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Congenital Left Atrial Appendage Aneurysm: A Case Report

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ABSTRACT

Introduction: Left atrial appendage aneurysm is an infrequent abnormality presenting as atrial tachyarrhythmia, progressive dyspnea, atypical chest pain, and systemic thromboembolism. Surgical excision is considered to prevent fatal complications, even in asymptomatic cases. The present study aimed to report a 20-year-old male who presented with sudden onset palpitation and dyspnea.

Case Presentation: The case was really novel, because congenital left atrial appendage aneurysm was accompanied by dilated cardiomyopathy and Left Ventricular (LV) non-compaction. Electrocardiography (ECG) showed atrial fibrillation with Rapid Ventricular Response (RVR) secondary to a massive congenital left atrial appendage aneurysm. At first, left-sided weakness was diagnosed. In the next Computed Tomography (CT) scan performed in the hospital, right temporoparietal hypodensity was detected, which was in favor of right Middle Cerebral Artery (MCA) territory infarction that progressed with hemorrhagic transformation.

Conclusion: The patient did not accept surgical treatment and was discharged with medical therapy (oral anticoagulants and antiarrhythmic medications). He was expired as a result of multiple strokes.

1. Introduction

Left Atrial Appendage Aneurysms (LAAA) without any valvular abnormalities are extremely uncommon. This condition may be caused by hyperplasia of the left atrium wall muscle (1-8). Despite being congenital, LAAA does not present at early ages. Hence, LAAA is usually diagnosed accidentally after the incidence of thromboembolic complications or when some cardiac manifestations such as palpitation, dyspnea, and angina occur (9-17).

2. Case Report

The present case was a 20-year-old gentleman presented with sudden onset palpitation and dyspnea. He came to the hospital and after some initial treatments, he immediately experienced left-sided weakness under the Electrocardiography (ECG) process. He was thus referred to our clinic for further examinations. The results of his physical examination were systolic murmur in the left sternal border, Point of Maximal Impulse (PMI) laterally displaced, elevated Jugular Vein Pressure (JVP), and irregular pulse rate. Additionally, neurological exam revealed left central facial palsy, left-sided hemiparesia (muscle power was one out of five in the left upper and lower extremities), and upward left plantar reflex. Moreover, ECG showed atrial fibrillation with Rapid Ventricular Response (RVR), and heart rate was 145 per minute. At the radiological workup, chest roentgenogram (chest X-ray) showed massive cardiomegaly with an increased cardiothoracic ratio (0.65) without pulmonary hypertension and a prominent convexity of the left border (Figure 1).

Brain Computed Topography (CT) scan showed right temporoparietal hypodensity in favor of the right Middle Cerebral Artery (MCA) territory infarction, which progressed with hemorrhagic transformation in the next brain CT scan performed in the hospital course (Figure 2).

The patient underwent Transthoracic Echocardiography (TTE), which demonstrated the Left Ventricular (LV)

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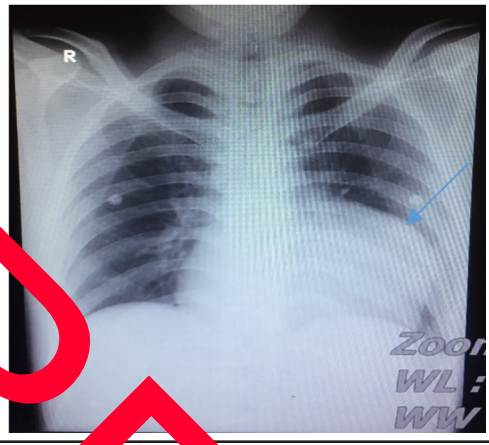

Figure 1. Posteroanterior Chest Radiography. A slightly Prominent Apex Left Heart Border (Blue Arrow) Is Seen.

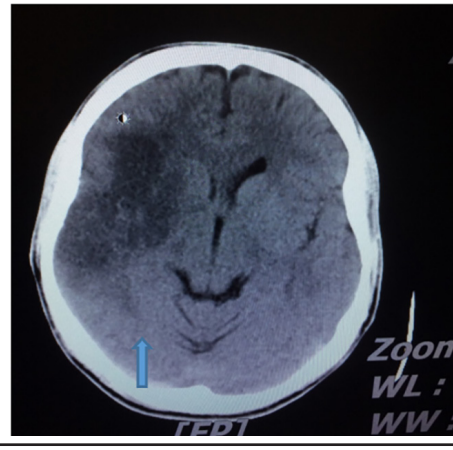

Figure 2. Hypodensity in the Right Temproparietal Lobe in Favor of Infarction (Blue Arrow)

