Massive Myocardial Calcification: A Rare Autopsy Finding

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Abstract

Introduction: Myocardial calcification is rare and occur by two mechanisms- dystrophic and metastatic. It can present with variable clinical manifestations like congestive heart failure, cardiomyopathy, arrhythmia and sudden cardiac death.

Case Report: Post mortem visceraof a 50 year old male was received in the department of pathology with alleged history of burns, who died after two weeks of hospital treatment. There was no past history or investigation available in the post mortem papers. Whole heart, pieces of brain, both lungs, liver, spleen and both kidneys were received. On gross examinationno abnormality was observed in any of these viscera. Microscopic examination revealed extensive calcification in anterior, lateral & posterior walls of left ventricle of the heart. Sections from kidney showed features of chronic tubulointerstitial nephritis.

Conclusion: Myocardial calcification is rare and mostly diagnosed incidentally or on autopsy. Extensive sampling of heart and other viscera might help in finding the etiology in such rare cases of massive calcification in myocardium. Antemortem diagnosis can be made with computed tomography (CT) scan and endomyocardial biopsy.

Keywords: Autopsy, dystrophic calcification, myocardial calcification, tubulointerstitial nephritis.

Introduction

Massivecardiac calcification also called as heart of stone, is very rare.[1] Cardiac calcification is characterized by the abnormal accumulation of calcium salts in various parts of heart like coronary arteries, cardiac valves, myocardium and pericardium of heart.[2]

Two basic types of calcification have been recognised namely dystrophic and metastatic. Dystrophic calcification represents the sequelae of local tissue damage and cellular necrosis. It is not associated with abnormalities in serum calcium level showever, hypercalcemia has shown to accentuate the process.[3] Dystrophic calcification is more prevalent than metastatic calcification. The most common etiology is previous myocardial infarction leading to myocyte necrosis.[4]

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Metastatic calcification represents the consequence of a systemic process like hypercalcemia andor abnormalities of calcium homeostasis and can occur in normal or diseased tissue. Any abnormality of calcium metabolism like renal failure, increased bone turnover, hyperparathyroidism and vitamin D-related disorders can lead to metastatic calcification. Metastatic myocardial calcification is most commonly reported in patients with chronic renal failure who are on hemodialysis.[8]

It is also important to emphasise that the etiology of myocardial calcification can often also be multifactorial in origin. For instance, several cases of extensive calcification have been reported in patients after heart transplant due to a combination of repeated cellular rejection, transient renal failure, cardiac trauma, steroid administration and septicemia.[5]

Massive myocardial calcification usually have variableclinical manifestations such as chronic heart failure, restrictive cardiomyopathy, arrhythmia and sudden cardiac death. It is associated with poor prognosis and high mortality.[6]Various imaging techniques like radiographs, CT scanand echocardiography can detect their presence. These imaging technologies are currently used widely for the diagnosis in the field of forensics. The final diagnosis can be confirmed by histologic examination.[7]

Knowledge of these potential etiologies associated with myocardial calcification and their imaging patterns can help to provide a concise and accurate differential diagnosis, however histological examination remains gold standard.[8] We here by present a rare case of cardiac calcification identified on autopsy.

Case Report

Post mortem viscera of a 50 year old male was received in the department of pathology with alleged history of approximately55-60% superficial and deep burns and who died after two weeks of treatment. No significant past history or investigation was available in the post mortem papers. Whole heart, pieces of brain, both lung, liver, spleen and both kidneys were received. No abnormality was identified in the viscera on gross examination.

Microscopic examination revealed extensive areas of myocardial calcification involving the anterior, lateral and posterior walls of left ventricle of heart. Single cell calcification of cardiac myocytes are seen in most of the areas. The sections from left ventricle and inter-ventricular septum also showed areas of myocardial fibrosis along with foci of calcification. Sections from coronary arteries showed the presence of pathological intimal thickening in the right coronary and left circumflex arteries. Left anterior descending artery also shows presence of atheromatous plaque with calcification obstructing upto 70 % of the lumen. Von kossa stain was used to further confirm the presence of calcification as the calcified regions were identified by black staining on the calcified areas on the slide.

The sections from kidney show prominent foci of interstitial fibrosis with interspersed areas of tubular atrophy and admixed areas of mononuclear leucocytes. These histomorphological features were consistent with chronic tubulointerstitial nephritis. Sections from piece of both lungs, liver and spleen showed areas of congestion.

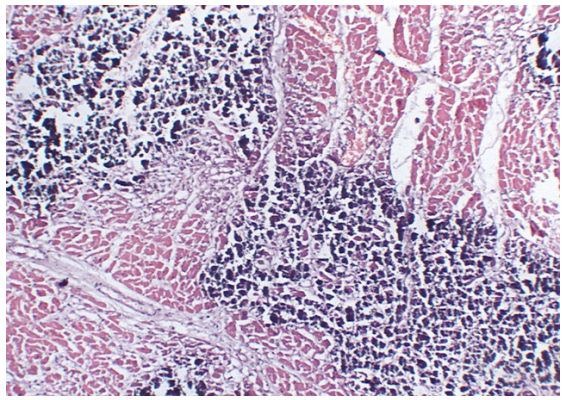

Figure 1: Microphotograph shows calcification in mvocardium(H&E x10).

