A giant left atrial appendage is a rare congenital anomaly that has been reported on only a few occasions. We report two symptomatic patients with atrial fibrillation combined with a cerebellar infarct in one and dyspnea in the other. Both patients were treated surgically with resection of the giant left atrial appendage and radiofrequency pulmonary vein isolation. Recognition of this uncommon pathology can lead to timely surgical intervention.


A giant left atrial appendage is a rare entity described only in case reports. Patients can be asymptomatic or exhibit supraventricular arrhythmias, systemic thromboembolism, cardiac dysfunction, or aspecific chest pain. Because of the risk of life-threatening complications, recognition of this pathology in an early stage is important and can lead to timely intervention.

Case Reports

Patient 1
A 39-year-old man with an unremarkable medical history was admitted to the hospital with an acute, extensive cerebellar infarction and palpitations owing to atrial fibrillation. The cerebellar infarction was treated immediately with thrombolysis. Hemorrhagic complications required two craniotomies, but only mild residual ataxia on the left side and atrial fibrillation persisted.

A chest radiograph showed a convex left upper cardiac figureation. Both transthoracic (TEE) and transesophageal echocardiography (TEE) demonstrated a giant left atrial appendage (LAA), with spontaneous echo contrast and a slightly enlarged left atrium. No thrombi were seen. No other structural or functional abnormalities were observed. On computed tomographic (CT) and magnetic resonance imaging (MRI) the presence of a giant LAA was confirmed.

The patient was referred to our hospital for surgical treatment. He underwent a median sternotomy and bipolar radiofrequency bilateral pulmonary vein isolation. On extracorporeal circulation with cardioplegic arrest and normothermia, the giant LAA was excluded primarily by closure of the appendix with a Blalock suture (Fig 1). Macroscopically, a mass measuring 7 × 7; 2.5 cm was resected. Histologic analysis revealed left atrial appendage tissue with minor fibrosis. The postoperative course was uneventful, and the patient was discharged to his referring hospital 4 d after surgery. Four weeks later, he was still in sinus rhythm and showed good recovery.

Patient 2
A 69-year-old man with a history of a pericardial cyst was admitted to the hospital because of dyspnea, stage New York Heart Association Class II, and permanent atrial fibrillation. He was scheduled for percutaneous pulmonary vein isolation. By routine CT imaging and echocardiography a giant LAA was visualized (Fig 2A, 2B). Therefore, the plan for a percutaneous approach was abandoned, and a surgical resection performed.

The patient underwent surgical resection of the LAA and pulmonary vein isolation. A mass measuring 7 × 7; 5.5 × 7; 2.5 cm was removed, and the histologic analysis showed normal left atrial appendage tissue (Fig 3). The patient was discharged to the referring hospital 5 d after surgery. Despite numerous attempts to restore sinus rhythm by electrical cardioversion and amiodarone (Cordarone), the patient is still in atrial fibrillation.

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Address correspondence to Dr van Herwerden, Department of Cardiothoracic Surgery, E.03.511, Postbus 85500, 3508 GA Utrecht, The Netherlands; e-mail: Lherwerd@umcutrecht.nl.

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Comment

A giant LAA is a rare anomaly, either congenital or acquired, and is described only in case reports [1]. In absence of another pathology, a giant LAA is considered to be a congenital anomaly, probably caused by congenital dysplasia of the atrial pectinate muscles [2]. It is important to distinguish acquired enlargement of the left atrium secondary to mitral valve disease, from pericardial defects with herniation of the left atrium and pericardial cysts. Proposed diagnostic criteria for giant LAA are: (1) origin from an otherwise normal left atrium, (2) well-defined communication with the left atrium, (3) position within the pericardium, and (4) distortion of the left ventricle free wall by the aneurysm [3]. These two cases fulfilled all these criteria.

Approximately 100 cases of the giant LAA are reported in the literature, which underscores the rarity of this anomaly. Usually this condition is asymptomatic; however with aging of the patient it is likely that these aneurysms increase in size, thereby predisposing the patient to supraventricular arrhythmias, cardiac dysfunction, atypical chest pain caused by compression, and an increased risk for thromboembolic complications [1]. To prevent these severe complications, it is important in young patients with atrial fibrillation to rule out a coexisting giant LAA.

