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LEFT VENTRICULAR MYXOMA PRESENTING AS CEREBELLAR ATAXIA IN A POST ANGIOPLASTY YOUNG PATIENT ; POSSIBLE MULTIPLE SYSTEMIC METASTASES? - A CASE REPORT WITH REVIEW OF LITERATURE

Cardiac Surgery

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ABSTRACT

Primary left ventricular myxomas are extremely rare. A common modality of presentation of such tumours is an embolic event or cerebrovascular stroke; rarely it may lead to left ventricular outflow tract obstruction. We are reporting a case of a young male, who was a known case of myocardial infarction , post angioplasty and stenting two years back, who presented with cerebellar ataxia. Preliminary radiological evaluation of the brain showed left superior cerebellar artery infarct. Later cardiologic evaluation showed presence of a left ventricular thrombus which on table was revealed to be a myxoma. The patient underwent subsequent surgical resection of the myxoma. This case is being presented due to the rarity of a left ventricular myxoma presenting in a young patient with ischaemic heart disease; and the possibility of the left anterior descending artery lesion being a systemic metastasis of the myxoma that was incidentally diagnosed earlier than the primary.

KEYWORDS

Myxoma, Left Ventricular Thrombus, Cerebellar Infarct

INTRODUCTION:

Cardiac myxomas are by far the most common primary tumours of the heart though primary left ventricular myxomas are extremely rare. A common modality of presentation of such tumours is an embolic event or cerebrovascular stroke; rarely it may lead to left ventricular outflow tract obstruction.

CASE REPORT:

A 27 year old male had reported to the emergency department of our hospital in June 2019 with an episode of syncope followed by ataxic symptoms. Past history revealed a previous diagnosis of ischaemic heart disease in 2017; coronary angiography had revealed proximal thrombotic lesion (70%) in the left anterior descending artery (LAD). A 2D echocardiography had showed regional wall motion abnormalities involving the apex and inter ventricular septum. There were no other abnormalities. Left ventricular ejection fraction was 45%. Following this he had undergone PAMI (Primary angioplasty in myocardial infarction) to LAD and the post operative period was uneventful.

Following his arrival after the attack of syncope, and after eliciting the history, an urgent MDCT was performed that revealed a well defined lesion with cytotoxic oedema in the left superior anterior cerebellar hemisphere, in the left superior cerebellar artery territory. Multiple small lacunar infarcts in the bilateral centrum semiovale regions. It also showed minimal ischaemic periventricular white matter pallor (Fuzeka Type I changes with few lacunar infarcts). In accordance with the previous history of angioplasty, a detailed cardiologic evaluation was done.

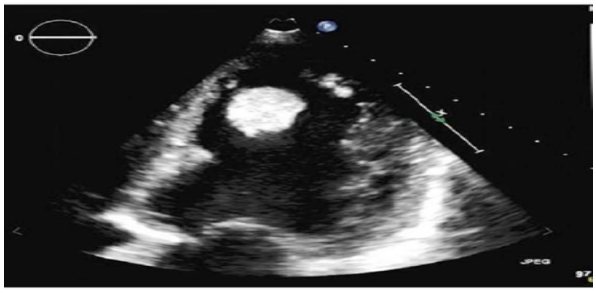

Figure 1

2D Echo (Fig 1) revealed a large mobile clot, 4 x 1.75 cm, in the left ventricular cavity, with basal, middistal, anterior, anteroseptal, anterolateral segment hypokinetic, The valves and septa were said to be normal. Left ventricular ejection fraction was 35%. The clot was suspected to have been formed as a result of left ventricular remodelling post myocardial infarction.

A cardiac Computed Tomography showed ballooning of the left ventricle with paradoxical movement.

The patient was taken up for emergency surgery after taking the necessary consents. After taking the patient on cardiopulmonary bypass, the left ventricle was opened and the clot demonstrated. Intra-operatively, the lesion was revealed to be a myxoma, of size approximately 5 x 4 x 3 cm.

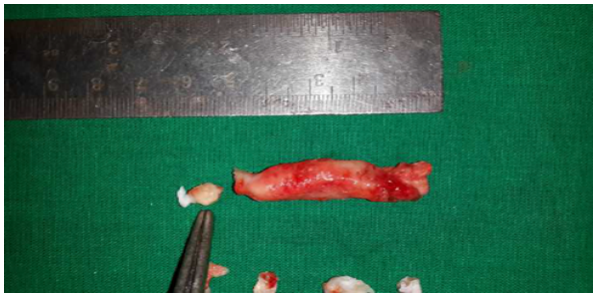

Figure 2

The tumour was extracted and sent for histopathological examination (Fig 2).

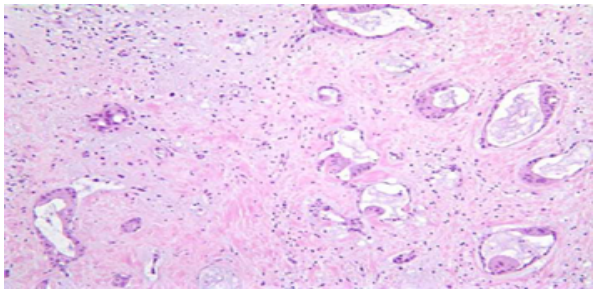

Figure 3

Histological features consisted of stellate or globular myxoma cells with abundant eosinophilic cytoplasm, indistinct cell borders, oval nucleus with open chromatin and indistinct nuclei, consistent with

myxoma (Fig 3).