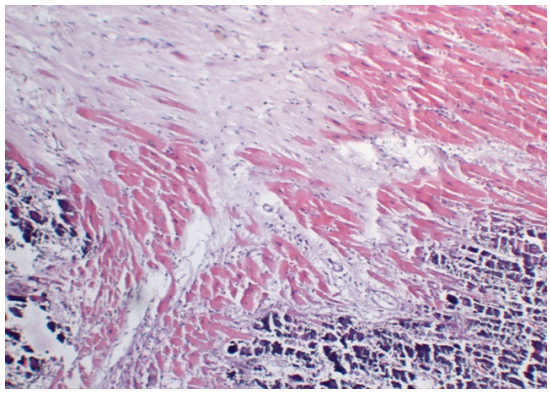

Figure 2: Microphotograph shows calcification in mvocardium with fibrosis (H&E x10).

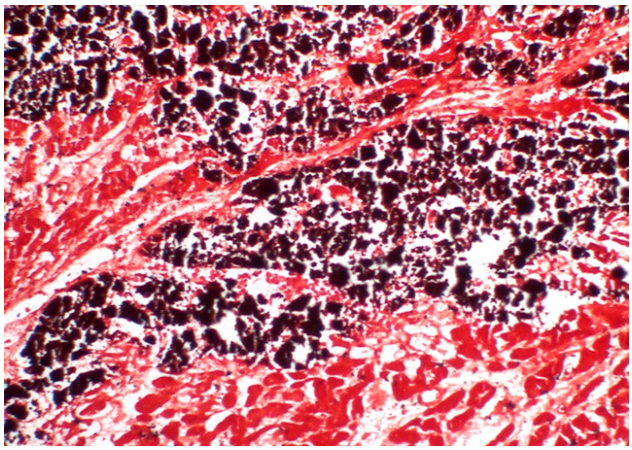

Figure 3: Microphotograph shows positive staining for calcium (Von Kossa x20).

Discussion

Cardiac calcification is not uncommon, however massive myocardial calcification with single cell infiltration is a rare entity usually detected incidentally at time of autopsy with limited published cases in the literature. [9,10] Although a certain degree of cardiac calcification is considered normal as a part of aging, it should be differentiated from pathological process as the latter can cause significant structural abnormalities and functional impairment.

Joo-Young Na[11] reported a case cardiac calcification on autopsy in a patient having medical history of chronic ischemic heart disease and endstage renal disease. The author observed cardiac myocyte dystrophy along with some myocytes showing partially calcified cytoplasm emphasising that calcification in this case was not from the interstitium such as the vascular walls. Thus the author considered that the isolated diffuse massive calcification of the heart was due to both dystrophic and metastatic calcification in this case.

In a retrospective study by Catellier[12] et al, four cases of cardiac calcification were reported on post mortem in which although healed myocardial infarctions were seen but calcitic deposits were not present within these scars in any case. It was observed that the distinction between dystrophic andmetastatic processes was difficult when the calcium deposition was observed in minimally damaged or apparently undamaged myocardium. It was suggested that calcium once present in heartis responsible for successive myocardial injury. Because of differences in tissue substrate, clinical diagnoses, metabolism and age complicate the chemistry of calcific deposition in the myocardium, it further suggested that use of either dystrophic or metastatic be avoided and these lesions may simply be described asmyocardial calcification.

In a prospective study by JoyLiBS[13] et al, they reported three cases of cardiac calcification diagnosed on CT scan with 5 - 13 weeks history of onset of calcifications in all three patients. It was likely that a relatively acute process such as sepsis was causing myocardial damage causing to the deposition of dystrophic calcifications. Histopathological examination of both these calcifications have been described to have identical findings of hydroxyapatite formation and both develop via a shared mechanism in which calcium enters myocardial fibers, initially becoming sequestered by the mitochondria but eventually accumulates throughout the muscle fibers.[14] It was concluded in the study that the calcifications were consistent with dystrophic calcifications as a plausible source of damage to the myocardium was identified. However, given that all three patients had a history of chronic kidney disease with calcium and phosphorus disturbances, there is likely that a component of metastatic calcification was also present in these patients.[13]

Shackley[15]et al reported a case of cardiac calcification in a patient with past history of coarctation of the aorta and pericardial effusions on thoracic CT scan which showed extensive myocardial calcification involving the interventricular septum, apex, anterior and posterior walls of the left ventricle. Calcifications were not identified in any other organ system and the patient's serum calcium levels were repeatedly in the normal range. Histologic examination revealed marked hypertrophy and massive calcification of the myocardium with focal fibrosis related to the calcified foci. Author stated that it was not possible to determine the cause of myocardial calcification in this case.

In another antemortem study by Aras[16] et al, calcification was suspected on X ray and confirmed on CT scan. However, since anatomicopathological study was not performed, etiology of the extensive

myocardial calcification in this patient could not be identified.

In the present study, there was no history or investigations available in the post mortem papers. Gross examination of the viscera revealed no abnormality, however the microscopic examination revealed presence of fibrosis along with coronary artery atherosclerosis and tubulointerstitial nephritis. Therefore, dystrophic calcification of the myocardium was possible. Further histopathology also revealed presence of calcification in individual myocytes rather than diffuse areas of calcification in foci of fibrosis which is better correlating to metastatic calcification which could possibly be due to hypercalcemia following renal failure. So, the possibility of either type of pathological calcification cannot be ruled out with the limited clinical details. This manuscript describes our experience of cardiac calcification which can act as a guide for physicians encountering this condition, as well as stimulate further research into this rare, although a very fascinating diagnosis.

Conclusion

Myocardial calcification is rare and mostly diagnosed incidentally on autopsy. Post mortem analyses and case reports provide valuable insights of this condition helping to shedlight on its rarity, potential predisposing factors and help to provide a concise and accurate differential diagnosis. Extensive sampling of heart and other viscera might help in finding the etiology in such rare cases of massive calcification in myocardium.Antemortem diagnosis can be made possible with use of CT scan and endomyocardial biopsy.

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Ethical Clearance: Not required

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