

Giant Left Atrial Appendage Aneurysm Presenting As Supraventricular Tachycardia

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Abstract: **Background:** The left atrial appendage aneurysm (LAAA) is extremely rare. It can be caused by congenital dysplasia of the atrial muscles or secondary to mitral valve disease. Most patients are asymptomatic, while palpitations, dyspnea, or chest pain can be found to be main symptoms. **Case:** we describe the case of 18 year old male with congenital aneurysm of left atrial appendage. The patient presented with atrial tachyarrhythmia. The diagnosis was made on transthoracic echocardiography, CECT of thorax and confirmed by histopathological examination (HPE) of excised left atrial appendage aneurysm. **Conclusion:** LAAA is a rare cardiac anomaly. Echocardiography is considered the initial diagnostic tool, and cardiac CT as well as MRI helps to differentiate it from other abnormalities. The associated high risk of life-threatening complications and the relative ease of surgical removal suggest that prompt evaluation should be considered in patients with lesions adjacent to the left heart border.

Keywords: Aneurysm, Left atrial appendage, Arrhythmia

1. Introduction

The left atrial appendage aneurysm (LAAA) is extremely rare. It can be caused by congenital dysplasia of the atrial muscles or secondary to mitral valve disease. Most patients are asymptomatic, while palpitations, dyspnea, or chest pain can be found to be main symptoms.

CASE: 18 year old male presented to our hospital with palpitations (Intermittent) and chest pain for 2 months. Patient did not have history of syncope or presyncope. On Physical examination his heart rate was 200 beats per minute, regular, BP of 90/60. There was no added sounds or murmur. Results of basic laboratory examination including metabolic panel, CBC was normal. 12 lead ECG showed atrial tachycardia with 2: 1 AV conduction. Chest X-ray showed cardiomegaly with Left atrial appendage

enlargement. Comprehensive 2D TTE and Doppler imaging showed normal ventricular size and function. In the apical views, a large, nonmobile, lobular, echo-free cavity measuring 90x60 mm was seen adjacent to the left ventricular anterolateral and left atrial free walls; only mild impingement of the left ventricle was noted. Focused imaging of the left atrium demonstrated the communication between this structure and the left atrium and revealed a giant LAAA. There was presence of spontaneous echo contrast in LAA and LA. CECT Chest showed huge dilatation of left atrial appendage (101x 86x 110mm) suggestive of aneurysm. He underwent surgical excision of LAAA through median sternotomy under cardiopulmonary bypass and was discharged in a stable condition. HPE of surgical specimen showed the wall of the aneurysm composed of myocardium and fibrotic tissue, consistent with LAAA.

