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A CASE REPORT OF CONGENITAL GIANT LEFT ATRIAL APPENDAGE ANEURYSM WITH THROMBUS PRESENTING AS ATRIAL ARRHYTHMIA.

Cardiology

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ABSTRACT

Congenital left atrial appendage aneurysm are a rare entities and are caused by congenital dysplasia of the atrial muscle. Less than 100 cases have been reported in literature so far. Patients usually present with dyspnoea ,palpitations ,thromboembolic phenomenon in their 3rd decade with slight higher preponderance in females.In asymptomatic individuals the condition is often diagnosed incidentally during cardiac imaging . We hereby report a case of 30 year old male with a giant LAAA containing thrombus presenting with palpitations diagnosed with echocardiography and cardiac MRI. The patient successfully underwent aneurysmal resection sugery, post which atrial arrhthias stopped and symptoms relieved. Therefore,even though rare,if a young patient presents with atrial tachyarrthymia with no other associated cardiac pathologies, a LAAA should be ruled out. Since this condition has been shown to be associated with increased morbidity and mortality. A transthoracic echocardiogram or even better,a TEE colour Doppler echocardiogram may suggest the diagnosis by demonstrating the exchange of blood between LA and LAAA. Surgical resection is the standard modalitiy of treatment even in asymptomatic individuals to reduce risk of associated complications. **Learning objectives :** It is a rare case of giant left atrial appendage aneurysm, few cases have been reported, LA appendage aneurysm usually present as atrial arrhythmias ,stroke or as incidental diagnosis on imaging.Though patients may be asymptomatic it still requires operative intervention since it has very high risk of atrial arrhythmias and thrombus formation which result into stroke.Therefore, if a young patient presents with atrial tachyarrthymia with no other cardiac pathologies, LAAA should be ruled out.

KEYWORDS

left atrial appendage aneurysm, atrial arrthymia ,left atrial appendage thrombus

INTRODUCTION

Congenital left atrial appendage aneurysm are a rare entities and are caused by congenital dysplasia of the atrial muscle.[1].This entitiy is different from the acquired conditions causing enlargement of left atrium like rheumatic heart disease or severe mitral regurgitation.Less than 100 cases have been reported in literature so far. Even though rare,this is not an innocuous condition. Although initially small and asymptomatic ,they are susceptible to grow over years and become symptomatic when they reach enough size .Patients usually present with dyspnoea ,palpitations ,thromboembolic phenomenon in their 3rd decade with slight higher preponderance in females.In asymptomatic individuals the condition is often diagnosed incidentally during cardiac imaging .

We hereby report a case of 30 year old male with a giant LAAA containing thrombus presenting with palpitations diagnosed with echocardiography and cardiac MRI .The patient successfully underwent aneurysmal resection sugery, post which atrial arrhthias stopped and symptoms relieved.

Case Report

A 30 year old male presented to us with complaints paroxysmal palpitations for the past 6months. Physical examination was unremarkable. Ecg on admission showed multiple atrial tachyarrthymia with about 130/min .Chest x-ray done suggestive of mild cardiomegaly with CT raio of 0.6 with abnormally prominent left heart border.(fig1a) .Transthoracic echocardiography demonstrated a giant echo free space adjacent to the posterolateral wall of the left ventricle communicating with the left atrium through a 2cm wide neck. Echo also demonstrated the presence of echogenic mass in the aneurysmal sac i.e thrombus. For further confirmation contrast echocardiography was performed which confirmed the origin of the aneurysmal sac from the Left atrium and also showed areas of no contrast uptake in the aneurysmal sac which confirmed the presence of thrombus.(fig2b).There was no valvular pathology and left ventricular function was normal .LAAA on echocardiography can sometimes be confused with pericardial cyst, mediastinal cystic tumors ,or left atrial herniation through the pericardium. Hence a cardiac MRI was done to confirm the diagnosis and provide further information on the structure of the aneurysm. MRI showed 7x5x4.5 cm aneurysmal dilation of left atrial appendage with 4x4x3 cm filling defect within with no contrast enhancement suggestive of thrombus.(fig2a)

Patient was started on beta blockers , anticoagulation and considered for urgent aneursymectomy to prevent from complications of systemic thromboembolism. Histopathology of the excised specimen confirmed the presence of left atrial appendage tissue with thrombus (fig1b)

DISCUSSION

LAAA is a extremely rare anomaly. LAAA is charaterised by localised outpouching or a diffuse enlargement of left atrial appendage .The first case of LAAA was reported by Parmley et al. in 1962 during a surgical procedure .[2] About 40 % of cases of LAAA are congenital and the rest are acquired in origin. Besides the acquired aneurysms, which are usually associated with LA enlargement due to chronic mitral valve disease (regurgitation or stenosis).[2]. LAAA can be classified into two forms of intrapericardial and extrapericardial. Intrapericardial-type aneurysms are congenital, caused by dysplasia of musculi pectinati and related atrial muscle bands, which they can bulge from the left atrial wall or from the left atrial appendage[3]. Extrapericardial-type aneurysm is associated with a defect in the pericardium resulting the atrial appendage herniating out through the pericardial defect.There is a slight female predominance .Despite the congenital cause ,symptoms usually do not arise until second or third decade of life.[2] The aneurysm probably increase in size as the patient ages .once it reaches a larger ,they predispose the patient to atrial arrthymias, cardiac dysfunction, atypical chest pain episodes due to compression of the left coronary artery or any of its divisions, an increased risk of intra-atrial thrombus formation and systemic embolization[4]. Fibrosis of the endocardium or myocardium is a common histopathological finding in a thin walled aneurysm.[2]. Our patient had episodes of intermittent palpitations and intra-aneurysmal thrombus formation. Therefore, even though rare, if a young patient presents with atrial tachyarrthymia with no other associated cardiac pathologies, a LAAA should be ruled

