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Full Length Research Paper

Endomyocardial Fibrosis: A Summary of Echo-Diagnosed Cases in a Tertiary Hospital in North Central Nigeria

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Endomyocardial fibrosis (EMF) is a form of restrictive cardiomypathy commonly seen in the sub-tropic region, characterized by apical fibrosis of the ventricles at late stage. Echocardiography was the mainstay of diagnosis. A retrospective view of echocardiographic parameters seen in the EMF cases seen at the Jos University Teaching Hospital. Echocardiography records from September 2010 to December 2015 showed ventricular fibrosis (EMF) in 14 of the 5,992, (2.3/1000) adult patients. Their age ranged between 18 and 73 years. Nine (9) patients had apical fibrosis in the right ventricles, while the remaining five (5) had it in the left ventricles. The mean and standard deviation of age of patients were 41.2 ± 16 years, LVIDD 41.3 ± 17.6mm, RVD 46.3 ± 9.7mm and EF ratio 2.1 ± 1.0. Other results were Deceleration time 188.6 ± 60.2msec, and Tricuspid regurgitation velocity (max) 3.0 ± 1.1 m/sec. EMF was seen in this region though prevalence appeared to be declining. Morbidity and mortality can be reduced if early method of diagnosis is defined.

Keywords: Endomyocardial fibrosis, Ventricular apical fibrosis, Echocardiography.

INTRODUCTION

Endomyocardial fibrosis (EMF) is a form restrictive cardiomyopathy commonly seen in the tropical and subtropical regions (Davis, 1948). It is characterized in the late stage by fibrosis of ventricular apical endocardium resulting in a restrictive cardiomyopathy. There may also be destruction of the subvalvular apparatus, though the fibrosis does not usually involve the valve leaflets (Kartha and Gupta, 1993). When the fibrotic process involves the papillary muscles and the subvalvular mechanism, valve immobility results in atrioventricular valve incompetence (Kartha and Gupta, 1993; Okeahialam and Osunkwo, 2002).

Its highest prevalence is in the poor regions of sub-Saharan Africa where it is usually found in children and young adolescents (Davis 1948). Okeahialam et al. 2003 reported a series of cases about 15yrs ago in this centre. Isiguzo et al. 2015 and Danbauchi et al. 2012 and other writers that have reported cases of EMF from this Savanna region, where it was thought to be rare or nonexistent (Okeahialam and Osunkwo, 2002). Generally, the prevalence of this disease is on the decline(Isiguzo et al., 2015). Perhaps, non specific improvement in livelihood and early interventions could be said to have played a vital role in preventing this diseases.

Echocardiography was the main diagnostic tool and supports the management of the disease in most patients as it helps in the assessment of severity and extension of endocardial fibrosis, quality of atrioventrivular valves and presence of intracardiac thrombi (Davis, 1948; Kartha and Gupta, 1993). EMF results in endocardial rigidity, atrioventricular valve incompetence and progressive reduction in ventricular size leading to a restriction in filling. It was first described by Davies and Conners and their coworkers in Uganda (Rutakingirwa et al., 1999).

In this study, we report additional echocardiography

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parameters (tricuspid regurgitation velocity/pressure gradient, site of dominant fibrosis.etc) not initially reported in Jos University Teaching Hospital, considering cases between 2010 and 2015.

MATERIALS AND METHODS

The study was a retrospective analysis of echo database records of the Jos University Teaching Hospital. All patients with echocardiographic diagnosis of EMF (Endomyocardial fibrosis) seen between 2010 and 2015 at the Jos University Teaching Hospital. Jos, Nigeria.

Echocardiogragraphic diagnosis of EMF was included on this study, based on the presence of intra-ventricular apical fibrosis, aneurysmal atrial dilation and significant pulmonary hypertension. A 3.5 MHz transducer probe of an ALOKA Echocardiography machine was used. Demographic data and clinical parameters were extracted from the ECHO Database of the cardiology division. Parameters included: site of dominant fibrosis, left ventricular measurements, and severity of pulmonary hypertension. Ethical approval for the study was obtained from the ethical committee of the Jos University Teaching Hospital

RESULTS

Echocardiography records from September 2010 to December 2015 showed ventricular apical fibrosis (EMF) in 14 of the 5,992 (2.3/1000) adult patients. Their age ranged between 18 and 73 years. Table 1

Nine (9) patients had fibrosis on the right ventricles, while the remaining five (5) had it in the left ventricles. The means and standard deviations of the age of patients were 41.2 $\pm$ 16 years, left ventricular internal diameter at diastole (LVIDD) were 41.3 $\pm$ 17.6mm, right ventricular diameter (RVD) were 46.3 $\pm$ 9.7mm and E/A ratio were 2.1 $\pm$ 1.0. Other results are deceleration time (Dec.t) 188.6 + 60.2ms, and tricuspid valve regurgitation maximum velocity (TRVmax) 3.0 $\pm$ 1.1 m/s using continuous- wave Doppler recording. Severity scoring was not done, but was likely to be moderate to severe because of the chamber sizes. The hepatic vein caliber was not determined (Table 2).

DISCUSSION

Initial reports from Nigeria were of the opinion that EMF occurs in hot and humid coastal areas with most of the cases seen in Ibadan being natives of the southwest (Kartha and Gupta, 1993). Several authors have however, reported cases of EMF among the dry and arid northern Nigeria (Okeahialam and Osunkwo, 2002; Isiguzo et al., 2015; Danbauchi et al., 2012). Jos is a cosmopolitan city with numerous ethnicities peacefully cohabiting. It lies between longitude $8^0 53^l$ and latitude $9^0 56^l$ covering 9,400km^2 in area in a Savanna region.

