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ENDOMYOCARDIAL FIBROSIS: DECREASING PREVALENCE OR MISSED DIAGNOSES?

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ABSTRACT

Objective: The prevalence of endomyocardial fibrosis is reported to on the decline worldwide. This study was carried out to determine the prevalence of endomyocardial fibrosis and the possible contribution of missed diagnoses, at the University of Nigeria Teaching Hospital, Enugu.

Materials and Methods: The case records of patients admitted into the pediatric and adult medical wards of the University of Nigeria Teaching Hospital, Enugu, between November 1993 and October 1999 inclusive, were reviewed. Echocardigraphy records in the same hospital between January 1991 and October 1991 inclusive, were equally analyzed.

Results: Nine of the total admitted patients and 16 of the echocardiographic records, were diagnosed as having endomyocardial fibrosis. This shown a prevalence of 0.09 % and 0.8% respectively. Four (44.4%) of the admitted endomyocardial fibrosis, and 31.2% of the echo-documented case, were within the first three decades of life. The rate of missed diagnosis was quite high (75%).

Conclusion: Compared with the earlier prevalence rate of 10%for the same environment, is the prevalence of endomyocardial fibrosis decreasing, or would missed diagnosis account for the trend obtainable now ?. More concerted efforts at the provision and use of diagnosis facilities is advocated to unravel the true picture.

KEYS WORDS: Endomyocardial fibrosis, Prevalence, Missed Diagnosis.

INTRODUCTION

Endomyocardial fibrosis (EMF) has been defined as a progressive disease of the heart (of as uncertain cause) characterized by fibrosis of the endomyocardial, affecting in particular the apex and subvalvular regions of the ventricles. It is classified under the Restrictive cardiomyopathies, whose salient feature is the restriction to the diastolic filling of the ventricle. Its nomenclature has evolved from Isolated myocarditis,[1] Endomyocardial Necrosis,[2] Endocardial fibrosis,[3] Davies' disease[4] to finally settle with Endomyocardial fibrosis.[5]

Historically, Konstan and Bedford, who observed a from of heart failure in Africa troops serving in the Middle East, first described this condition. 1 These soldiers , usually aged 20-30 years, and mostly from West Africa, presented in heart failure with gross cardiomegaly. Seventeen of their 40 patients died and had autopsy. Finding at autopsy showed extensive subendocardial fibrosis/ necrosis which clinicopathological features Konstan and Bedford considered as corresponding to Isolated myocarditis. It then fell on Davies and his colleagues to produce shortly there a more complete picture of what they termed Endomyocardial Fibrosis (EMF).[5,6]

Endomyocardial Fibrosis affects predominantly children and young adults in the 2nd and 3rd decades. It is endemic in the hot, humid and low lying areas of the tropical rain forest of Africa (notably Uganda, Nigeria, less so in Sudan, the Congo, Kenya, Tanzania, Zambia, Gabon, Ghana, Ivory Coast, Cameroon), South India, Sri Lanka, Colombia, Venezuela, Brazil, Thailand, Ceylon, and Malaya.[3,7,8] A very occasional case of an individual who had never been to / lived in the tropics had, however, been reported by Bishop et at.

So many possible aetiological postulates of this disease have been made. These include bacterial, viral, 5-0H tryptamine derived from plantain diet,[10] cyanide from garri,[11] abnormal immune reaction,[12] schistosomiasis,[13] toxoplasma gondii,[14] and microfilariasis.[15,16,17,18] While the global prevalence of EBF is as yet undetermined, the rate in Nigeria in the sixties and seventies was put at about 10%[19] This study was thus carried out to determine the prevalence of endomyocardial fibrosis and the possible contribution missed diagnoses in the nineties, at the University of Nigeria Hospital, Enugu.

MATERIALS AND METHODS

A review of the total admissions into the paediatric and adult medical wards of the University of Nigeria Teaching Hospital, Enugu, covering a 6- year period (from November 1993 to October 1999) was carried out. A study of the 6614 cases admitted into the medical wards and 3494 paediatric admissions over this period is reported.

