

Huge Interatrial Septum Aneurysm

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Abstract Atrial septum aneurysm (ASA) is a saccular aneurysm of the interatrial septum, bulging into either atrium during the cardiac cycle. It is mostly congenital in nature and is often associated with patent foramen ovale (PFO). ASA is increasingly gaining attention as a possible cause of cryptogenic stroke. We here present a case of ASA in an elderly female in whom ASA possibly contributed to atrial fibrillation and mitral valvular prolapse; she denied any thromboembolic event. Transesophageal echocardiography remains the imaging modality of choice for diagnosis of ASA. Management guidelines for initiation of antiplatelet therapy, anticoagulation and the need for endovascular or surgical closure of associated PFO are lacking.

Keywords: atrial septum aneurysm, patent foramen ovale, stroke, transesophageal echocardiography

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1. Introduction

Atrial septum aneurysm (ASA) is a rare clinical entity where interatrial septum bulges as a saccular aneurysm into either or both the atria during cardiac cycle. Most of the reported ASA are congenital, though acquired cases have been reported. ASA is often associated with other congenital cardiac abnormalities including patent foramen ovale (PFO). ASA is attributed in cardiogenic stroke, cardiac arrhythmias, migraine and mitral valvular prolapse. We here present a case of 95 year old lady with huge type 1R ASA with PFO that was incidentally detected on transthoracic echocardiography. Atrial fibrillation in the patient be partly attributed to ASA; she never had any cardioembolic episodes. Transesophageal echocardiography is the imaging modality of choice for the diagnosis of ASA. Guidelines on management of ASA with or without complications are lacking. Use of antiplatelet therapy with or without anticoagulation (warfarin) and surgical closure or endovascular closure of an associated PFO are the available treatment options.

2. Case Presentation

A 95 year-old-women with a past medical history of hypertension, hypothyroidism, allergic rhinitis, Alzheimer's dementia, pulmonary hypertension, atrial fibrillation, mild to moderate mitral regurgitation and mild tricuspid regurgitation presented with worsening bilateral lower extremity swelling. History of present illness was limited by patient's dementia. On arrival she was not in any

distress, vitals were stable and physical examination was significant for systolic murmurs in mitral and tricuspid area, rales heard in infrascapular area bilaterally, lower extremity edema till knees associated with scaling and hyperpigmentation of the knees were noted. Laboratory investigations were significant for an elevated D-Dimer level (3630 ng/ml) a computed tomography angiography of the chest ruled out pulmonary embolism and venous duplex ultrasonography of bilateral lower extremity ruled out deep vein thrombosis. Chest radiography was significant for cardiomegaly and pulmonary congestion. EKG revealed normal sinus rhythm and left bundle branch block. Troponin levels trended were not elevated. A transthoracic echocardiography obtained revealed a giant interatrial septal aneurysm within the right atrial cavity (**Figure 1**). Following injection of agitated saline contrast via a peripheral vein, a moderate to large number of microbubbles entered the left atrium, consistent with the presence of a right to left shunt across a patent foramen ovale and/or small atrial septal defect (**Figure 2**). Colorflow Doppler evaluation revealed a trivial left to right shunt across a tiny secundum defect. Diffuse left ventricular hypokinesia with an ejection fraction of 25-30%, right atrium and right ventricle were dilated and pulmonary artery systolic pressure was elevated (60 mm of Hg).

3. Discussion

Atrial septum aneurysm (ASA) is a rare cardiac clinical entity that was first reported by Lang and Posselt in 1934. ASA is characterized by bulging of interatrial septum into either or both atria during the cardiac cycle [1]. The

prevalence of ASA in autopsies is 1% and the prevalence in transesophageal echocardiography studies in unselected patients ranges from 2 to 10% [2]. Most of the reported cases of ASA are detected incidentally and majority of the reported cases are congenital in nature. Though rare, increase in atrial pressures can result in development of

ASA [3]. ASA is associated with congenital cardiac abnormalities such as patent foramen ovale (PFO) [4], atrial septal defect [3], mitral valvular prolapse [5,6], tricuspid valvular prolapse, marfan's syndrome, sinus of valsalva aneurysm and aortic dissection [7].