out [5] Since this condition has been shown to be associated with increased morbidity and mortality. Physical examination usually in unremarkable. ECG shows the presence of atrial arrthymias in most patients. Chest x-ray shows non specific cardiomegaly with an unusually prominent left heart border[6]. A transthoracic echocardiogram or even better, a TEE colour Doppler echocardiogram may suggest the diagnosis by demonstrating the exchange of blood between LA and LAAA . Even contrast echocardiography is helpful to establish the communication between LA and the aneurysm and also to confirm thepresence of thrombus. When echocardiography is unable to visualize the precise extent of the giant LAAA, CT scan and MRI can provide more detailed anatomic visualization, especially in excluding differential diagnoses such as mediastinal or cardiac tumors [7]. Foale and associates proposed the following diagnostic criteria for congenital LAA aneurysm: (i) origin from an otherwise normal LA, (ii) well-defined communication with the LA, (iii) position within the pericardium and (iv) distortion of the LV free wall by the aneurysm [8]. Our patient fulfilled all the four criteria for the diagnosis of an LAA aneurysm and the diagnosis was made during screening echocardiography and MRI. To prevent the occurrence of severe complications, such as myocardial dysfunction, atrial fibrillation and systemic embolism, early surgical intervention is recommended once the diagnosis is established even in asymptomatic cases. Various successful approaches to aneurysmectomy with or without cardiopulmonary bypass have been described, including median sternotomy, left thoracotomy, mini- thoracotomy and endoscope [1, 9] . In our case, the large size of this LAAA made us choose resection via median sternotomy and cardiopulmonary bypass (fig3). This approach provides a clear and motionless field so that we can have complete and accurate resection of the aneurysm to prevent the recurrence of thrombotic events, coronary artery injury and obstruction of the left superior pulmonary vein. The prognosis after surgical treatment is favorable as none of the patients in the systematic review done by Madan raj et al. reported recurrent symptoms, arrhythmia, or thromboembolic events at up to 180 days follow-up. In those not undergoing surgery due to various reasons medical management should be directed to treat atrial tachyarrhythmia and thromboembolic complication[10]

CONCLUSION

Aneurysm of left atrial appendage is rare and often an incidental diagnosis during echocardiography. It is important to recognize this entity, as it is associated with cardiovascular morbidity and mortality by predisposing to atrial tachyarrhythmia and thromboembolism. Surgical resection is the standard modalitiy of treatment even in asymptomatic individuals to reduce risk of associated complications. Medical management is directed at treatment of thromboembolism and atrial tachyarrhythmia.

Conflict of interest : "The author(s) declare(s) that there is no conflict of interest "

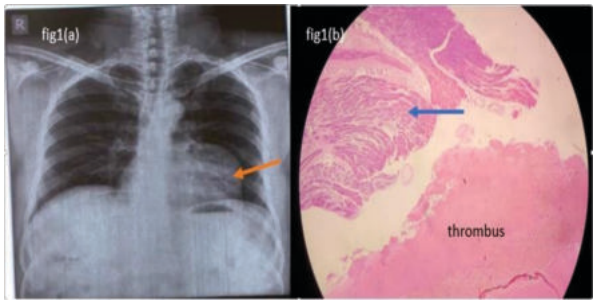

Fig1: (a) X-ray showing prominence of left heart border (arrow) (b) Histopathology section confirming left atrial appendage tissue (Arrow) with thrombus.

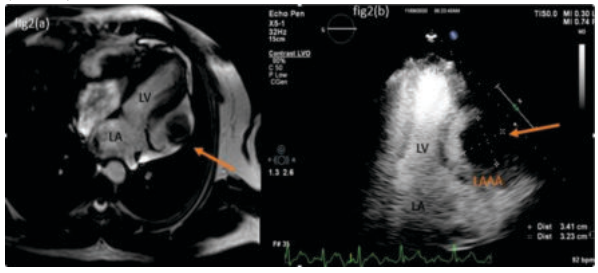

Fig 2: (a) Cardiac Mri showing left atrial appendage aneurysm with thrombus (arrow) (b) Contrast echo demonstrating communication between LA and aneurysm and hypoechoic area within the aneurysm due to absence of uptake of contrast, suggestive of thrombus (arrow) LA: Left atrium, LV: Left ventricle, LAAA: Left atrial appendage aneurysm.

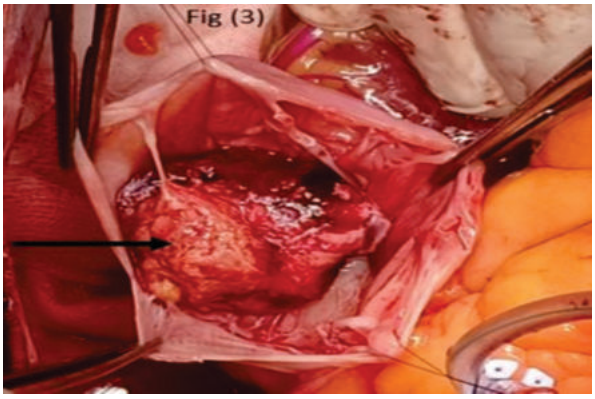

Fig3: Intra-op image showing opened aneurysmal sac with presence of thrombus within it (arrow).

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