Case Report

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Calcified Amorphous Tumour in an Eight Year Old Child

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Abstract

Non-neoplastic tumors are rarely found in the heart. Calcified amorphous tumor [CAT] of the heart is one such tumor. When present, it is often mistaken for a calcified myxoma, thrombus, vegetation or malignancy. It is found subendocardially with areas of calcification in an amorphous degenerating fibrin background causing obstruction to the outflow tracts. It has to be excised and subjected for histopathological examination to confirm its diagnosis. We found a CAT in an eight-year-old boy who was being evaluated for fever of unknown origin. Heterogenous tumor masses were found in the right ventricle and right ventricular outflow tract. These masses had embolised further into both the left and right pulmonary arteries. Surgery was done on conventional cardiopulmonary bypass and all the tumor masses were excised. Histopathological examination revealed features consistent with CAT. Very few cases of CAT have been reported in the world literature. Resection is curative but if incomplete, recurrences are known to occur.

Keywords: Calcified amorphous tumour; Non-neoplastic cardiac tumor.

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Introduction

Calcified amorphous tumor is a rare, non-neoplastic tumour of the heart of unknown etiology. It usually presents with symptoms of flow obstruction or embolisation. It is often mistaken for cardiac myxoma, fibroma, calcified mural thrombus, vegetations of infective endocarditis, prominent eustachian valve, Chiari's network, malignancy or may be an incidental finding on imaging.[1] Patients with chronic or end-stage renal disease are also known to present with calcium deposits on the mitral valve annulus, leaflets and the coronary arteries.[2]

CAT is usually attached to the endomyocardium by dense connective tissue and has to be differentiated from mimicking lesions by histopathological examination. Since most often patients are symptomatic, they have to be excised in toto. We report a rare case of calcified amorphous tumor in an 8-year-old child who was evaluated for pyrexia of unknown origin and eventually found to have a mass in the right ventricular cavity which had embolised downstream into branch pulmonary arteries. Surgery was done to excise the entire tumour. Histopathologically it was confirmed to be CAT. Patient has no recurrence of tumour at 8 months follow up.

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Case report

Eight-year-old male child presented with fever of 3 months duration. After extensive evaluation by pediatricians, he was found to have multiple masses in the right ventricular cavity. His vitals were stable. He had normal heart sounds: no murmurs or added sounds. He had normal vesicular breath sounds. His erythrocyte sedimentation rate was raised. C-Reactive protein was positive. Three consecutive blood cultures were negative. His vitamin D3 and serum calcium levels were normal. He had no hypercoagulable state. 2D echocardiography, computer tomography scan and magnetic resonance imaging revealed multiple, well-defined, ovoid, heterogenous masses varying from 1 to 5 cm in size, in the right ventricle, right ventricular outflow tract and main pulmonary artery with occlusion of the right lower lobar artery (Fig. 1).

He underwent surgery on conventional CPB. Branch pulmonary arteries were dissected up to hila. Right atrium and main pulmonary artery were opened. Large masses were seen in the right ventricle (Fig. 2) and its outflow tract measuring 5 and 3 cm respectively (Fig. 3). Tumor emboli were seen filling the left upper, left lower, right upper and right middle lobe branch pulmonary arteries. All masses were excised after opening the branch pulmonary arteries. Right lower lobe branch pulmonary artery was totally occluded by the embolus and could not be removed. Histopathological examination confirmed CAT. It showed predominantly fibrinous acellular material with large areas of dystrophic calcification and focal collection of inflammatory cells admixed with proliferating fibroblasts (Fig. 4). At 8 months follow-up, 2D echocardiography showed no residual mass or recurrence.

