Apical Hypertrophic Cardiomyopathy with Giant T Waves: Experience in Pakistan.

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SUMMARY

Idiopathic cardiac hypertrophy limited to the left ventricular apex alongwith giant negative T waves particularly in the chest leads as described initially from Japan exists in Pakistan. Presented are four Pakistani cases with documented Apical Hypertrophic Cardiomyopathy. As this disease exists in Pakistan, physicians should suspect the diagnosis in any symptomatic or asymptomatic patient with deep inversion of T waves in the ECG who has no apparent reason for such changes. Diagnosis may be confirmed non-invasively by 2-D Echocardiography and occasionally cardiac catheterization may be needed to confidently exclude other associated disease processes especially Coronary heart disease or for tissue biopsy confirmation.

INTRODUCTION

Cariomyopathies are a heterogenous group of disorders characterized by heart muscle dysfunction (1, 2). Hypertrophic cardiomyopathy which is the second most common type of cardiomyopathy is now also known to have many clinico-pathological types including the rarest and the most recently recognized variety called "Apical Hypertrophic Cardiomyopathy" described by the Japanese (3). This variety which was thought to be peculiar to the Far East only has been noted to occur sporadically, in the western populations (4, 5, 6). This paper describes the occurence of this relatively rare form of disease in Pakistan.

CASE HISTORIES :

Patient 1—A 47 years old male patient was referred to the NICVD in 1979 with the diagnosis of ischemic heart disease. He had complained of both typical and atypical chest pains for 2 years. He also had been diagnosed as having suffered an anterior sub-endocardial infarction but without any convincing data. He was physically active and lead a normal life otherwise and had no other known medical disease. No significant past history. No known family history of heart disease or sudden death. Physical examination on admission was unremarkable. Patient was obese but comfortable with normal TPR. All system reviews revealed no abnormality. Cardiovascular examination was essentially normal with brisk pulses, no venous congestion, no clinical L.V.H. but a questionable soft S4 gallop. A resting ECG (Fig. 1) showed LVH by voltage criteria and ST depression with giant negative T waves (more than 10 m.m. deep in V leads) especially in chest leads. An X-Ray chest was normal with no cardiomegaly and no pulmonary venous congestion. An M — mode Echocardiogram was done and was thought to be within normal limits. No 2-D Echo was done as it was then not available at the NICVD. Cardiac catheterization was done and showed normal cardiac pressures except for a moderately elevated LVEDP of 20 m.m. Hg. There was no resting or post ectopic L.V. outflow tract gradient. Left ventriculography showed a non-enlarged left ventricle with hypertrophy localized to the L.V.

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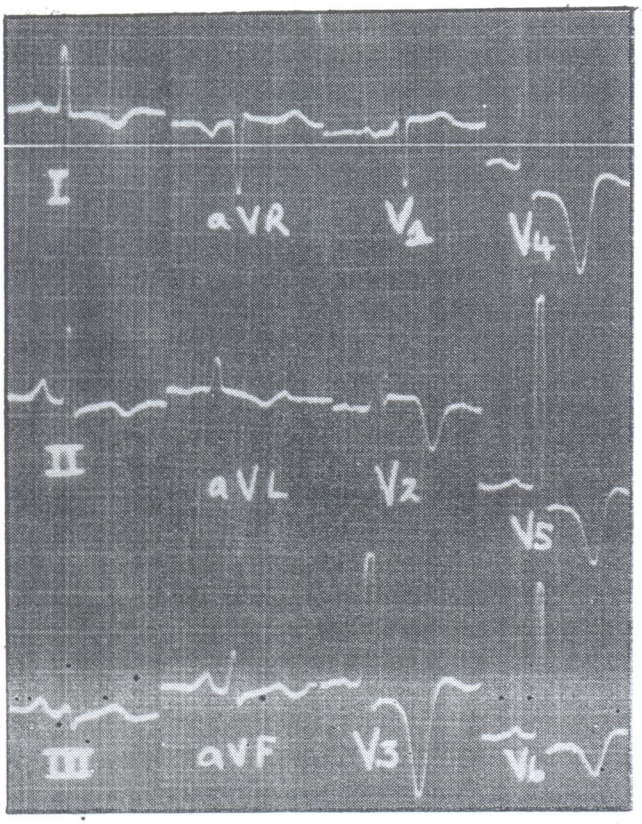

FIGURE – 1.