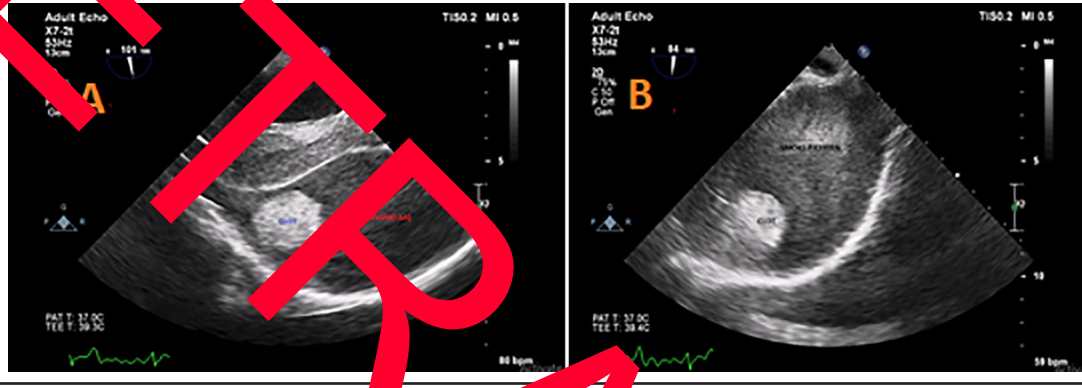

Figure 3. A Giant Left Atrium Aneurysm with Large Organized Clot (A) with a Smoke Pattern (B)

Ejection Fraction (EF) of 40% by Simsom method, diastolic dysfunction, and left-sided pleural effusion. TTE followed by Transesophageal Echocardiography (TEE) revealed a dilated left atrium with smoke pattern, a giant LAA aneurysm with a severe dense smoke pattern, and a large organized clot (Figure 3). He had a dilated left ventricle with global hypokinesia and hyper trabeculae apex, which could be in favor of LV Non-Compaction (LVNC) and EF = 35%.

The patient did not accept surgical treatment and was discharged with medical therapy (oral anticoagulants and antiarrhythmic medications).

Discussion

Congenital giant left atrial appendage aneurysms are extremely rare (2, 18-22). These aneurysms probably increase in size as the patient ages. Once they reach a large size, they enlarge the entire left atrium and can predispose the patient to atrial fibrillation, as in the present case (23-31). A large left atrial aneurysm can also cause symptoms by compressing the heart and resulting in cardiac dysfunction (32-39). Moreover, as shown in the patient reported in the present study, large atrial aneurysms increase the risk of intra-atrial thrombus formation and the possible embolic complications. Therefore, even though rare, if a young patient presents with atrial fibrillation with no other associated pathologies, a left atrial aneurysm should be ruled out. Although these aneurysms can remain silent, they can cause considerable morbidities (40-45). Hence, once they are diagnosed, they should be treated. If they are secondary to another pathological process, they can be resolved once the underlying problem is treated (46-49). 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 (39, 50-53). Up to now, various successful approaches to aneurysmectomy with or without cardiopulmonary bypass have been described including median sternotomy, left thoracotomy, mini-thoracotomy, and endoscopy (54, 55). If they are congenital, however, the only possible treatment is to respect the aneurysm. The present case showed some evidence of dilated cardiomyopathy and LVNC, suggesting a relationship between left atrial appendage aneurysm, and dilated cardiomyopathy and LVNC. Since all these diseases are often genetic condition, all the patients diagnosed with LVNC or dilated cardiomyopathy are recommended to be checked for congenital left atrial appendage aneurysm. Overall, this congenital problem is lethal and physicians should be alert about it.

Although the present case received oral anticoagulants and his International Normalized Ratio (INR) was in the therapeutic range, he experienced multiple clots and expired. Thus, oral anticoagulants should not be administered in these patients and they have to undergo surgical treatments.

Ethical Approval

This study was approved by our ethical committee of Shiraz University of Medical Sciences IR.SUMS.MED.REC.1401.039.

Informed Consent

The "Inform Consent" was obtained.

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There is no acknowledgement.

Authors' Contribution

H[illegible] designed the study, A.M. helped in patient manage[illegible]ent. A.S., R.F., and S.S. drafted the manuscript. A.Z and r[illegible]ised the manuscript

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