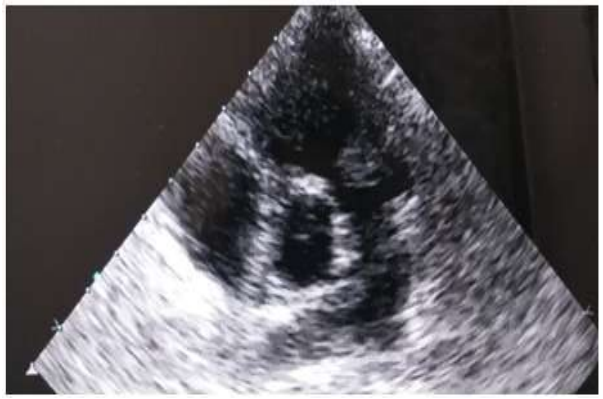

Fig. 1: Echo image of the tumour in right ventricle

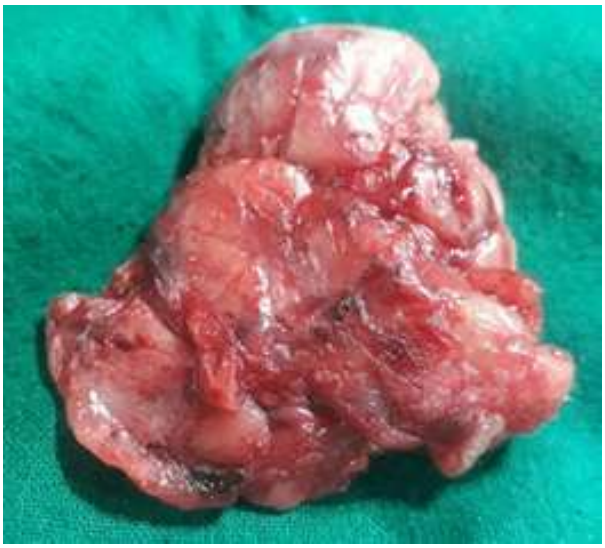

Fig. 2: Tumour from right ventricular cavity

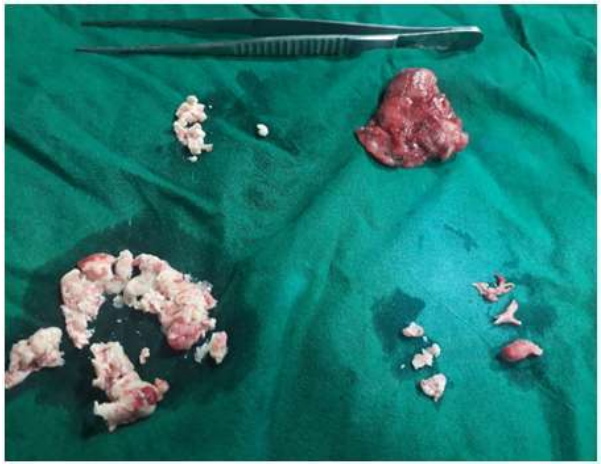

Fig. 3: Entire tumour tissue excised from right ventricle upto branch pulmonary arteries

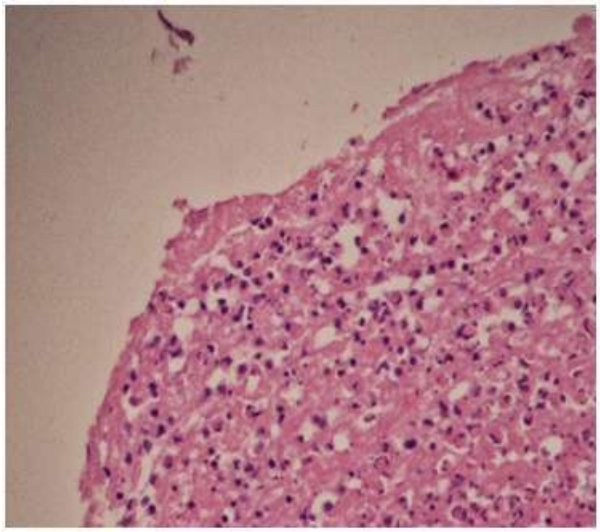

Fig. 4: Histomicrograph showing amorphous material [pink] with specks of calcium [blue]

Discussion

Cardiac CAT is a rare entity and was first described by Reynolds *et al.* in 1997[1]. These tumors are often seen in the left ventricle though they are known to occur in all four chambers and on the mitral valve. They are known to occur more commonly in females than the males; more often in adults than children.

They are mobile and are often mistaken for a myxoma, fibroelastoma, mitral annular calcification, hydatid cyst, echinococcal cyst or an organized thrombus, embolus or vegetation.[1] The presence of thrombotic tendencies in several of the patients supports the possibility that this entity is thrombotic in origin with heterogenous initiating events.[1] They are also known to occur in elderly patients with end-stage renal disease with exuberant mitral annular calcification. Abnormalities in the calcium, phosphoros and vitamin D3 metabolism is thought of as a probable cause of CAT in patients with chronic kidney disease.[2] Patients present with symptoms of dyspnea, syncope, embolic episodes or chest discomfort. They are usually diagnosed by a 2D echocardiography which delineates their morphology, location and echogenicity. To confirm, other investigations like transesophageal echocardiography, computer tomography scan or magnetic resonance imaging may prove helpful. Subjecting the excised mass for histopathological examination is needed to diagnose and decide on further course of treatment.

Histologically, CAT shows calcified nodules in an amorphous, eosinophilic, degenerated fibrin background with focal inflammation.[1,3] Though very rarely seen in association with rheumatic heart disease, when they do, they tend to occur following a degenerative, calcific process that extends to the posterobasal myocardium.[4] These tumors are covered with fibrin and attached to the subendocardium with dense connective tissue or granulation tissue. They need to be excised to prevent embolization and flow obstruction. Most often, excision is curative and long-term prognosis is excellent but if resection is incomplete, recurrences can occur. Left ventricular tumors undergo high shear stress during systole; hence, tend to break off and embolise. Hence they should be resected at the earliest.[4,5] Right sided lesions tend to embolise into main pulmonary artery and its branches.

Conclusion

Calcified amorphous tumors are non-neoplastic masses that are known to occur more commonly in the ventricles, causing flow obstruction and embolic episodes. Although complete resection is curative, recurrences have been reported from residual tumor masses in some of the patients. We report an interesting case of CAT that had embolised into the pulmonary artery and its branches and was successfully excised. It is very rarely seen in children such as our eight-year-old patient.

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