A typical ECG of a case of Apical Hypertrophic cardiomyopathy showing increased left ventricular voltage and giant negative T waves.

apex and a 'spade -- like' configuration in diastole (Fig. 2) with a vigorous systolic contraction pattern and obliteration of the apical cavity. Coronary Arteriograms (Fig. 3) showed a completely normal coronary arterial tree except for a small area of systolic milking (bridging) on the mid anterior descending artery. The patient was reassured and as he was only mildly symptomatic (intermittently) with a normal effort tolerence on exercise testing, no treatment was given. He continues to do well without treatment.

Patient 2 – This 56 years old teacher and socially very active person presented to the NICVD in 1984 with a history of syncope and jerky movements of the body during unconsciousness occurring thrice over a period of two years. He had been labelled an epileptic after a CT scan was reportedly normal but a borderline EEG abnormality was noted. He was on phenobarbitone 30 mg. twice daily. He had no other medical problems. No history of chest pain or dyspnea etc. No significant past history. No

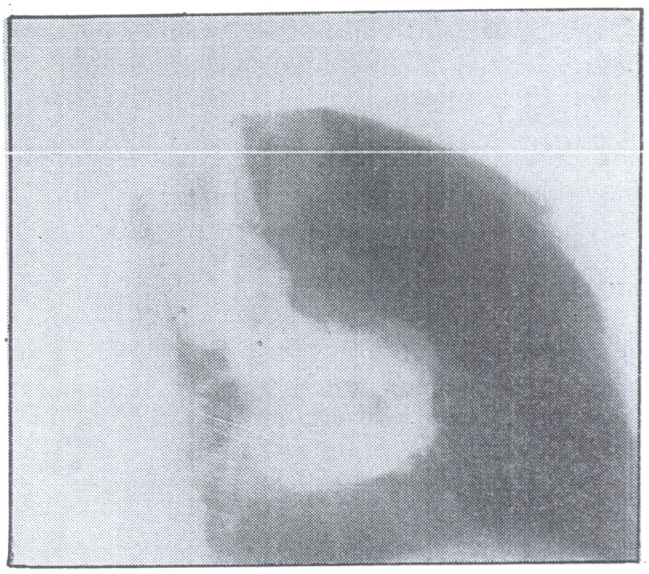

FIGURE -- 2.

30° RAO left ventriculogram in diastole showing the typical 'spade-like' configuration.

family history of known heart disease but a child in the family was diagnosed as having epilepsy. All three episodes of syncope had been sudden complete loss of consciousness without aura, some involuntary bodily movements during syncope but no grand - mal type seizures and complete recovery after several minutes without any post-ictal states. He had not had a recurrence of syncope since being started on phenobarbitone since the last episode a couple of months ago. Physical examination was unremarkable and showed a normally built, alert person looking younger than his stated age and without discomfort. TPR and a systems review was normal. The cardiovascular examination was essentially unremarkable except for suggestion of LVH on apex palpation in the left lateral position and an S4 gallop (remarked at that time as WNL for age). An ECG showed left ventricular hypertrophy and marked ST and T wave changes especially giant negative T inversions across the precordium. An X-Ray chest was normal with no cardiomegaly and no definite pulmonary venous congestion. An ETT was done and showed normal effort tolerence for age and no chest pain or inappropriate dyspnea and no fall in B.P. during or post exercise with persistence of the abnormal ECG pattern throughout the ETT and no arrythmis recorded any time. An M – mode Echocardiogram was initially read as normal but at re-evaluation was thought to show suggestion