Information was collected from the hospital medical admission and discharge records-using a preform. Data including age, sex, data, (time of the year) and diagnosis of the patients using clinical diagnosis criteria.

Equally, a retrospective study of the 2060 patients scanned in the Echocardigraphy laboratory of the same institution, never a 9 year period between January 1991 and October 1999 was done. Information collated, using a preform, included age, sex, date of scanning, pre-Echo (clinical referral)and Echocardiographicdiagnoses respectively.

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RESULTS

A total of 10,108 admitted patients were studied, with only 9 EMF cases (0.09%) discovered. Seven of the EMF case were seen in the adult medical wards and only 2 in the paediatric wards. There were 5 males and 4 females, with a male to female ration of 1.3.:1 for the adults, and 1:1 for the paediatric patients. (see Table 1).

Only 4 out of the total 9 admitted cases (44.4%) were within the first 3 decades of life. One each were in the 4th ,5th and 8th decades, while 2each were in the 3rd and 7th decades. The mean age was 38.3 years, with the age range from 7 to 74 years. Two of the cases each were admitted in 1993 and 1998. One each was admitted in 1995 and 1997. There was non-admitted in 1994 and 1996, while 3 cases were admitted in 1999. Five of the adult cases (71.4%) were seen in the predominated rainy season months of May to October while 2 were seen between November and April. the 2- paediatric cases were also seen during the rainy season months.

Table 2 shows the Echocardiographic diagnosis of EMF, from January 1991 to October 1999. Sixteen of the 2060 scanned patients (0.8%) were EMF cases. the male to female ration was 3.1. The mean age was 39 years, with the age range from 2 ½ to 60 years. Only 5 out of the 16 cases (31.2%) were within the first 3 decades. The predominant rainy season months accounted for 10 out of the 16 cases(62.5%).

DISCUSSION

From tables 1 and 2 it can be seen that the prevalence of EMF among the admitted cases and the Echo-scanned ones are 0.09% and 0.8% respectively. This definitely contrasts with the about 10% prevalence rate in the country of study (Nigeria), obtained in the sixties and seventies[19].

There is, however, a correlation in prevalence with the work of Farrer-Brown and Tarbit in Kampala, where 0.08%(11) of the 147 hearts obtained at autopsy from patients who died in Mulago Hospital had characteristic appearances of EMF.[20] Falase also recently observed that there are indications that the prevalence of EMF may be much less now.[21] Enugu, where the study was done, is located in the Guinea Savannah geographical belt, up from the location possible influence the prevalence rate recorded in the study? this is based on the earlier works in the same country of reference which noted that patients with EMF rarely came from Ibadan itself (the place of study) or the savannah region to the North and East, but predominantly from the rainforest area[15].

The sex distribution in this from admitted cases (in table 1) and the Echo-diagnosed cases (Table 2) are 1.3:1 (for adult), 1:1 (paediatric) and 3:1 male to female ration, respectively. This compares with the 1:1 ration obtained by D' Arbela et al in Uganda on 56 cases of autopsy[22]. Andy J and co-workers also obtained male to female ration of 3.1:1 in 41 cases from South Western Nigeria, and 1.3:1 in 48 patients from South Eastern Nigeria, in a study that spanned over an 18- year period.[18] The exact significance of the male: female prevalence ration, however, from the Echo diagnosed EMF may be associated with more readiness to get males receive health than the females, for cultural reason.

The age ranges found in the study are point for careful consideration. Only 4 (44.4%) of the 9 cases in the wards and 5 (31. 2%) of the 16 Echo cases were within the first 3 decades of life. The age ranges varied from 7-74 years (from the wards) and 2 ½ to 60 years from Echo diagnosis, with mean ages at 38.3 and 39 years respectively. Andy et al , however had an age range of 6-76 years with mean age of 21.4 in their 89 studied cases.[18] Shaper and others in an autopsy series in Uganda covering a 16 year period (1950-65), on 128 patients, found that more than two- thirds(86) of the EMF subjects were less than 35 years of age.[23] D' Arbela and colleagues in a later work had 37 of the 56 autopsied EMF cases (>63%) from the 1st 3 decades.[22] It has been documented that EMF has been recorded in a 3 year old and in subjects over 60 years of age , so that age may not be a useful factor in the individual diagnostic problem.[24] One of the EMF cases from our study was 2 ½ years old, and anther 74 years of age. Much literature, however, point to the preponderance of EMF mainly in the 2nd and 3rd decades of life.

