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UNUSUAL PRESENTATION OF PRIMARY PLEOMORPHIC RHABDOMYOSARCOMA OF HEART: COMPLETE ATRIOVENTRICULAR BLOCK AND SEVERE MITRAL REGURGITATION

Cardiology

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ABSTRACT

Primary cardiac tumours are rare and difficult to diagnose because most are asymptomatic or have varied non-specific presentations. This report describes a 29-year-old man presenting with complete heart block, primary cardiac tumour in the left atrium, and severe mitral regurgitation. In view of the primary severe mitral regurgitation and complete heart block, mitral valve repair and pacemaker insertion were planned. Mitral valve repair was done with 29mm St Jude tailor annuloplasty ring, and the biopsy was taken from the nodules noted in the left atrium; temporary right ventricular epicardial pacemaker implantation and CABG with SVG to PDA graft were done to look for the recovery of complete heart block. Histopathological examination revealed pleomorphic rhabdomyosarcoma. The patient developed renal failure and liver failure during the postoperative period and expired after 10 days.

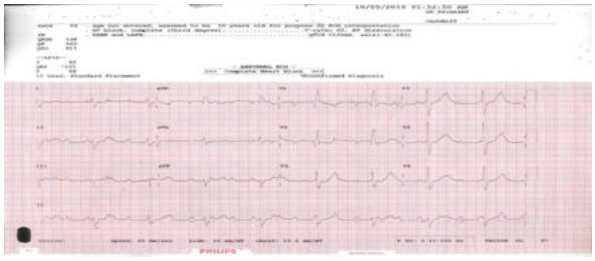
KEYWORDS

INTRODUCTION –

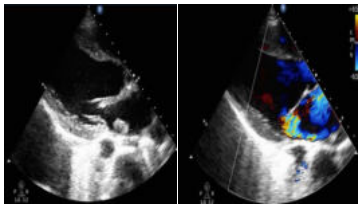
Cardiac tumours are rare and have an incidence of less than 0.33%.[1] 3/4th of the tumours are benign, and only 1/4th are malignant. Out of the malignant tumours, rhabdomyosarcoma is the second most common malignant tumour of the heart after angiosarcoma. It is a very aggressive tumour with high mortality. Herein, we report a case of primary pleomorphic rhabdomyosarcoma, which had a deceptive presentation.

Clinical details - A 29-year-old male patient presented to the hospital with acute pulmonary oedema. He is hypertensive, non-diabetic, non-smoker, and non-alcoholic. He had a history of CVA (cerebellar stroke) one year back, from which he recovered after two months.

12 lead ECG showed complete heart block with wide complex escape rhythm with RBBB morphology with left anterior fascicular block with a rate of about 54/min.



2D- echocardiography revealed a non-pedunculated mass lesion in the left atrium near the posterior wall with prolapse of anterior mitral valve leaflet in parasternal long-axis view. Colour doppler in parasternal long-axis view revealed severe eccentric mitral regurgitation directed towards the posterior wall.

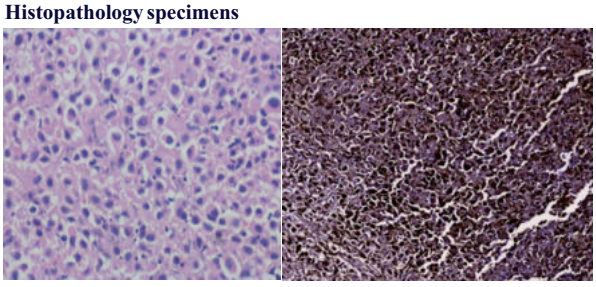


Management - Immediately, the patient was stabilized with temporary pacemaker insertion and appropriate medical therapy. The mass lesion was initially thought to be thrombus as the patient had a history of a cerebrovascular accident at a young age and as it disappeared after 5 days of therapy with anticoagulation therapy. In view of the primary severe mitral regurgitation and complete heart block, mitral valve repair and pacemaker insertion were planned.

Intervention -

Coronary angiogram showed ectasia of coronary vessels with an intermediate lesion in the proximal part of the right coronary artery. Necrotic granulomatous pericardial cavity near the aortic pericardial reflection, multiple nodules on the floor of LA and near the roof of LA near the fibrous skeleton of mitral and tricuspid annular junction. The Mitral valve was distorted with annular dilatation involving P2 and P3 areas. Mitral valve repair was done with a 29mm St Jude tailor annuloplasty ring. Biopsy was taken from the nodules of the left atrium, temporary right ventricular epicardial pacemaker implantation, and CABG with SVG to PDA graft was done to look for the recovery of complete heart block.

Histopathology specimens



Histopathology specimens showing sheets of large pleomorphic cells with a hyperchromatic nucleus, cells are polygonal, and few cells have tennis racket-like appearance and vimentin-positive suggestive of rhabdomyosarcoma. The patient developed renal failure and liver failure during the postoperative period and expired after 10 days.

DISCUSSION –

Cardiac masses frequently pose significant diagnostic and therapeutic challenges. In most cases, they are detected as an incidental finding,

and further evaluation may lead to confirmation of a cardiac tumor. The finding of a tumor is generally an uncommon event. However, other masses, such as thrombi or vegetations, are much more common. Thrombus is expected in the presence of regional wall motion abnormality or aneurysm with left ventricular systolic dysfunction, whereas vegetations are to be suspected if they have a history suggestive of infective endocarditis.

Cardiac tumors are mainly divided into two groups, primary and secondary tumors. Primary tumors are very rare, with an autopsy incidence of 0.001% to 0.03%.[2]They include benign or malignant neoplasms that may arise from any tissue of the heart. Secondary or metastatic cardiac tumors are 30 times more common than primary neoplasms, with an autopsy incidence of 1.7% to 14%.[3] Primary cardiac tumours are very rare, with the most common being myxoma. The majority of the myxomas are found in the left atrium, along with female dominance. Primary cardiac sarcomas are the most common malignant primary cardiac tumor that constitutes approximately 1% of all soft tissue sarcomas.4 The age of presentation for cardiac sarcomas ranges from 1 to 76 years, with a mean age of around 40.4 Angiosarcomas and unclassified sarcomas account for approximately 76% of all cardiac sarcomas, of which angiosarcomas are the most common[5] Rhabdomyosarcoma is the most common form of cardiac sarcoma in children.

First-degree AV block commonly occurs in patients with myxoma. However, there are rare reported cases of AV dissociation in patients with LA tumors.6,7 Since the AV node is a right-sided structure; direct compression can be more easily explained if the mass is located in the right atrium. In the present case, intraoperative findings revealed the nodules present on the roof of the left atrium near the fibrous skeleton of mitral and tricuspid annular junction. These findings also explain the presence of both the right bundle brank block and left anterior fascicular block.

In the present case report, the patient had a history of stroke one year back, which may be due to embolism of the small fragments of the tumour. Hence, even though rare, primary cardiac tumors should be excluded, particularly in young patients without comorbidities who present with stroke.

CONCLUSION -

Preoperative diagnosis of rhabdomyosarcoma of the heart is difficult as in this case because of varied and deceptive presentation. Surgical excision of the tumour is the management strategy in localized tumours to provide palliative relief and improve short-term survival followed by chemotherapy or radiotherapy. Prognosis is very poor, with survival not more than two years in most patients despite all the therapeutic strategies.

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