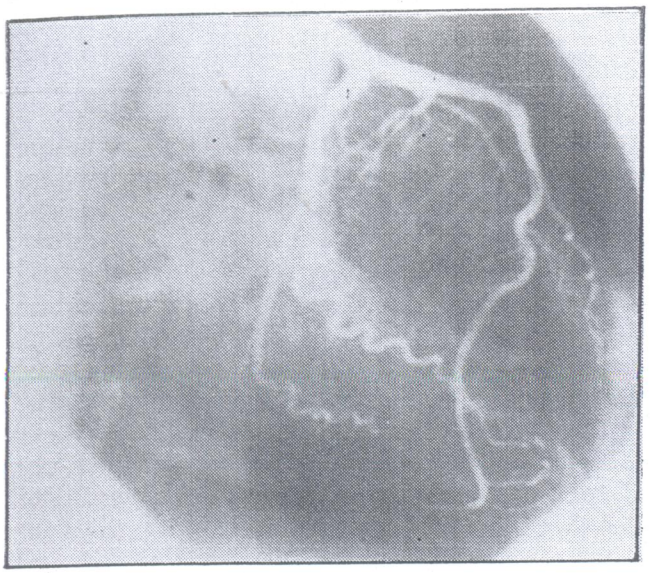

FIGURE – 3.

30° RAO view of the normal left coronary artery.

of abnormal apical thickening and contraction pattern. A 2-D Echo showed the presence of hypertrophy localized to the apex of the left ventricle. Cardiac cateheterization showed normal cardiac pressures except for a severely elevated LVEDP of 32 m.m. Hg. (despite the absence of orthopnea, evidence of pulmonary venous congestion or significant effort dyspnea). There was no resting or post ectopic L.V. outflow gradients. Left ventriculography (Fig. 4) showed same findings as patient No. 1 and coronary angiogrames (Fig. 5) were normal. Patient has been empirically put on verapamil 40 mg TDS and the neurologist, has continued phenobarbitone. He is fully active and free of syncope for last 1½ years. Consultations by the patient in U.K. have resulted in continuation of same treatment.

Patient 3 — This 61 years old male patient started with chest pains in 1977. The pains were mostly atypical, non-exertional, of variable duration and easily bearable. For the past one and a half years he had started experiencing chest heaviness which was mostly, but not always, exertional and somewhat relieved by sublingual nitroglycerine but after 20 minutes or more as a rule. There was also a feeling of dyspnea on exertion alongwith the chest heaviness. There was no history of dizzy or syncopal spells. Patient had been hospitalized in 1977 with a diagnosis of "heart attack" but there were no other admissions or any other major illnesses. He had been

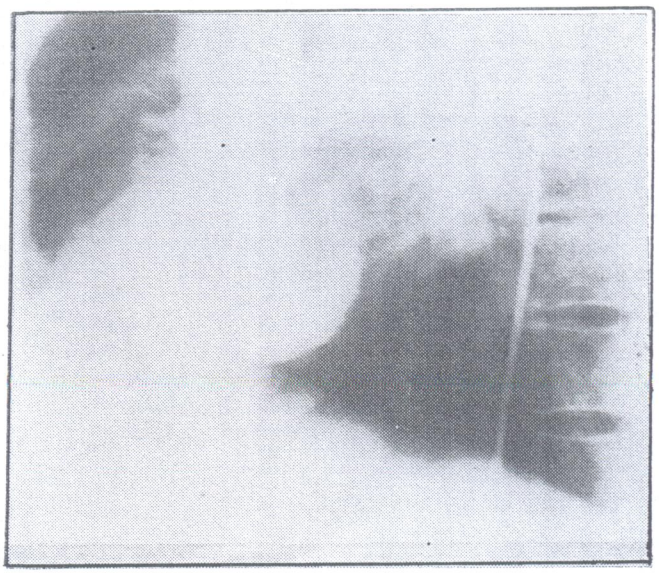

FIGURE – 4.

69° LAO left ventriculogram in diastole showing the localized left ventricular apical hypertrophy and wide outflow tract.