It may be that most of our patients could have developed the problem much earlier, but due to the social- economic and cultural trends and practices, turn up much later, in the Teaching hospital, after attempting several orthodox and non-conventional remedies.

The monthly distribution pattern shows 7 cases from the ward and more than 62% of the Echo- diagnosed EMF cases as presenting within the predominant rainy season of the year in this geographic belt (viz April to October). This is in keeping with earlier works done in the country, in which initial sickness was noted to usually begin in the rainy season and relapses also tended to occur during this season.[25,26]

Table 2 shows that the clinical diagnosis of EMF tallied with the Echo diagnosis only in 4 out of the 16 cases (25%). This may be a reflection of a low suspicion index of EMF among the doctors generation. It may possibly contribute to the low prevalence rate of the disease, by way of missed diagnosis.

Noted in table 2 too is the preponderance of Right sided EMF as against left sided or even biventricular EMF. Fourteen of the 16 cases were RVEMF (which is 92%) of the total cases. D' Arbela and colleagues reported electro-cardiographic features of EMF with left ventricular hypertrophy in 6, and right ventricular hypertrophy in 34 of their 56 patients studied at autopsy in Uganda.[22] The 3 patients in Okuwobi's study at Lagos presented EMF of the right ventricle.[27] Yet another work in Uganda over a 16 year period by Shaper et al, on 128 autopsy cases of EMF showed occurrence of pure RV lesions in about 10%, pure LV lesion in about 40% and biventricular in 50% of the case.[23] Hutt and co in anther work surmised that although clinical right sided involvement was more frequent, the LV was more often affected at post mortem.[28] Falase had deposed that EMF was most commonly found in RV, followed by biventricular disease and finally the LV.[19] In a recent work, scanning over 18 years of prospective study (1976-1994)on 89 patients with EMF, the gross pathology and histopathology in four of five cases that had post-mortern was typical of biventricular EMF.[18] It appears, however, from all these, that RVEMF appears more predominant clinically but autopsy may reveal more of LV and biventricular involvement.

CONCLUSION

Is EMF really getting rarer in prevalence, or may missed diagnosis influence its prevalence rate? This will need further epidemiological, and possible longitudinal, studies to elucidate. This may be made more plausible with the availability of better facilities, both clinically and in autopsy, especially in the endemic regions of EMF, while lie around the tropics.

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Table 1. **Total Admissions vs Emf Cases.**

WARDS	TOTAL ADMISSIONS	EMF CASES MALE	FEMALE	TOTAL
Medical	6614	4	3	7
Paediatric	3494	1	1	2
Total		5	4	9

Table 2: **Echocardiographic Diagnosis of Emf**

DATE	SEX	AGE	PRE-ECHO DIAGNOSIS	ECHO DIAGNOSIS
12/8/92	M	60	PERICARDIAL EFFUSION	RVEMF
2/12/93	F	25	EMF	RVEMF
12/1/94	M	37	MVD	RVEMF
17/3/94	F	28	MVD	RVEMF
27/7/94	M	26	VSD	RVEMF
30/1/97	M	21/2		LVEMF
10/4/97	M	37	HHD	RVEMF
22/5/97	M	19	CCF 2° DCM	RVEMF
10/7/97	M	44	EMF	RVEMF
2/2/98	F	50	DCM R/O PLEURAL. EFFUSION	RVEMF
16/2/98	M	55	DCM	RVEMF
4/5/98	M	41	CARDIOMYOPATHY	RVEMF
11/5/98	M	60	CVA	RVEMF
5/10/98	M	60	EMF	RVEMF
3/6/99	M	50	DCM	LVEMF
26/8/99	F	45	PLEURAL. EFFUSION R/O EMF	RVEMF

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