Spontaneous Left atrial intramural multiseptated dissecting hematoma

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Abstract

Our case was initially admitted with presumptive diagnosis of Non-ST elevated myocardial infarction in congestive heart failure and was later found to have large left atrial (LA) mass. Apart from complete echocardiography study, we took help of multimodality imaging to better characterise this LA mass.However we did not have a confirmed diagnosis. Cardiac surgery was performed and surprisingly revealed large LA mass with pockets of fresh blood inside LA. Pathological specimen confirmed the presence of hematoma ruling out other atrial neoplasms. Though contrast echocardiography and cardiac magnetic resonance imaging were closest to intra-operative diagnoses, In the absence of any pre-disposing factors, final diagnosis was made at surgery and histopathological diagnosis.

CASE PRESENTATION

Our patient is 69 years old gentleman known to have non-insulin dependent diabetes mellitus, essential hypertension, dyslipidaemia and mild renal impairment. He had atypical angina 7 years back with normal epicardial coronaries on coronary angiogram (CAG). He again had atypical chest pain one year back and underwent bike stress echo, which was negative for inducible ischemia at high workload. His echo till then showed preserved biventricular size and systolic function. He had mild LA dilatation with diastolic dysfunction.

He presented this time with 5 days history of variable threshold angina and progressive shortness of breath. He had associated upper back pain of similar intensity. His electrocardiogram (EKG) showed diffuse concave ST elevation in precordial leads and PR segment depression with highest highly sensitive troponin-I leak of 0.77ng/ml (normal value 0.02-0.06). He had pulmonary congestion on chest X-ray. In light of refractory angina and heart failure despite optimal medical management, He was taken up for urgent CAG, which again revealed normal epicardial coronaries. Subsequent labs were unremarkable. Later he underwent transthoracic echo (TTE) which showed a large mass occupying the whole of Left atrium (LA) and attached to the posterior wall and interatrial septum. The mass appeared inhomogeneous and cystic with multiple septations **(Figure 1a and 1b).** There was no colour flow observed through the mass**.** There was underlying moderate LA dilatation with interatrial septum (IAS) bulging into right atrium suggestive of high LA pressure. Left ventricular size and systolic function were normal. We went ahead with contrast echocardiography, which showed complete non-opacification of LA mass suggestive of avascular nature **(Figure 2 and supplementary video 1).** Computed tomography (CT) with contrast was primarily performed to rule out aortic dissection because of his presentation. But it as well was suggestive of large mass in the LA **(Figure 3).** To further characterise the mass, we performed transoesophageal echo (TEE) showing large well encapsulated mass with echo-density lower than underlying LA wall. There were multiple irregular sized

cystic spaces with loosely bound septations **(supplementary video 2).** The mass was attached to IAS and extending till the base of posterior mitral leaflet and whole of posterior LA wall **(Figures 4a, 4b and supplementary video 3 to 6).** There was mild inflow obstruction with mean gradient across mitral valve of 5mmhg. There was no pulmonary venous obstruction noted. Our differential diagnosis at this point was cardiac hydatidosis, polycystic LA myxoma, LA dissection and LA hematoma.

Finally, cardiac magnetic resonance (CMR) imaging was performed which showed a large smoothly marginated LA mass measuring 5.1*6.3 centimetres related to posterior wall and IAS **(Figures 5a and 5b)** . It was not attached to Mitral valve with no extension beyond the confines of LA. The mass had slightly heterogenous signal intensity being moderately hypertense on spin echo sequences (black blood imaging, T2 weighted) and moderately hypointense on gradient echo (white blood imaging, T1 weighted cine). There was a suggestion that this mass could be thrombus.