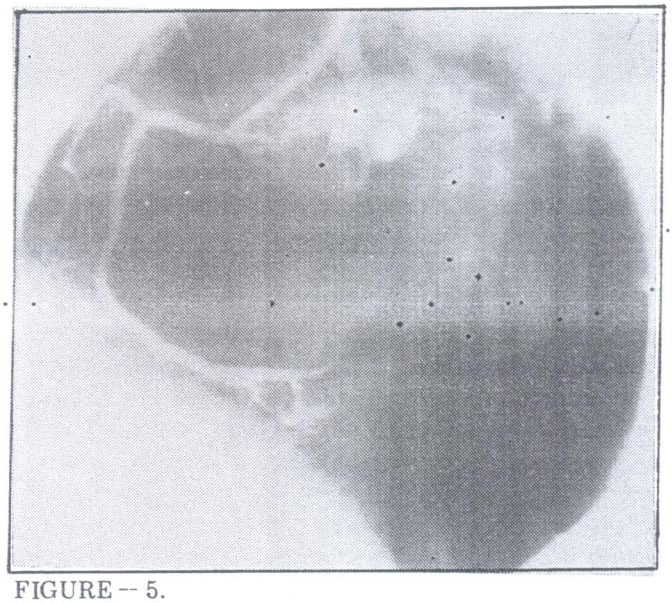

FIGURE – 5.

Right coronary angiogram showing normal coronary vessel.

on various anti-anginal drugs since 1977 and was presently on nitrates, beta blocker and calcium blocker. There was no family history of sudden deaths but one brother had coronary artery disease confirmed by angiography. Pre sently, patient was working and in functional class-II (C.H.A.).

Physical examination revealed an overweight, well built person in good general health. Normotensive, relaxed and with a resting pulse rate

of 60/ minute. Systems review was unremarkable. Cardiovascular examination was essentially normal with no definite left ventricular hypertrophy (thick chest built) but an S4 was audible without any cardiac murmurs. The ECG showed LVH voltage criteria in aVL and anterolateral T inversion with small Q waves (diagnosis of anterolateral infarction in 1977!). 2-D Echo was very difficult to perform but the picture was suggestive of hypertrophy localized to LV and RV apex. There was no ASH or SAM and the upper half of the LV and M — mode Echo appeared normal. Cardiac catehterization showed a very high resting LVEDP = 32 m.m. Hg. All other pressures were normal and there were no valvular of sub-valvular gradients. L.V. angiography revealed a hypercontractile ventricle with systolic obliteration of the apical L.V. cavity and thickening of the apical and papillary muscle region. No mitral or aortic regurgitation was seen. The coronary arteries were completely normal. The patient was continued on a combination of beta-blocker and calcium blocker. A mild diuretic was added and nitrates were slowly withdrawn.

Patient 4 — This 57 years old patient had a history of mild hypertension of recent onset and typical angina of effort. His physical examination was unremarkable but a resting ECG showed left ventricular hypertrophy and marked S T and T wave changes. The finding at cardiac catheterization were unremarkable including a normal LVEDP of 8 m.m. Hg. However, the left ventricular showed severe hypertrophy of the L.V. apex and papillary muscles and a typical spade — like appearance in diastole as in the previous cases. There was no L.V. outflow obstruction. There was, however, some disease in the left anterior descending and circumflex coronary arteries. The left ventricular systolic contraction was supra-normal. The patient is on beta-blockers presently and has responded well symptom — wise. He has two diseases i.e. Apical Hypertrophic Cardiomyopathy and coronary artery disease.