DISCUSSION :

Primary cardiac tumours are uncommon with incidence of 0.02%[1][2]. Commonest left ventricular mass is a thrombus[3]. Benign cardiac myxomas constitute 88% of cardiac tumor cases[1][4]. LV myxomas account for only 2.5% of cases[4][5]

Cardiac myxomas are associated with embolism, obstruction to the outflow tract, arrhythmia, and other constitutional symptoms. Embolism occurs in 30% to 40% of all cases. The risk of embolisation mainly depends on the morphology of myxoma rather than its size. There are 2 types of cardiac myxomas: friable polypoid type and smooth-surface rounded type, out of which the polypoidal type tends to result in embolism because of its friable consistency and intracavitary location[1][6]. Obstructive symptoms (valvular ball-valve obstruction) occur in 54%–95% of patients[7][8][9]. Many cases masquerade as mitral valve disease[7]. Systemic emboli occur in 10%–45% of myxoma patients[7][8][9][10]. More than two-thirds of myxomatous emboli migrate to the central nervous system[7][11][12], but any arterial bed may be affected, leading to a great variety of symptoms and signs. Recorded cases document emboli in the upper and lower extremities, aortic saddle, coronary arteries, kidneys, liver, spleen, eye, skin, and more[7][13]. Cerebral emboli may lead to numerous fusiform aneurysms, described as typical of cardiac myxomas[7][14].

Coming to our case, there were quite a few points that demanded our attention; these being simultaneously the points that distinguished this particular presentation from the ones that have been published over the years.

These are:

A. Age of presentation - myxomas have been reported in people ranging from 3 to 83 years in age. However, the mean age of sporadic presentation is 56 years, whereas that for familial cases, it is 25 years[15]. Our case was a sporadic one that occurred in a young male in his twenties; there was no history of myxoma presentation amongst his relatives.

B. Sex: 75% of sporadic myxomas occur in females[15]; however our case had a sporadic presentation in a young male.

C. Ventricular myxoma: The incidence of a LV myxoma is approximately 2.5% of cases[16][17].

D. Chances of a primary atherosclerotic lesion also cannot be ruled out, more so as the timing of presentation of the myxoma and the ischaemic heart disease was different.

E. A probable coronary embolus manifested as the sole lesion, with no evidence of a primary in 2D echoes done post myocardial infarction; and later the same patient returned with features of a cerebellar infarct with the primary myxoma present in the left ventricle. This could be explained in two manners:

1) Either a primary atherosclerotic lesion that manifested in the first instance

2) A coronary embolus of a primary occult myxoma manifested as ischaemic heart disease, and the myxoma presented with cerebellar emboli 2 years later, with itself being grown in size. In this context it becomes a case of multiple metastases from a primary myxoma that manifested in different periods.

Hypothesising primary atherosclerosis and myxoma as a separate entity: Our patient, prior to his diagnosis of ischaemic heart disease, had been a healthy young male with no reported history of ischaemic heart disease in the family or without any cardiac risk factors. Hence after the diagnosis of a myxoma after 2 years, it appeared plausible to us that the prior myocardial ischaemia could actually be caused by coronary metastasis from an occult primary myxoma that was undetectable in the 2D echo performed in 2017. Also, no similar association has been reported in the literature.

Hypothesising coronary embolisation from an occult primary: Quite a few cases of myocardial infarction or ischaemic heart disease caused due to coronary embolisation from myxomas have been reported in literature.

However all the available literature documents that all these cases have been due to left atrial myxomas. Moreover, the atrial myxomas were detected at the time of presentation with ischaemic heart disease.

Jayaprakash et al have reported a case of a myxoma manifesting as an acute ST elevation myocardial infarction in a 45 year old lady[18]. Braun S et al have reviewed 40 cases of myocardial infarction due to left atrial myxoma from 1970-2002. They found that the common culprit was right coronary artery with inferior wall myocardial infarction seen in most of the cases[19]. Al Zahrani et al in 2014 have compiled the reported cases of atrial myxoma associated myocardial infarctions from 2003 to 2013 and found 16 cases[20]. We have gone through the available literature to compile the published cases of ischaemic heart disease and myocardial infarction post 2013 till date. Totally we could find 6 cases, the results have been presented in the table below:

Case No	Year of publication	First author	Site of Obstruction	Source of embolus	Age of patient
1	2013	Arcenas[21]	Obtuse marginal artery (OM)	Left atrium	53
2	2014	Al Zahrani	Not mentioned	Left atrial myxoma	22
3	2016	Chaudhuri[22]	RCA	Left atrium into the left venrticle	70
4	2016	Al-Fakhouri[23]	LAD, D2 (second diagonal), OM2	Left atrium	50
5	2016	Demir[24]	LCX	Left atrium	77

As it is evident from the literature, all the cases have left atrial myxomas as the source of embolus. A left ventricular myxoma has not been reported to be the probable source of embolus. Though the commonest is RCA (right coronary artery), yet a few cases of LAD (left coronary artery) involvement have been reported.

Hypothesising an occult primary: Not much documentation is available about occult myxomas till date. However, it has been hypothesised that transthoracic echocardiogram may not be able to identify tumours smaller than 5 mm. Whenever a very small tumour is suspected, trans oesophageal echocardiography is required[25][26][27].

Hypothesising a multi system embolisation: Having a coronary embolus and cerebellar emboli that manifested one after the other makes our case unique, with limited sources in the literature[28][29][30].

CONCLUSION :

Ventricular myxoma, just like atrial myxoma, can have multiple modes of presentation. In patients presenting with left ventricular thrombotic masses in 2D Echo, the chances of systemic embolisation should always be taken into account. Surgical resection is the treatment of choice.

Ethical Approval:

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/ or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent:

Informed consent was obtained from all individual participants included in the study.

Conflict Of Interest:

The authors declare that they have no conflict of interest.

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