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ASSESSMENT OF CARDIOMYOPATHIES IN AUTOPSY SPECIMEN AT TERTIARY CARE HOSPITAL , BHAVNAGAR

Pathology

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ABSTRACT

Introduction: Cardiomyopathies can cause sudden death. Many of them have histologically detectable changes. The percentage of different type of cardiomyopathies from the region of this study is not available. The purpose of this study was to determine the frequency of cardiomyopathy in autopsy to estimate the frequency of ischemic and non ischemic and to detect primary and secondary cardiomyopathy with respect to age and sex of the deceased. **Materials And Methods:** A retrospective hospital based study of cardiomyopathy was done on autopsy during January 2020 to December 2020. All specimens were photographed and gross description recorded. H&E stained slides were reviewed for presence of morphologically detectable cardiomyopathy **Results:** Out of 209 cases, 19 cases showed cardiac myopathies. Among them, 12 cases were of Ischemic and 07 cases of Non ischemic cardiomyopathy. 04 cases were of primary and 15 cases of secondary cardiomyopathy. All primary cardiomyopathy cases were of hypertrophic cardiomyopathy. Secondary cardiomyopathies included 12 cases of ischemic cardiomyopathy and 3 cases of myocarditis. **Conclusion:** Autopsy study can diagnose undetected/ silent cardiomyopathies that were not known prior to death.

KEYWORDS

Autopsy of heart, Hypertrophic cardiomyopathy, Ischemic Cardiomyopathy, Myocarditis, Restrictive cardiomyopathy.

INTRODUCTION

Cardiomyopathies are of heterogeneous group of disease of the myocardium associated with mechanical and /or electrical dysfunction that usually exhibit inappropriate ventricular hypertrophy or dilation. They occur due to variety of causes that frequently are genetic.[1] They classified as either ischemic and non ischemic and also classified as primary and secondary cardiomyopathy. Cardiomyopathies are clinically unsuspected till a late stage. Cardiomyopathies are also under reported at autopsy [2].Among all three patterns dilated cardiomyopathy is most common clinically and restrictive cardiomyopathy is less frequent. No data is available regarding the percent of different type of cardiomyopathies from the region of this study, the purpose of this study was determine the frequency of cardiomyopathy in autopsy.

MATERIALS AND METHODS

A retrospective study of hearts received for autopsy cardiomyopathy after approval from IRB. All consecutive non autolysed autopsy specimens received between January 2020 to December 2020 were received.

Ischemic cardiomyopathy was considered when either there was history of ischemic heart disease or there was evidence of atherosclerosis in the coronary arteries. Hypertrophic cardiomyopathy was considered when weight of heart was twice the normal along with microscopic evidence of myocyte hypertrophy, replacement fibrosis, thickening of intramural arteries and myocyte disarray. Dilated cardiomyopathy was considered when heart was enlarged, weighing two to three times normal, flabby, globoid shaped and had no features of inflammation, granuloma, Amyloidosis, enlarged, bizarre, hyper chromatic nuclei, multinucleation and hyperchromasia with lipofuscin deposition. Restrictive cardiomyopathy was considered when the right atrial or ventricular wall was severely attenuated due to loss of myocytes, accompanied by massive fatty infiltration and focal fibrosis. Myocarditis was considered when myocardium was infiltrated by inflammatory infiltrate [3]. All specimens were photographed and gross description recorded. Sections were given from the ventricles and atria if available were reviewed for presence of cardiomyopathy

RESULTS AND ANALYSIS

Total 247 autopsies were received during period of January 2020 to December 2020. Among them 212 autopsies contained specimen of heart and 3 of them were autolysed which were excluded from study as per exclusion criteria.

Table 1 Age and etiology wise distribution of cardiomyopathies

Age group (years)	Cardiomyopathies			
	Non ischemic	Ischemic	Primary	Secondary
0-10	00	00	00	00
11-20	00	00	00	00
21-30	01	00	00	01
31-40	02	00	00	02
41-50	01	03	01	03
51-60	02	03	02	03
61-70	01	05	01	05
71-80	00	00	00	00
>80	00	01	00	01
Total	07	12	04	15

Table 1 shows that 19 cases of cardiomyopathies were detected from 209 specimens accounting to 9 % of these ischemic cardiomyopathies were almost double that of non ischemic. Secondary cardiomyopathies markedly outnumbered the primary cardiomyopathies. Non ischemic cardiomyopathy were seen earlier than ischemic.