DISCUSSION :

Cardiomyopathies are a group of disorders affecting the heart muscle and of unknown or ill understood etiology and not secondary to any known disease (1). As characterized by John Goodwin (2) and the W.H.O. (1) the two commonest varieties are the Dilated Cardiomyopathy which as the name implies is characterized by dilatation of the ventricles usually without any hypertrophy and a marked diminution in the systolic contractile function resulting in congestive heart failure and ultimately death — *in simple terms systolic function failure.* The second commonest variety is the Hypertrophic Cardiomyopathy characterized by hypertrophy of the left and/or the right ventricles involving part or whole of these chambers. This results in an excellent, supranormal function but the small cavity (with anterior mitral displacement may result in dynamic outflow obstruction in many cases) and thickened and stiffer walls result in difficulty in diastolic stretching and filling and the high diastolic filling pressures in the left heart result in pulmonary congestion and edema and sometimes secondary to this in right heart failure — *in simple terms this is a disease of diastolic function failure.* The two other rare forms of cardiomyopathy are the Restrictive and Obliterative forms (see figure 1).

Our experiences of Dilated and Hypertrophic Cardiomyopathy in Pakistan have been published (7, 8, 9, 10, 11). The cases of Hypertrophic Cardiomyopathy encountered in Pakistan have had all the classical features of the disease as well described in the English language medical literature (1, 2). Hypertrophic Cardiomyopathy has been described by pathologist for years (12) but was first correctly identififed by the British pathologist Donald Teare in 1958 (13) and later placed in its proper perspective amongst the Cardiomyopathies by John Goodwin (2). Since then it has been slowly realized that Hypertrophic Cardiomyopathy is not one disease but a heterogenous group of diseases with varying types of hypertrophy and significantly different clinical features and presentations. The latest sub-group of Hypertrophic Cardiomyopathy described is the so called "Apical Cardiomyopathy" described in the late seventies by the Japanese (3) and thought to be peculiar to the orientals. However reports of isolated cases have been published from the west as well (4, 5, 6) though with some cases exhibiting features different from the classical Japanese description. This paper describes four cases of the classical Japanese variety diagnosed for the first time in Pakistan. Personal communications with cardiologists in India, Bangladesh, Sri Lanka and Nepal have failed to document experience with such cases to date, though this information

is only anecdotal and cases may be diagnosed as awareness of this relatively newly recognized entity spreads.

The remarkable feature of Apical Cardiomyopathy is the relative lack of symptoms and especially absence of significant signs on physical examination contrasting with the severely abnormal ECG showing giant negative T – waves alongwith increased left ventricular voltage and ST depression. While the ECG changes are surely due to apical hypertrophy, the exact etiology remains obscure (14). It is very likely that many of these cases were labelled ischemic heart disease or sub-endocardial infarction in the past. The diagnosis was difficult to confirm as simple M – mode Echocardiography could completely miss the localized apical abnormality (4). However, the availability of 2-D Echo has made the diagnosis a simple matter and can show the extent of the hypertrophy clearly (4), rendering Cardiac Catheterization unnecessary except in complex cases with suspected associated coronary disease or other forms of cardiomyopathy (e.g. Obliterative Cardiomyopathy and apical obliteration) or for research purposes where coronary angiography and ventriculography and endomyocardial biopsies are necessary (14). In our own cases while cardiac catheterization was done (and as expected showed only high diastolic L.V. pressures and no gradients or M.R.), no myocardial biopsies were taken due to lack of facilities to process such specimens. So that while all other features of the disease are exactly the same as described by the Japanese, the presence of myocardial fiber disarray was not established in these cases. However, for clinical purposes neither catheterization nor biopsy is necessary as the clinical and 2-D Echo features of this disease are very specific.

The importance of the recognition of this disease lies in the ease with which it can be confused with other diseases and the fact that treatment with vasodilators and digitalis and diuretics which may be appropriate in cases of coronary heart disease and other cases of left ventricular dysfunction, are inappropriate for Apical Cardiomyopathy. While the best treatment for this new entity is yet to be established, verapamil and mild diuretics (if congestive symptoms are present) may be used. The use of amiodarone would appear to be most appropriate if significant arrythmia are documented. The natural history of this condition is not fully elucidated but may be better than some other forms of Hypertrophic Cardiomyopathy (3). However, in view of the serious nature of this class of diseases (15) these patients should be closely followed until further data is forthcoming.

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