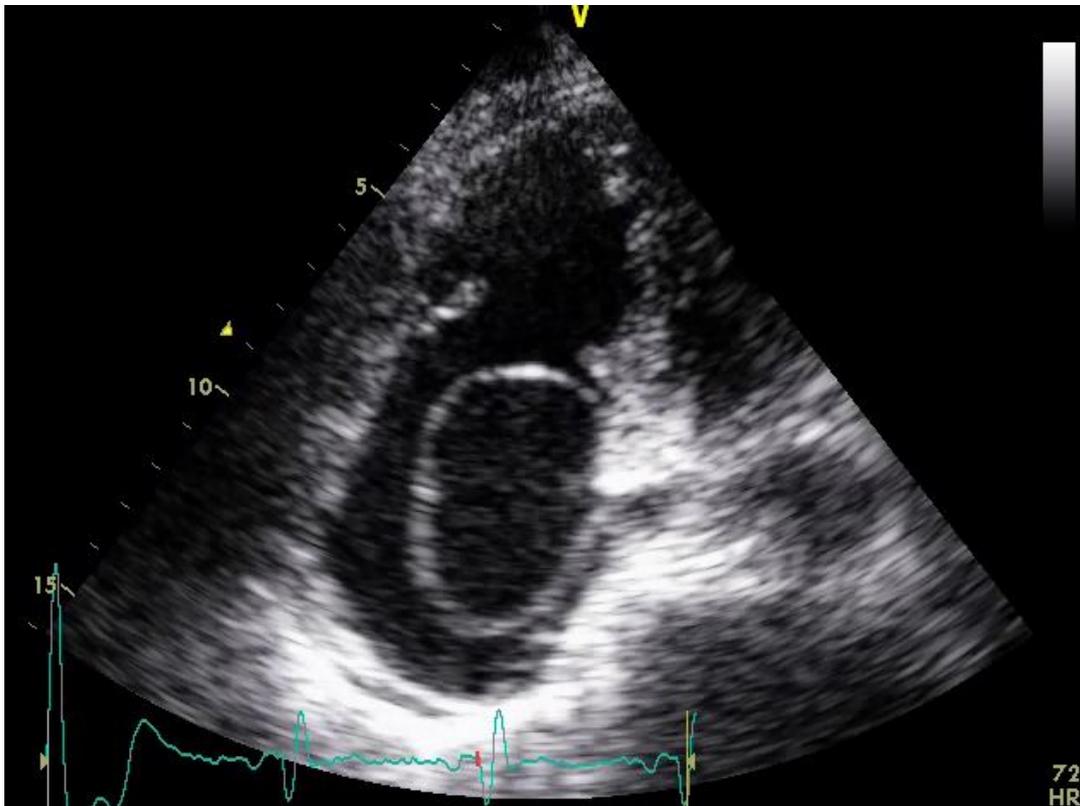


Figure 1. Large ASA bulging into right atrium (type 1R) as seen in transthoracic echocardiography