Table 2 Gender wise distribution of cardiomyopathies

Type of cardiomyopathy	Gender		
	Male	Female	Total
Ischemic	11	01	12
Non ischemic	05	02	07
Primary	03	01	04
Secondary	13	02	15

Secondary cardiomyopathy included ischemic cardiomyopathy and myocarditis. Primary cardiomyopathy composed of hypertrophic cardiomyopathy.

Table 2 shows that the male sex appears to have more number than female sex, the overall cases of female in the 209 specimen was very low. So this difference is likely to be purely due to sample bias.

Table 3 Percentage of histomorphological diagnosis with gender distribution

Histomorphological diagnosis	Gender			
	Male	Female	Total	Percentage%
Acute MI	29	02	31	14%
Ischemic cardiomyopathy (Figure 1)	11	01	12	5.7%
Hypertrophic cardiomyopathy (figure 2,3)	03	01	04	1.9%
Myocarditis (Figure 4A,B)	02	01	03	1.4%

Chronic pericarditis	01	00	01	0.5%
Metastasis	01	00	01	0.5%
No remarkable pathology	115	44	159	76%
Total	161	48	209	100%

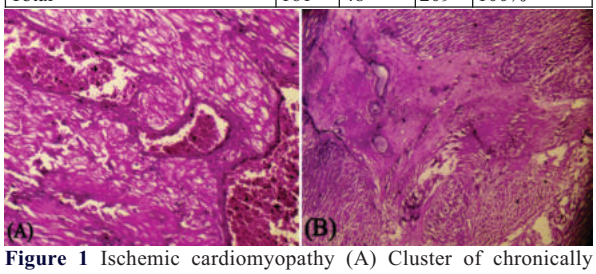

Figure 1 Ischemic cardiomyopathy (A) Cluster of chronically ischemic vacuolated myocytes along with marked congestion of intramural vessels [40x H&E] (B) Sclerosis [10x H&E]

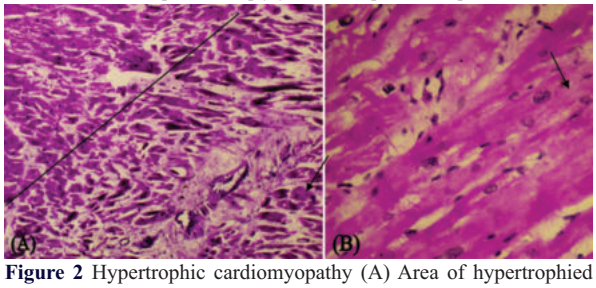

Figure 2 Hypertrophic cardiomyopathy (A) Area of hypertrophied myocardium (arrow) below line and normal myocardium (arrowhead) above line [10x H&E] (B) Hypertrophied myocytes shows mild enlarged, hyper chromatic nuclei (arrow) [40x H&E]

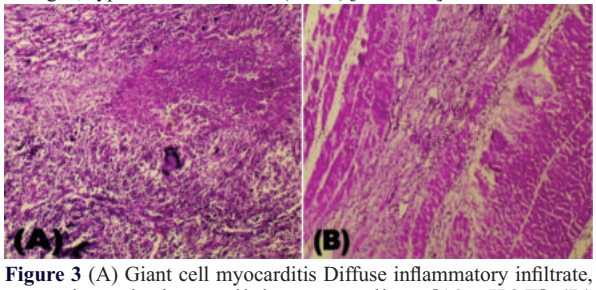

Figure 3 (A) Giant cell myocarditis Diffuse inflammatory infiltrate, necrosis and giant cell in myocardium [10x H&E] (B) Hypersensitivity myocarditis- Patchy interstitial inflammation (arrow) [10x H&E]

DISCUSSION

Cardiomyopathies[4]- Primary Cardiomyopathy predominantly involves heart due to genetic or acquired causes. Secondary cardiomyopathy involves the heart as part of systemic or multiorgan dysfunction. It can also be classified according to anatomical abnormalities fall into three distinct pathological patterns [1] - dilated cardiomyopathy including arrythmogenic cardiomyopathy, restrictive cardiomyopathy and hypertrophic cardiomyopathy. The prevalence of **dilated cardiomyopathy** is 1:2500 with incidence of 7/100,000/year [1]. Its incidence at autopsy is 4.5/100,000/year while clinically its incidence is 2.45/100,000/year [3]. It may manifest clinically at a wide range of age. Prevalence of **hypertrophic cardiomyopathy** is 1:500[1]. It is one of the most common causes of sudden unexpected cardiac death in young people and athletes and is most commonly present in second or third decade of life. Prevalence of restrictive cardiomyopathy is not known.

