

Case Report

Intramyocardial choristoma-a rare entity: first-ever case report and literature review

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Received: 17 May 2023

Accepted: 13 July 2023

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ABSTRACT

We report the first-ever case of 'Intramyocardial choristoma' documented in English-language literature. A 71-year-old woman, during her preoperative echocardiogram assessment, noted to have a large left ventricle (LV) mass with severe mitral regurgitation (MR). The patient was asymptomatic, with CT scan reported a large dense calcified LV mass. She underwent mitral valve replacement with LV mass resection operation. The mass was completely excised. Histopathological examinations show intramyocardial choristoma, with the lesion shows cardiac muscle with areas of calcification and mature bone tissue, with clear margins. Discharged well post operatively. Commonest clinical scenarios for intracardiac masses are myxomas, infective endocarditis with vegetation and thrombus. Primary cardiac tumours are very rare. Choristoma are rare benign tumour, with histologically normal tissues in an abnormal location, with a very rare recurrence rate. Transthoracic echocardiogram is reliable for the evaluation of the intracardiac masses, but the use of contrast echocardiogram gives additional benefit in analysing the characteristics of the intracardiac mass. For complex lesion, such as intramural component, extension into the inflow or outflow and into pericardial/extracardiac, MRI have added benefits. The treatment of choice is complete surgical resection. Follow up surveillance scans are recommended, as there is a very low risk of recurrence with uncertain prognosis. A multidisciplinary team approach is highly recommended in managing the patient pre-operative, intra-operative and post-operatively. Post operatively, regular follow up with imaging is recommended.

Keywords: Intramyocardial choristoma, Intracardiac masses, Choristoma

INTRODUCTION

The commonest differential diagnosis for intracardiac masses is myxomas, infective endocarditis with vegetation and thrombus.¹ Besides these, other diagnosis of intracardiac mass includes cardiac tumours. We report possibly the first-ever documented case of 'Intramyocardial choristoma' in English-language literature. We would be discussing about the case, the challenges in investigating and managing this very rare disease based on few clinically reported studies which are not intramyocardial in nature, but generally on choristomas. Our approach, hopefully would be a standard of care of management to be used in the future.

CASE REPORT

A 71-year-old woman, during her preoperative echocardiogram assessment for an elective urology operation, noted to have a large LV mass with severe MR and was radiologically reported as myxoma.

The patient was otherwise asymptomatic. No constitutional symptoms of malignancy. She was treated empirically as infective endocarditis and was started with an intravenous antibiotic which was stopped after 2 weeks as all the cultures came back negative. Transthoracic echocardiogram shows large LV mass, measuring 6×2 cm. CT scan of thorax shows large dense

calcified LV mass, measuring approximately 3.2×7.4×5.1 cm. She was scheduled for elective operation of mitral repair with LV mass resection.

Intraoperatively, there were calcified nodules at the left atrium (LA) wall and the consistency of the LA wall at the interatrial septum was hard and bony. The mitral valve and peri-valvular apparatus were also calcified. A large regular LV mass, with hard and bony in consistency noted with attachment to the lower part of the posterior mitral valve leaflet (LV part) and was excised completely together with the mitral leaflets. Mitral valve replacement done with bioprosthesis mitral valve.

The HPE reported as intramyocardium choristoma. Microscopically, the lesion shows cardiac muscle with areas of calcification and mature bone tissue. No malignant cell was seen with clear tissue margin. Post operation, the patient recovered and discharged well with a scheduled follow-up. On follow up echocardiogram in 3 months and 6 months, noted no intracardiac mass, with functioning bioprosthesis mitral valve. Planned to repeat yearly surveillance echocardiogram.