Imaging studies are necessary to diagnose a giant LAA. A chest radiograph can may show nonspecific cardiomegaly with a prominent left cardiac border. TTE and eventually TEE are sufficient to recognize the giant LAA, reduced atrial blood flow, and thrombi. MRI and CT can be helpful to confirm the diagnosis and clarify relations with surrounding tissue or concomitant pathology [1, 4].

Surgery is also recommended in asymptomatic patients to prevent atrial fibrillation and thromboembolic complications. The preferred approach is a median sternotomy for adequate exposure. Cardiopulmonary bypass provides safety of excision in a motionless field and decreases the risk of embolization during manipulation or in the presence of intraaneurysmal thrombi [1, 5].

Although resection alone is usually adequate, in some cases, especially with biatrial enlargement, an additional Cox-Maze III procedure or pulmonary vein isolation is performed [6]. Hof et al. [7] reported a case of a patient with a giant LAA and atrial fibrillation who was treated successfully with pulmonary vein isolation [7]. However, the causality between giant LAA and atrial fibrillation has not yet been proved.

This case report describes two patients with a congenital LAA typically presenting with atrial fibrillation (Patients 1 and 2) and thromboembolic complications (Patient 1). These patients were treated with surgical resection (and ablation).

References

Nontraumatic Ascending Aortic Disruption

Richard Y. Highbloom, MD, Yosef Y. Schwartz, Michael Rosenbloom, MD, Frank W. Bowen, MD, and Neeta Datwani, MD

Division of Cardiotoracic Surgery, Department of Surgery, and Division of Cardiology, Department of Medicine, Cooper University Hospital of Rowan University, Camden, New Jersey

This report documents a case of completely spontaneous ascending aortic disruption. A 54-year-old African American male day laborer presented with severe retrosternal chest and back pain and shortness of breath. He had no history of hypertension, smoking, or trauma and was taking no medications. The computed tomographic angiography scan performed to exclude pulmonary embolism instead demonstrated a hemorrhagic pericardial effusion and an ascending aortic pseudoaneurysm. He was taken emergently to the operating room for repair of his ascending aorta. The histopathology report was normal.

Comment

Truly spontaneous aortic disruptions that arise without any predisposing factors—chronic hypertension, sudden trauma, heritable connective tissue disorders or other cardiovascular diseases—are extremely rare. The exact number of genuinely spontaneous, nontraumatic aortic disruptions is difficult to quantify primarily because the term "spontaneous" is loosely applied and is ill defined in the surgical literature. Although just over 30 cases of spontaneous aortic ruptures have been reported (using the most liberal definition) [1], the majority of were attributed to atherosclerosis exacerbated by chronic hypertension and other cardiac ailments [2,3]. The actual number may be as small as 11 previously recorded cases, all of which had a history of hypertension; five had cystic medial necrosis, three had atherosclerosis, and the histology of the others was unknown [4].

One case of truly spontaneous aortic rupture stands out [2]. Similar to our event, a previously healthy patient with no history of hypertension, trauma, or other cardiac symptoms was admitted following sudden collapse and shock. A transesophageal echocardiogram was performed. An aortic rupture was found and A chest radiograph was unremarkable. Chest computed tomographic angiography performed to exclude pulmonary embolism instead demonstrated a hemorrhagic pericardial effusion and an ascending aortic pseudoaneurysm (Fig 1). He was taken emergently to the operating room for repair of his ascending aorta.

Intraoperative transesophageal echocardiography revealed early cardiac tamponade, but failed to identify the etiology conclusively. At surgery, the findings confirmed the radiologic diagnosis of ascending aortic rupture, contained only by adventitia (Fig 2). There was no dissection or valvular involvement (Fig 3). Ascending aortic replacement was performed with a 28 mm woven Dacron graft placed in the supracoronary position under cardiopulmonary bypass and aortic crossclamping. His postoperative course was unremarkable, and he was discharged home on postoperative day 4.