

HYPERTROPHIC CARDIOMYOPATHY, CORONARY ECTASIA MYOCARDIAL BRIDGING & INTERNAL CARDIOVERTER DEFIBRILLATOR IMPLANTATION

TARIQ ASHRAF, AZAM SHAFQUAT, ASADULLAH KUNDI

SUMMARY

Hypertrophic Cardiomyopathy is one of the causes of sudden cardiac death. Here we present one case with Hypertrophic Cardiomyopathy with ectatic vessel and myocardial bridging who later underwent Internal Cardioverter Defibrillator implantation.

Key words: Hypertrophic Cardiomyopathy, Coronary Ectasia, Myocardial bridging, Internal Cardioverter Defibrillator.

INTRODUCTION

The clinical course of Hypertrophic Cardiomyopathy (HCM) remain stable over long periods of time with 25% of HCM cohort achieving normal longevity¹⁻⁵. A high prevalence of Myocardial bridging & HCM has been reported⁵. Myocardial bridging may be found at multiple sites in HCM⁶. Here we present one case with HCM, ectatic coronary vessels, myocardial bridging & Internal Cardioverter defibrillator (ICD) implantation.

CASE REPORT:

A 50 year old male was evaluated for exertional angina Functional class III with history of sudden death of his brother at age of 45. ECG reveals Sinus rhythm with Right axis deviation & T-wave inversion in V Leads (Fig 1). Echocardiography showed asymmetrically hypertrophied Left ventricle with septal thickness of 33mm and posterior wall thickness of 12mm (Fig 1). The patient underwent coronary angiography which showed Ectatic left main, Left Anterior descending artery (LAD), Left Circumflex (LCx) & Right Coronary Artery (RCA) with myocardial bridging in mid LAD, LCx and RCA. (Fig 2). Left Ventricular systolic function was normal with left ventricular end diastolic pressure (LVEDP) of 26mmHg. The patient was already on optimal dose of b-blockers & aspirin. As there was history of sudden cardiac death in family and maximum left

ventricular wall thickness of 33mm on Echocardiography ICD was implanted for primary prevention of sudden cardiac death.

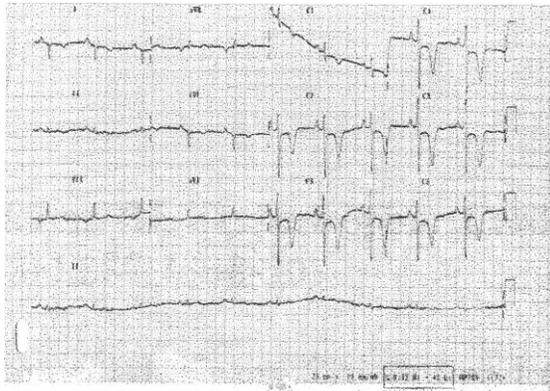
DISCUSSION:

The vascular Ectasia supplying Hypertrophic myocardium suggest that vascular dilatation was related to functional and or structural changes of left ventricular wall. In HCM phasic characteristics of coronary arterial blood flow may be exaggerated to the extent that even systolic flow reversal has been described⁷.

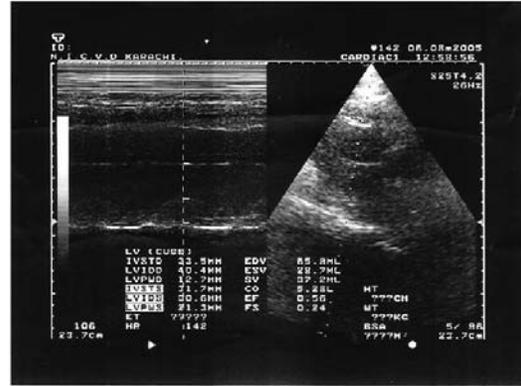
The clinical symptoms of angina in presence of massive coronary ectatic could be due to mismatch of metabolic demands of myocardium and blood supply⁸ compromised by intermittent flow obstruction by myocardial bridges and possible co-existent small vessel disease⁹⁻¹⁰. In literature ectatic vessel with myocardial bridge has been reported in single vessel only¹¹. Our case is unusual in which ectasia with myocardial bridging in HCM were present in three vessels i.e. LAD, LCx & RCA. Moreover there was history of sudden cardiac death in family for which ICD was implanted as primary prevention¹².

* Senior Registrar, Incharge Cath Lab, National Institute of Cardiovascular Disease, Karachi.

Figure - 1
Hypertrophic Cardiomyopathy (a) ECG showing Right Axis Deviation & T-wave inversion in V-leads. (b) Echocardiography showing septal wall thickness of 33mm & posterior wall thickness of 12mm.