Inspite of extensive literature search, in autopsy or other histopathology on cardiomyopathy could not be found. Hence hardly any literature is available for comparison of the study. Study *Shah* has mentioned about hypertrophic cardiomyopathy so it has been included for comparison. Few clinical studies - *Hare* and *Tyaminka* have been included as they were found relevant to this topic.

In present study, histological evidence of ischemic cardiomyopathy was observed in age group of 60-70 years. Similar findings were reported by *Shah* [5] though from an earlier age. It is corroborates with the known fact that risk of development of ischemic cardiomyopathy increases with age in patients of heart disease. In present study, 7 cases of non ischemic cardiomyopathy were noted which included 4 cases of hypertrophic cardiomyopathy and 3 cases of myocarditis. Though *Shah* [5] have mentioned the term hypertrophic cardiomyopathy for 20 cases. It is not clear whether they meant hypertensive cardiomyopathy or true hypertrophic cardiomyopathy.

All the cases of hypertrophic cardiomyopathy in this study were of primary cardiomyopathy as only the heart was involved in these cases and no specific etiology was found like history of hypertension or coronary heart disease were found in these cases.

Table No. 3: Comparison of % of cardiomyopathy in different studies

Cardiomyopathy	Hare (6)	Tyaminka (7)	Present study
Ischemic	34%	65%	63.1%
Non Ischemic	66%	35%	36.8%

In present study, 63% cases of ischemic cardiomyopathy were found which is similar to study by *Tyaminka* [7] Concordance was seen with non ischemic cardiomyopathy also

However, *Hare*[6] noted ischemic cardiomyopathy in 34% cases. It is a study of Boston, USA and the area may be having fewer cases of ischemic heart disease and/or post ischemic cardiomyopathies due to availability of better health care. Hence their % of ischemic cardiomyopathy may be less.

In cases of ischemic cardiomyopathy - subendocardial or transmural replacement fibrosis myocyte hypertrophy and vacuolization are seen as a result of chronic ischemic changes were seen.

Hypertrophic cardiomyopathy was found in 1.9% cases. Similar findings were present in studies of 1.3% by *Shah*[5], *1.7% by Garg*[8] ,3.2% by *Sonawane*[9],.In cases of hypertrophic cardiomyopathy mild myocyte hypertrophy, hyper chromatic nuclei, areas of hypertrophy and atrophy of myocytes, intramural artery thickening and fibrosis were seen.

Myocarditis was found in 4 cases in this study. *Joshi C* [10] detected myocarditis in 9% and *Garg*[8] in 3.5% on the autopsies. In present study, we observed two types of myocarditis- one case of giant cell myocarditis and two cases of probable autoimmune/ drug induced or hypersensitivity myocarditis. G**iant cell myocarditis** showed diffuse infiltration of myocardium by lymphocytes, eosinophils, plasma cells and prominent giant cells with areas of necrosis. In 2 cases showed interstitial and perivascular infiltration of abundant eosinophils, lymphocytes, macrophages and plasma cells suggestive of hypersensitivity myocarditis.

2 cases had infiltration in right ventricle. However section of right atria for confirming diagnosis of restrictive cardiomyopathy were not available.

Chronic pericarditis was found in 0.5% case, whereas *Garg* [8] reported 2.8% cases and C. *Joshi*[10] 0.8% cases. Primary pericarditis is uncommon and is mostly secondary to concurrent cardiac infection. It was not analyzed further in this study as it is not part of study.

Limitation Of Study:

In the present study, post-mortem investigation to detect myocardial ischemic change like, histochemical staining by triphenyl tertazolium chloride and special stain like Masson trichome were not done.

CONCLUSION

Autopsy study can diagnose undetected/ silent cardiomyopathies. Ischemic cardiomyopathies were the most common followed by hypertrophic cardiomyopathy and myocarditis.

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