SURGERY AND HISTOPATHOLOGY

Based on all above findings and symptomatic status of our patient, it was unanimously decided by our multidisciplinary team to operate on this LA mass. Cardiac surgery was performed with median sternotomy and vertical pericardiotomy. Cardiopulmonary bypass established via ascending aorta and bicaval cannulation with myocardial protection delivered by antegrade cardioplegia. After opening the right atrium and exposing LA via fossa ovalis, a large LA mass extending from pulmonary veins superiorly and mitral annulus inferiorly with attachments to IAS and almost all of the posterior wall of LA was seen. After opening the mass, it looked like organised pockets of fresh clots**.** The mass occupying the posterior wall separated endocardium and epicardium. All the clots were evacuated followed by marsupialisation of LA cavity. The posterior wall and IAS were repaired with bovine pericardial patch. Histopathological examination of mass confirmed organised thrombus with fibrin **(Figure 6).** Biopsy of the LA septal tissue confirmed focal necrosis of the endocardium with thrombosis. No findings related to malignancy, hydatidosis, myxoma, endocarditis and amyloidosis was observed. The post-operative course was uneventful and patient was discharged on sixth day post-surgery. Pre-discharge echo showed no residual hematoma with patched LA wall on septal and posterior aspect.

DISCUSSION

Spontaneous left atrial intramural dissecting hematoma is exceedingly rare entity with unknown true incidence. Most of the information is available from isolated case reports. From aetiological perspective LAIH causes could be grouped into 4 major categories based on available published literature. Firstly, it can be secondary to complications of post cardiac surgeries like mitral and aortic valve replacement. Secondly it can be a result of iatrogenic complications secondary to percutaneous interventions (mainly due to high-risk coronary interventions, and during radiofrequency Cath ablation in electrophysiology lab **[2-5].** Thirdly it has been reported to occur in conditions like amyloidosis, mitral annular calcification and/or annular abscess, blunt chest trauma and dissecting aortic aneurysms and lastly as in our case it can be spontaneous**[6-9].** Usually LAIH has predilection to posterior LA wall (81%) due to lower quantity of fibrous tissue; however, in our case it occupied the whole of left atrium. Our patient posed a diagnostic dilemma as he did not have any predisposing factors and closely mimicked acute coronary syndrome (ACS)/pericarditis at presentation which mandated coronary angiogram. Retrospectively we thought that PR segment depression on presentation EKG could be secondary to atrial ischemia rather than ACS/pericarditis though there was no PR segment elevation. The pathological report did show necrosis of underlying LA endocardium. We used unfractionated heparin for ACS treatment initially which could be hypothesized to cause cystic degeneration of LA intramural thrombus. There is a close overlap between left atrial dissection and LAIH. The former is thought to originate from contained atrioventricular separation allowing pressurised blood flow from left ventricle to dissect the layers of posterior LA. In our case surgeons observed separation of endocardium and epicardium in posterior LA wall, henceforth it might be appropriate to term this LA mass as spontaneous intramural multiseptated dissecting hematoma. There are no definitive guidelines for management with some cases having predisposing factors being observed conservatively and majority of the cases went to operating theatre, as they were symptomatic. The work up of LAIH starts from imaging as clinical and laboratory

parameters are mostly variable and misleading.

CONCLUSION

This case posed us with a diagnostic dilemma throughout the work up including high-resolution multimodality imaging. Though contrast echocardiography and cardiac MRI were closest to intraoperative diagnosis but in the absence of predisposing factors, we were left with differential diagnosis until our surgeons did gross examination in the operating theatre along with microscopic pathological diagnosis. In view of the rarity of such peculiar finding and the challenging presentation along with lack of clear consensus for diagnosis and management we thought to share our experience.

FIGURES LEGENDS

Fig 1a & 1b

Parasternal long axis and Apical 4 chamber view suggestive of large polycystic inhomogeneous mass filling the entire LA.

Fig 2

Contrast echocardiography showing complete non-opacification of LA suggestive of avascular mass.

Fig 3

CT showed large mass in LA without any clear attachments to IAS and free from Mitral valve.

Fig 4a & 4b

2DTEE Long axis mid-esophageal view showing large well encapsulated mass in LA with loosely bound septations and lower echogenicity than the underlying endocardium. Live 3DTEE from the same 2D view shows the attachments to IAS and posterior wall sparing the mitral valve.

Fig 5a & 5b

CMR showing smoothly marginated LA mass (5.1*6.3) cms being moderately hyperintense on T2 weighted imaging and moderately hypointense on T1 weighted cine.

Fig 6

Low power microscopic examination of the specimen showing fibrinous material with blood consistent with hematoma without any evidence of other pathology. (Hematoxylin & eosin stained).

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