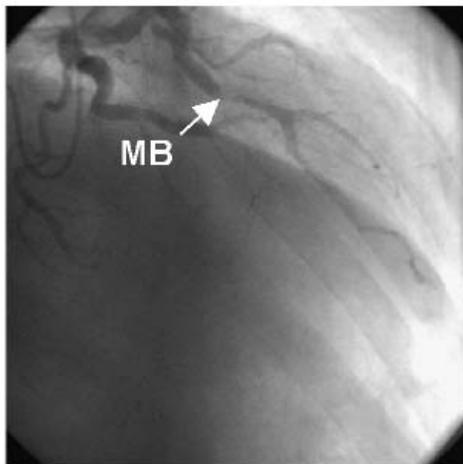


(a)

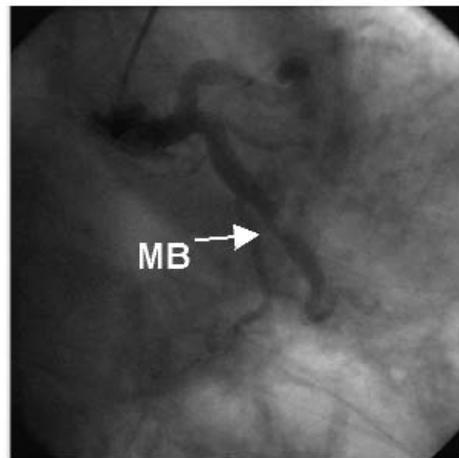


(b)

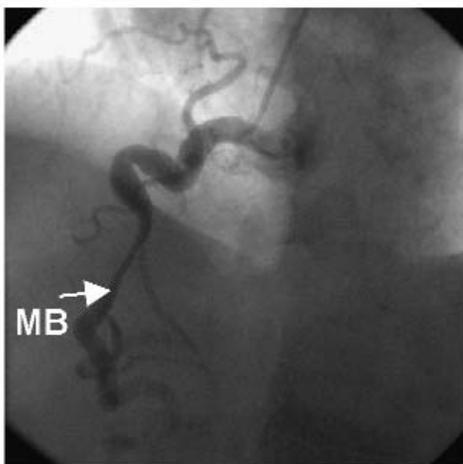
Figure - 2
Hypertrophic Cardiomyopathy, coronary ectasia & myocardial bridging (MB) in mid (a) LAD (b) LCx (c) RCA & (d) Left Ventriculogram.



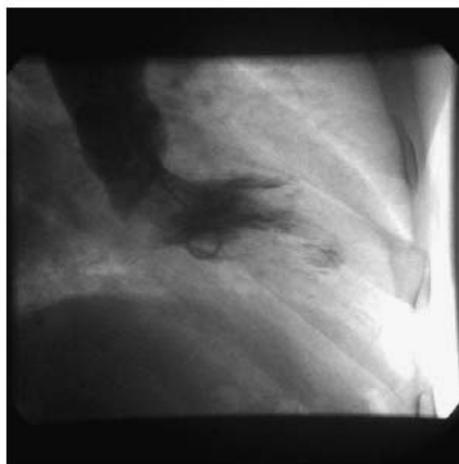
(a)



(b)



(c)



(d)

REFERENCES:

1. Maron BJ. Hypertrophic Cardiomyopathy: a systematic review. *JAMA* 2002;287:1308-20.
2. Maron BJ, Casey SA, Poliac LC, Gohman TE, Almquist AK, Aeppli DM. Clinical Course of Hypertrophic Cardiomyopathy in a regional United States Cohort. *JAMA* 1999;281:650-5.
3. Maron BJ, Casey SA, Hauser RG, Aeppli DM. Clinical Course of Hypertrophic Cardiomyopathy with Survival to advance age. *J Am Coll Cardiol* 2003;42:882-8.
4. Fay WP, Tabiercio CP, IL Strugh DM, Tajik AJ, Gersh BJ. Natural history of Hypertrophic Cardiomyopathy in the elderly. *J Am Coll Cardiol* 1990;16:821-6.
5. Takagi E, Yamakado T, Nakano T. Prognosis of Completely asymptomatic adult patients with Hypertrophic Cardiomyopathy. *J Am Coll Cardiol* 1999;33:206-11.
6. Cannan CR, Reede GS Bailey KR, Melton LJ III, Gersh BJ. Natural history of Hypertrophic Cardiomyopathy. A population based study, 1976 through 1990. *Circulation* 1995;92:2488-99.
7. Akasaka T, Yoshikawa J, Yoshida K, Maede K, Takagi T, Miyake S. Phasic Coronary flow characteristics in patients with Hypertrophic Cardiomyopathy. A study by coronary Doppler catheter. *J Am Soc Echocardiogr.* 1994;7:9-19.
8. Nienaber CA, Gambhir SS, Mody FV, Ratib O, Huang SC, Phelps ME, Schelbert HR. Regional myocardial blood flow and glucose utilization in symptomatic patients with Hypertrophic Cardiomyopathy. *Circulation* 1993;87:1580-1590.
9. Dissmann R, Schultheiss H. Ischemic in patients with Hypertrophic Cardiomyopathy. Various causes & symptoms and the difficulties of ischemic screening tests. *Eur Heart J.* 1996;17:982-984.
10. Maron BJ, Wolfson JK, Epstein SE, Roberts WC. Intramural ("Small Vessel") coronary artery disease in Hypertrophic Cardiomyopathy. *J. Am. Coll. Cardiol* 1986;8:545-557.
11. Stephan Gielen Ruth H. Strasser, Wolfgang Kubler, Christlub Haller Coronary artery ectasia and systolic flow. Cessation in Hypertrophic Cardiomyopathy. *Circulation* 1998;97:2372-2374.
12. Maron BJ, Shen W-K, Linse MS, et al. Efficacy of implantable Cardioverter - defibrillators for the prevention of sudden death in patients with Hypertrophic Cardiomyopathy. *N. Eng. J. Med* 2000; 342:365-73.