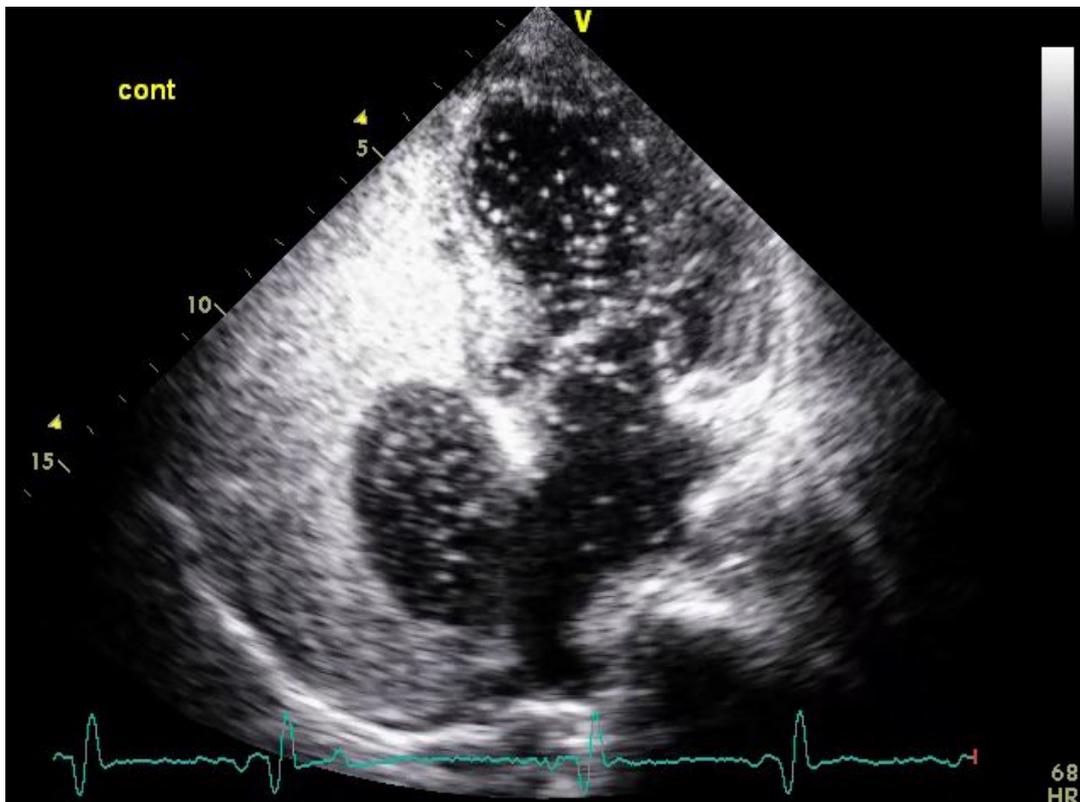


Figure 2. Microbubbles entered the left atrium, consistent with the presence of a right to left shunt across a patent foramen ovale and/or small atrial septal defect

Most reported cases of ASA are incidental findings [3]. ASA is increasing gaining clinical significance as it is attributed cardio-embolic stroke [8], atrial arrhythmias, ventricular arrhythmias [9,10], and migraines with aura [11] and MVP [5,6]. 70% of the patients with ASA have patent foramen ovale and the two together increase the risk of stroke by 33-folds [12]. Despite aspirin therapy patients with both ASA and PFO are at increased risk of recurrent stroke as compared to patients without either of these abnormalities [13]. The mechanisms that lead to increased risk of stroke in patients with ASA are: i) paradoxical embolism a venous thrombus through an associated PFO, ii) embolism of thrombus formed in ASA, iii) associated atrial arrhythmias that increase the risk of clot formation [14] and iv) any perforations that develop in ASA can result in paradoxical embolism [15]. ASA >5mm in thickness have a greater association with stroke; thickness in such cases represent thrombus in ASA [8]. Atrial repolarization delay due to ASA is attributed in the development of atrial fibrillation [16]. Patients with ASA are known to be at increased risk of ventricular tachycardia, though it is not clear if such arrhythmia are solely due to ASA or associated cardiac abnormalities [8]. The increased risk of migraine in patients with ASA is attributed to shunting of vasoactive substances from venous circulation to systemic arterial circulation through PFA [17]. The redundancy of subendocardial tissue in ASA also partly explains its association with MVP [10].

Echocardiography is the imaging modality of choice for ASA diagnosis; transesophageal echocardiography is more sensitive than transthoracic echocardiography [18]. Computed tomography [19] and contrast enhanced dynamic magnetic resonance imaging with valsalva [20] are other imaging modalities used in diagnosis of ASA. Based on the direction and movement of ASA Olivares-Reyes et al. proposed a new classification, the same has been tabulated in Table 1 [2]. Clear guidelines on regarding management of ASA are lacking; incidentally found ASA without any symptoms do not need any treatment. Though debate continues on antiplatelet therapy with aspirin alone versus oral anticoagulation therapy with warfarin in patients with PFO, no significant difference in primary end points of recurrent stroke or death at the end of 2 years was observed in aspirin alone versus warfarin alone treated groups [21]. Newer percutaneous transcatheter closure devices have a PFO closure rates ranging from 65 to 100% [22,23]. Following the placement of percutaneous transcatheter closure device the risk of recurrent embolic or neurological event lasted from 0 to 3.8% per year [24]. Conflicting results exist regarding the efficacy of surgical closure of PFO. A meta-analysis established superiority of PFO surgical closure over antiplatelet therapy alone [25]. Another study established the safety of PFO surgical closure but it did not establish the superiority of the same over medical therapy [26]. Devuyt et al. in their study demonstrated that in patients with ASA with PFO surgical closure is beneficial; no recurrence of stroke or transient ischemic attack was noted at the end of two year follow up period during which patients were not on antithrombotic therapy [27].

The patient we present here had 1R type of ASA. She had no prior thromboembolic events or migraine. It is unclear if the mitral regurgitation is an incidental finding

or if the same led to elevated left atrial pressures and hence ASA. Also atrial fibrillation may be comorbid condition or secondary to ASA.

Table 1. Olivares-Reyes et. classification of ASA based on the direction and movement of ASA

ASA type	Description
Type 1R	Bulging of ASA into right atrium only
Type 2L	Bulging of ASA into left atrium only
Type 3RL	Major excursion bulges into the right atrium and the lesser excursion bulges towards the left
Type 4LR	Major excursion bulges into the left atrium and the lesser excursion bulges towards the right
Type 5	ASA movement is bidirectional and equidistant to both atria during the cardiorespiratory cycle

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