In general, women of reproductive age and children are more commonly affected than men (Davis 1948). This was corroborated by our finding of 1:2 male-female ratio. Earlier study in this centre showed female preponderance, this could be due to the relatively small sample sizes and variation in mean age, 26.7years in old study compared to 41.7years in this study. The Ibadan series however, showed no sex predilection, while East African study reported 2:1 in favor of males (Kartha and Gupta, 1993).

We had a hospital prevalence of 2.3/1000 compared to initial prevalence of 20% implying a decline in prevalence. Similar late fibrotic stage of EMF was observed in a screening study in rural Mozambique, however 20% of a random sample of 1063 subjects of all age groups had echocardiographic evidence of this disease (Moccumbi et al., 2008). Akinwusi and Oyedemi 2002 reported a decline in prevalence of EMF in South West Nigeria (from 10% in the 1960s and 1970s to 0.02% to 0.04%. Our study was in support of a decline in prevalence of EMF.

The rate of progression of EMF is variable and has been described in hospital-based studies (Akinwusi and Oyedemi 2002; Moccumbi et al., 2008a). Most patients have a rapidly progressing heart failure, leading to death within 2 years of the initial insult (Fernandez et al., 2003). Ventricular dilatation was not a feature seen in our study. Left ventricular systolic function was preserved, much higher with those with left ventricular apical fibrosis. Diastolic function was impaired slightly more in those with right ventricular fibrosis. Doppler studies showed significantly high pulmonary pressure which is not surprising because EMF is associated with severe tricuspid regurgitation. They may also have sudden death from massive pulmonary embolism, systemic embolic events or arrhythmias (Ashok et al., 1982).

Macroscopically, there is cardiomegally due to severe atrial dilation that is related to both impedance of ventricular filling and massive atrioventricular valve regurgitation from tethering of the papillary muscles. All these were observed in most of the cases reviewed. The scar tissue may be massive, engulfing and fusing the trabeculae carnae, obliterating the tricuspid valve apparatus, thus resulting in severely dilated tricuspid annulus and free tricuspid regurgitation (Mocumbi et al., 2008b, Vijayaraghavan and Sivasankaran, 2012).

Moccumbi et al. 2008b had earlier observed that Pulmonary hypertension can be seen in both left and right EMF, depending on the stage of the disease and severity of structural lesions. Vijayaraghavan and Sivasankaran 2012 reported severe pulmonary

Table 1. Demographic characteristics and echocardiographic parameters of subject

Sn	age	sex	Ht(m)	Wt (kg)	Side of fibrosis	TRVmax (m/sec)	PG (mmHg)	LVIDD (mm)	RVD (mm	EF	E/A	Dec t (ms)
1	50	F			R			61	63	26		
2	16	F	1.50	44	L	3.98	20.6	17	40	81	2.9	77
3	18	F	1.68	55	R			50	42	85		
4	40	F			L			17	41	81		
5	35	F	1.67	55	L			24		65		
6	38	F	1.62	62	R			44		56	1.46	187
7	73	F	1.43	60	R			42	48	62	3.60	213
8	41	M	1.78	77	L	3.05	37.5	22	47	85	0.81	160
9	37	F	1.59	55	R	0.62		33	25	71	1.50	130
10	60	M	1.65	70	R	3.82	58.6	62	45	45	1.70	134
11	40	F	1.56	46	R	3.05	37.1	48	48	61	3.90	264
12	49	M		82	R	3.63	52.8	69	57	40	1.40	312
13	21	M	1.56	45	L	3.07	37.6	31	55	82	1.70	228
14	60	M	1.7	77	R	2.90		58	44	36	1.80	181

Keys: ht-height, wt-weight, L-left, R-right, TRVmax- tricuspid valve regurgitation maximum velocity, PG- pressure gradient, LVIDD- left ventricular internal diameter at diastole, RVD- right ventricular diameter, EF- ejection fraction, Dec. t- deceleration time.

Table 2. Echcardiographic parameters (mean values) based on side of ventricular fibrosis

	RIGHT SIDED EMF	LEFT SIDED EMF
LVIDD (mm)	52.4	22.2
RVD (mm)	46.5	45.8
EF (%)	54	78
E/A	2.2	1.8
Dec. t(msec)	203	155

LVIDD- left ventricular internal diameter in diastole, RVD- right ventricular diameter, EF- ejection fraction, Dec.t- deceleration time

hypertension to be more common in left sided EMF and is associated with retrograde increase in pulmonary pressure caused by diastolic dysfunction and mitral valve abnormalities in right EMF, pulmonary hypertension is due to acute or chronic thromboembolism. It was therefore not surprising that most of the EMF patients had pulmonary hypertension. (Table 2)

CONCLUSION

EMF continues to remain a public health concern in African countries. Sporadic cases are still being reported in Savanna regions of the continent, more frequent in the socially disadvantaged area, in women. Echocardiography remains an important tool in diagnosing the disease unfortunately at late stages. There is therefore need for more research aimed at early detection, perhaps the use of modern software is the way forward.

Author's Contributions

This work was carried out in collaboration between all authors. Author A (Chundusu CM) designed the study, and wrote the first draft of the manuscript. Authors B

(Rafeal SC, Gomerep VS) and C (Kumtap YC) managed the literature searches of the study. Author D (Danbauchi SS and Okkeahialam BN) managed the analyses of the study. All authors read and approved the final manuscript.

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