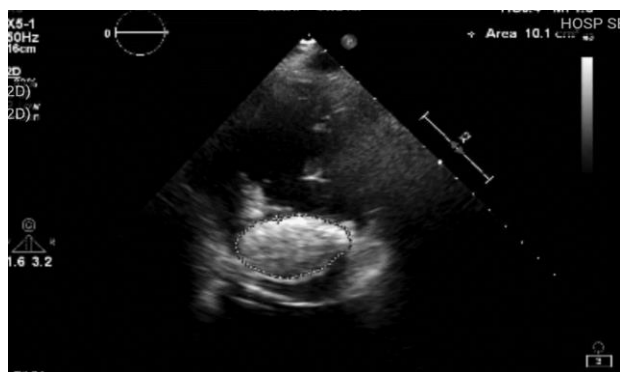


Figure 1: Echocardiogram shows large LV mass, measuring 6×3.2 cm.



Figure 2: CT thorax of the patient, shows large dense calcified mass seen at LV, measuring approximately 3.2×7.4×5.1 cm.

DISCUSSION

The commonest differential diagnosis for intracardiac masses are myxomas, infective endocarditis with vegetation and thrombus.¹ Besides these, other diagnosis of intracardiac mass includes cardiac tumours. Cardiac tumours are most commonly metastatic in origin, with the common primary malignancy is of thyroid, lung, breast, malignant melanoma and lymphomas.¹ Primary cardiac tumours are very rare with the incidence of 0.0017% to 0.28%, and usually benign (3/4 of all) with myxomas compromising 50% of it.¹

Choristoma lesions are documented as rare benign tumour, with histologically normal tissues in a site at which it is not normally detected (abnormal location).² The rate of recurrence after surgical excision is very rare.³

The documented examples of choristomas are like choristoma of the eyelid or orbit, corneal choristoma, salivary gland choristoma, choristoma of the oral cavity and osseous choristoma.^{3,4}

As an example, osseous choristoma is a tumour-like growth of lamellar bone in a location where bone does not normally form.³ The growth of a choristoma is normally regulated, compared to a neoplasm.⁹

In view of the very rare nature of this disease, the challenge is to diagnose it accurately (with a few differential diagnosis) and for a proper standardised management of the disease.

Transthoracic echocardiogram is reliable for the evaluation of the intracardiac masses in terms of location, attachment, shape, size, mobility and hemodynamic derangements.⁵

The use of contrast echocardiogram gives additional benefit in analysing the characteristics of the intracardiac mass: such as, thrombus shows complete lack of enhancement, myxomas shows partial/incomplete enhancement, and intracardiac tumour shows complete enhancement.⁶

For complex lesion, such as intramural component, extension into the inflow or outflow, obstruction of outflow tract, extension into pericardial/extracardiac, MRI have an added benefit, especially to analysis the complete morphology and functional evaluation of the mass.⁷

Complete surgical resection is the treatment of choice, documented for all the other types of choristoma.^{3,8} The recurrence is very rare, but still have been documented and the prognosis of the recurrence disease is uncertain.³ Thus, a proper follow up, with surveillance scans should be the standard of care for this patient post operatively.

In our institution, we recommend, if transthoracic echocardiogram shows features of complex intracardiac lesion, an MRI should be the next step of imaging preoperatively. A multidisciplinary discussion would be done with the cardiac surgeons, radiologist and the anaesthesiologist, prior to the surgical management. Post operatively, a regular multi-modulated follow-up by clinical symptoms and imaging (transthoracic echocardiogram in 3 months post-operative, 6 months and yearly) is the current standard of care in our institution.

CONCLUSION

As a conclusion, this is a very rare disease, thus a multidisciplinary management approach involving the cardiac surgeons, anaesthetists and radiologists should be done from the preoperative stages, intraoperative and until post-operative care. From the various reported studies of choristoma from various areas of the body, it's clear that a proper surgical excision with a clear margin is the treatment of choice. A proper follow up with routine scans is also necessary, as the risk of recurrence is present in this rare disease.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Francis T, Kuthubuddeen MH, Ranganathan R, Hamzah MK, Arif MNM, Jasid AM. Intramyocardial choristoma-a rare entity: first-ever case report and literature review. Int Surg J 2023;10:1385-7.