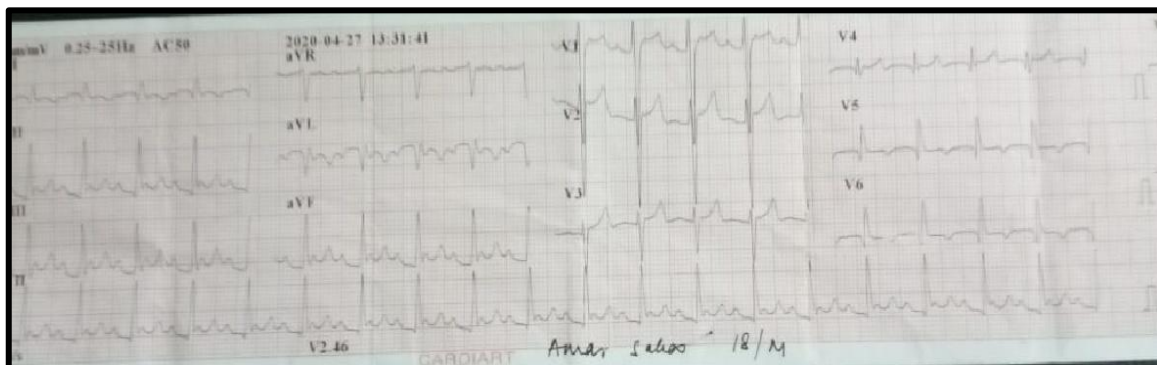


Figure 1: 12 Lead ECG showing Atrial tachycardia with 2: 1 AV block

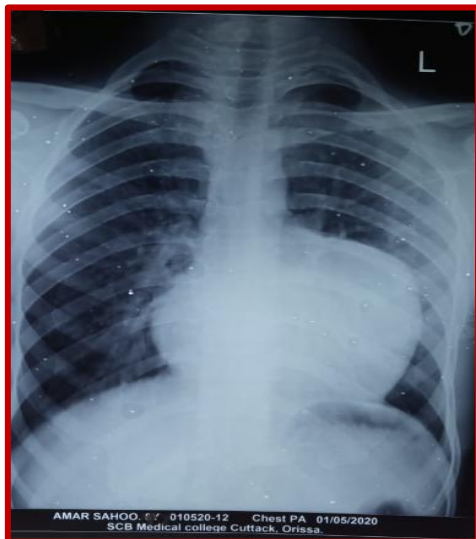


Figure 2: The posteroanterior chest x-ray demonstrating abnormal left heart border in the region of LAA.

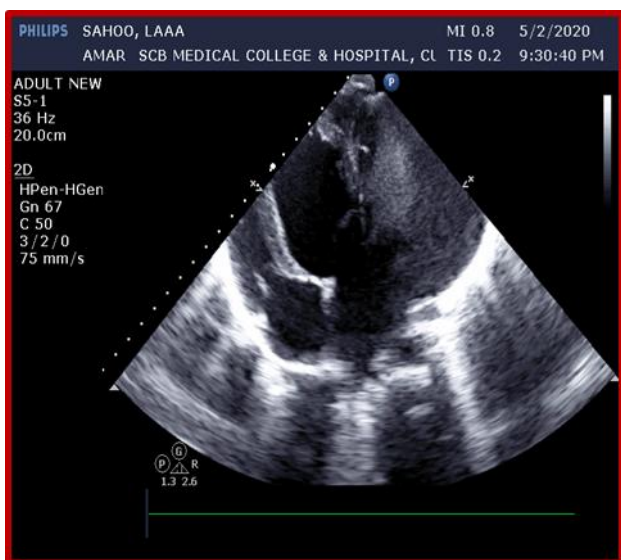


Figure 3: Apical four chamber view showing a large, nonmobile, lobular, echo-free cavity measuring 90x60 mm was seen adjacent to the left ventricular anterolateral and left atrial free walls, communicating with LA suggestive of LAAA. Dense spontaneous echo contrast in LAA seen.

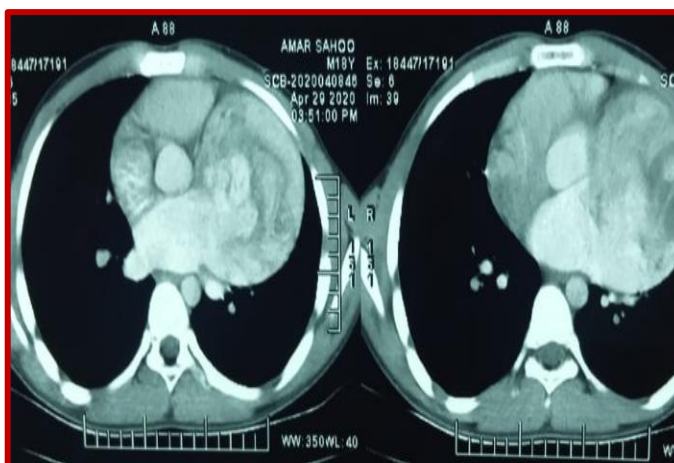


Figure 4: CECT Chest showed huge dilatation of left atrial appendage (101x86x110mm) suggestive of aneurysm.

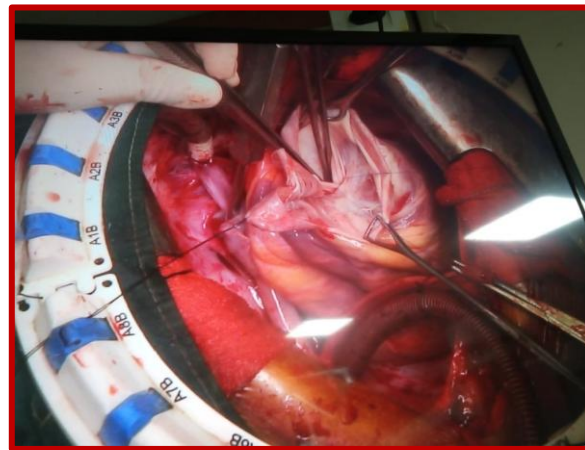


Figure 5: showing Intraoperative LAA after surgical Excision through median sternotomy

