaction represents an arrest in endomyocardial morphogenesis, which normally occurs

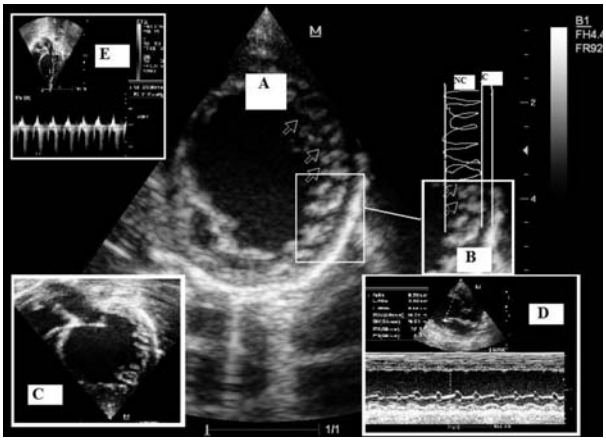

FIG.1 (*A*) *Parasternal short axis view. The end- systolic still frame shows the two layered structure with thin epicardial layer and an extremely thickened endocardial layer with prominent trabeculations and deep recesses (arrows). (B) To quantify the thickness of the noncompacted layer at the site of maximum thickness, the end- systolic ratio of the noncompacted (NC) to compacted (C) layer is determined. (C) Apical four-chamber view still frame. (D) M-Mode study demonstrating LVEF- 30%. (E) mitral valve Doppler demonstrating regurgitation.*

between 5- 8 weeks of fetal life and is characterised by gradual compaction of myocardium, transformation of large intertrabecular spaces into capillaries, and evolution of the coronary circulation [1]. Clinically, patients may be asymptomatic or present with CHF, arrhythmia and embolic events [4-6]. The presenting symptoms include tachypnea, cyanosis, syncope, or failure to thrive, Chin, *et al*. [7] have described facial dysmorphism, including a prominent forehead, low set ears, strabismus, high- arched palate and micrognathia.

LVNC can be diagnosed by two-dimensional echocardiography and color Doppler [7,8]. The criterion for diagnosis is ratio of noncompacted to compacted layer of >2 measured at end systole.

There is no specific therapy. The mainstay of treatment is diuretics, ACE inhibitors and beta- blockers (Carvedilol) to improve the left ventricular systolic function. Anticoagulation (target INR – 2.0-3.0) is recommended when LVEF <40% [3]. Pignatelli, *et al*.[4] recommend oral aspirin and a metabolic cocktail including thiamine, coenzyme Q10, riboflavin and carnitine. Poor prognostic markers predicting death or heart transplant are age at onset, NYHA functional class III–IV, sustained ventricular arrhythmias, ratio of noncompacted to compacted layers, number of affected segments, LVEDD and abnormal lateral mitral tissue Doppler Ea velocity [3,9].

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REFERENCES

1. Elshershari H, Okutan V, Celiker A. Isolated noncompaction of the ventricular myocardium. Cardiol Young. 2001;11:472-5.
2. Oechslin EN, Attenhofer Jost CH, Rojas JR, Kaufmann PA, Jenni R. Long- term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. J Am Coll Cardiol. 2000;36:493-500.
3. Oechslin E, Jenni R. Left ventricular non-compaction revisited: a distinct phenotype with genetic heterogeneity? Eur Heart J. [online]. Available from: URL: http://www.eurheartj.oxfordjournals.org/content/early/2011/01/31/eurheartj.ehq508.full. Accessed February 9, 2011
4. Pignatelli RH, McMahon CJ, Dreyer WJ, Denfield SW, Price J, *et al*. Clinical characterization of left ventricular noncompaction in children: A relatively common form of cardiomyopathy. Circulation. 2003;108:2672-78.
5. Suvarna JC, Deshmukh CT, Hajela SA. Left ventricular noncompaction: a cardiomyopathy often mistaken. Indian J Med Sci. 2009;63:303-7.
6. Ichida F, Hamamichi Y, Miyawaki T, Ono Y, Kamiya T, Akagi T, *et al*. Clinical features of isolated noncompaction of the ventricular myocardium: long- term clinical course, hemodynamic properties, and genetic background. J Am Coll Cardiol. 1999; 34:233-40.
7. Chin TK, Perloff JK, Williams RG, Jue K, Mohrmann R. Isolated noncompaction of left ventricular myocardium: A study of eight cases. Circulation. 1990;82:507-13.
8. Jenni R, Oechslin E, Schneider J, Attenhofer Jost C, Kaufmann P. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. Heart. 2001;86:666-71.
9. McMahon CJ, Pignatelli RH, Nagueh SF, Lee VV, Vaughn W, *et al*. Left ventricular noncompaction cardiomyopathy in children: characterisation of clinical status using tissue Doppler- derived indices of left ventricular diastolic relaxation. Heart. 2007;93:676-81.

Angiokeratoma Circumscriptum of the Tongue

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Angiokeratoma circumscriptum is rare cutaneous disorder. It usually presents as multiple,red, blue or black asymptomatic papules on lower extremities. Oral involvement, common in systemic form, is rare in localized forms. We report a case of angiokeratoma circumscriptum of tongue, involving both dorsal and ventral aspects.

Key words: *Angiokeratoma, Tongue*.

Angiokeratoma is the term applied to describe quite distinct clinical conditions that share a clinical presentation with asymptomatic hyperkeratotic cutaneous vascular lesions and a histological combination of superficial dermal vascular ectasia with overlying hyperkeratosis. The following five varieties are generally recognized: (*i*) generalized systemic type angiokeratoma corporis