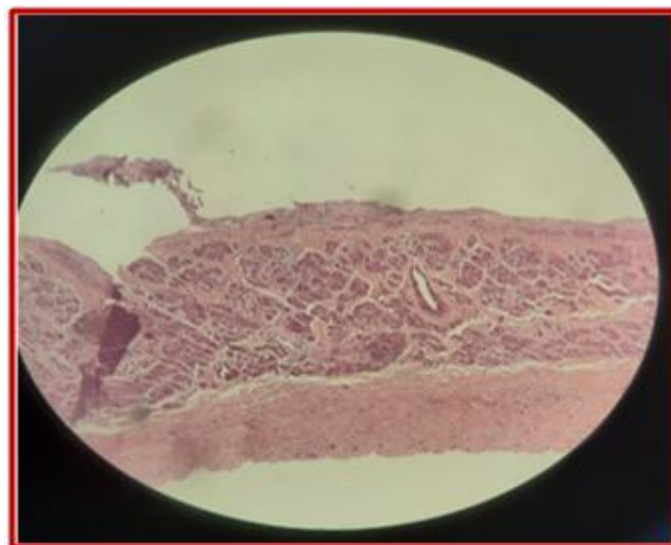


Figure 6: Histology showing the wall of the aneurysm composed of myocardium and fibrotic tissue (hematoxylin and eosin, ×50)

2. Discussion

Epidemiology and pathology

Parmley^[1] first reported LAAA in 1962.^[1] It can occur among patients of all the age groups, with a mean age 30 ± 20 years (range: fetus at 28 weeks to 88 years), although most patients (24.8%, 25/101) are in their third decade. This is probably because of progressive enlargement of the aneurysm with age. Although previous studies reported that there was no gender difference in the prevalence of LAAA, the percentage of female (53/101, 52.5%) patients seems to be slightly higher than that of male (45/101, 44.6%) patients. This is consistent with a previous report by Aryal et al^[2] that included 82 patients LAAA has been classified as congenital or acquired,^[3] with ninety percent of the cases being congenital. The reason is not clear, although the anomaly is probably due to congenital dysplasia of the atrial pectinate muscles.^[4] Acquired LAAA is often secondary to mitral valve disease, or other conditions leading to elevated left atrial pressure.^[5-7] The common histopathological finding of both congenital and acquired LAAA is fibrosis of the endocardium or

myocardium. Hypertrophied myocardium with an increase in interstitial fibrous tissue may be seen in some cases.^[8, 9] Rarely, LAAA has been reported to be associated with congenital cardiac abnormalities,^[7, 10-13] including 5 patients with atrial septal defect, 1 patient with ventricular septal defect, 1 with anomalous pulmonary venous drainage, 1 with mitral valve cleft, and 1 with tricuspid atresia.

Symptoms

The symptoms associated with LAAA often occur during the second to the fourth decades of life. Thirty-four percent (34/101) of patients are asymptomatic and are diagnosed incidentally. On the other hand, among patients with symptoms, the main complaint is palpitation in 45 cases of the patients (44.6%), followed by dyspnea on exertion (28.7%, 29/101) and chest pain (11.9%, 12/101). When the aneurysms reach a larger size, they probably compress the left coronary artery or any of its divisions, leading to myocardial ischemia and atrial arrhythmia. Therefore, patients may experience palpitations, dyspnea, and chest pain. Palpitations on ECG proved to be atrial arrhythmia or supraventricular tachycardia. Electrocardiographically signs of atrial fibrillation/flutter were seen in 27 (26.7%) patients, supraventricular tachycardia in 10 (9.9%) patients, and sinus rhythm in 60 (59.4%) patients. A plausible explanation for the occurrence of supraventricular arrhythmias is the LAAA increases tension on the conduction system or a congenital defect in the conduction tissue exists. Thromboembolic events may also present during the progression of LAAA. The dilatation of LAAA results in the stasis of blood, therefore increasing the risks of thrombi formation and systemic thromboembolism. In our study, 6 patients (5.9%) had systemic thromboembolism events.

Diagnosis

Methods used to diagnose LAAA include a chest x-ray, echocardiography, CT, and MRI. A standard chest x-ray was done in 88 (85.4%) patients. Although the findings were abnormal in all, they were nonspecific and might be interpreted as pericardial cyst, cardiac tumor, or mediastinal mass. Echocardiography is considered to be the primary method of diagnosis. It shows a large sacular structure associated with the left atrium. Eighty-six patients underwent echocardiography. TTE diagnosed LAAA accurately in only 24% (21/86) patients due to limited echo window. Although TTE has a low sensitivity for detection LAAA, it is useful to evaluate left ventricular function, abnormal myocardial movement, and valve regurgitation caused by the compression of LAAA. Besides, it helps to exclude other cardiac abnormalities. TEE provides clear visualization of the structures surrounding atrium, making it effective for the diagnosis of LAAA.

According to our data, TEE identified 30 cases with LAAA among 36 patients who underwent TEE. In the remaining 6 patients, the finding was reported either as enlarged left atrium or pericardial cyst. Due to its precise delineation, TEE should be mandatory if the diagnosis is

ambiguous after evaluation by TTE.^[14] Other imaging modalities, including CT and MRI are useful for diagnosing LAAA as well as ruling out other differential diagnoses. MRI has the highest temporal resolution, making it the optimum approach for assessing the surrounding structures and cardiac anomalies. Ninety-one percent of patients (31/34) with LAAA were identified by cardiac MRI. However, MRI does have certain drawbacks, such as requiring a regular heart rhythm and exposing the patient to nephrotoxic contrast agents. Cardiac CT helps to evaluate the anatomy of coronary artery if compression of left coronary artery or its divisions is suspected. Twenty-seven patients underwent CT scan, and LAAA was detected in 24 cases. However, CT cannot provide functional data as accurately as echocardiography or MRI. Management of LAAA.

Surgical treatment is often recommended even in asymptomatic patients, as it can prevent the potential thromboembolic complications and treat associated atrial arrhythmias. A variety of surgical approaches for surgical resections have been reported.

Median sternotomy aided with cardiopulmonary bypass is the most commonly reported operative approach. This approach has been recommended in patients with intracardiac thrombus, for aortic clamping in this technique prevents systemic embolization during the resection of aneurysm. This approach is suitable for removal of large aneurysms with associated thrombi, but it comes at the cost of increasing invasiveness. Resection through a left lateral thoracotomy has been reported. Compared with the median sternotomy, this technique provides a better visual field and reduces the degree of invasiveness, although this is offset by an increase in procedural operator difficulty.

Seventy-four patients reported in the literatures were surgically treated. Eighty-five percent (63/74) of patients underwent median sternotomy. The size of the aneurysm was highly varied, from 4×3 to 22×15 cm. The average size of LAAA was $11 \pm 5 \times 7 \pm 3$ cm. Thrombi were diagnosed in 17 patients. In contrast, 10% of patients (11/101) without thrombi received the left lateral thoracotomy, with smaller LAAA size (the average size $7 \pm 3 \times 4 \pm 2$ cm). The median sternotomy is considered safer, especially in giant LAAA with intra-aneurysmal thrombi, whereas the left lateral thoracotomy is an option for patients without intra-aneurysmal thrombi seeking for less invasive approach. Both above surgical approaches are considered safe, as no patient died postoperatively. In fact, the prognosis is favorable, for freedom from recurrent symptoms and arrhythmia has been reported from 10 days to 8 years follow-up.^[15-17] Minimal endoscopic techniques have been less frequently described,^[18-20] although the outcomes have also been good. A nonsurgical approach has been reported in 11 patients. The reason included denial of surgery with sinus rhythm ($n = 5$),^[21-25] right femoral artery embolism,^[26] the failure of addressing the occlusion of the right coronary artery in a patient with acquired LAAA,^[27] Eisenmenger syndrome in a patient with atrial and ventricular septal defect,^[7] older age (68 and 76 years, respectively) without intra-aneurysmal

thrombus,^[11, 28] and death secondary to massive cerebral embolism.^[3]

3. Conclusion

LAAA is a rare cardiac anomaly. Echocardiography is considered the initial diagnostic tool, and cardiac CT as well as MRI helps to differentiate it from other abnormalities. The associated high risk of life-threatening complications and the relative ease of surgical removal suggest that prompt evaluation should be considered in patients with lesions adjacent to the left heart border.

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