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Vital Importance of Delineation of Coronary Artery Anatomy in Atypical Congenital Giant Right Atrial Aneurysm

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Abstract

Introduction: Giant congenital right atrial aneurysm is a very rare congenital heart lesion, which may be asymptomatic or present a variety of symptoms, particularly supraventricular arrhythmias and intracardiac thrombosis formation.
Case Presentation: This is a report on a 3.5-month-old male infant with imperforated anus and an unusual-shaped congenital giant right atrial aneurysm with retro-ventricular extension. This unusual shape prevented appearance of cardiomegaly on the chest X-Ray. Surgical resection of the aneurysm was attempted. However, posterior descending coronary artery, which was embedded in the wall of the aneurysm, was irreversibly damaged during the operation. The patient died in the operation room. We concluded that pre-operative delineation of coronary arteries in cases with congenital giant right atrial aneurysm (CGRAA) with extension to the posterior left ventricle is mandatory. Despite the current data that surgical excision of the aneurysm is the treatment of choice, our case required simple closure of the aneurysmal neck from inside the right atrium to be an easier and safer surgical approach for treatment of CGRAA with a tricky anatomy.
Conclusions: This case indicates that delineation of coronary artery anatomy in atypical congenital giant right atrial aneurysm is of vital importance. Closure of the aneurysmal sac, instead of aneurysmal resection, is a safer and more simple approach in atypical cases.

Keywords: Congenital Giant Right Atrial Aneurysm, Coronary Artery Anatomy

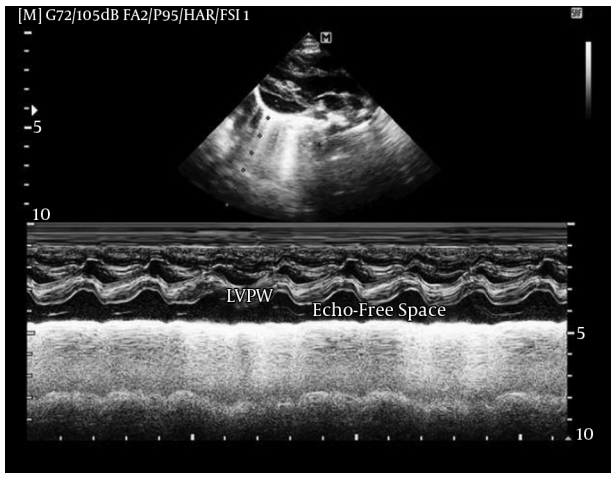
1. Introduction

Congenital giant right atrial aneurysm (CGRAA) is a very rare disease with diverse clinical manifestations. Surgical excision of the aneurysm is the treatment of choice (1, 2). To the best of our knowledge, to date, no CGRAA with retro-left ventricular extension involving coronary artery has been reported. We report a case with an unusually huge congenital right atrial aneurysm with extension to the posterior aspect of the left ventricle with subtle adherence of the posterior descending coronary artery to its wall.

2. Case Presentation

A 3.5-month-old infant with the diagnosis of imperforated anus and hypospadias was referred to our clinic with a history of colostomy in the neonatal period. On physical examination, he was acyanotic with normal cardiac auscultation. Electrocardiogram and chest X-Ray were normal. On echocardiography, an echo-free space was seen behind the left ventricular posterior wall, both in the parasternal long-axis and modified four-chamber views (Figure 1 echo videos 1 and 2 in the supplementary file). There were tissue strands within this space, mimicking the presence of purulent pericardial effusion. Lowering the Nyquist frequency revealed blood flow in this huge effusion-like, echo-free space (Figure 2). Thorough examination showed an unusually shaped congenital giant right atrial aneurysm (CGRAA), with an oblique lie, extending from the right side and anterior of the heart to the left and posterior of the left ventricle (Videos 3 - 5 in the supplementary file). The infant experienced episodes of witnessed transient supraventricular tachycardia during echocardiographic examination and afterwards during admission. He underwent surgical operation for resection of aneurysm (Video 6 in the supplementary file). Immediately, after weaning from cardiopulmonary bypass (CPB), he developed hypotension. The CPB was re-established. Posterior descending coronary artery (PDA) was damaged and the patient died in the operation room.

Figure 1. M-Mode of Parasternal Long-Axis View



An echo-free space mimicking a pericardial effusion is seen posterior to the left ventricular free wall. This is in fact, the retro-left ventricular segment of the Congenital Giant Right Atrial Aneurysm (CGRAA).

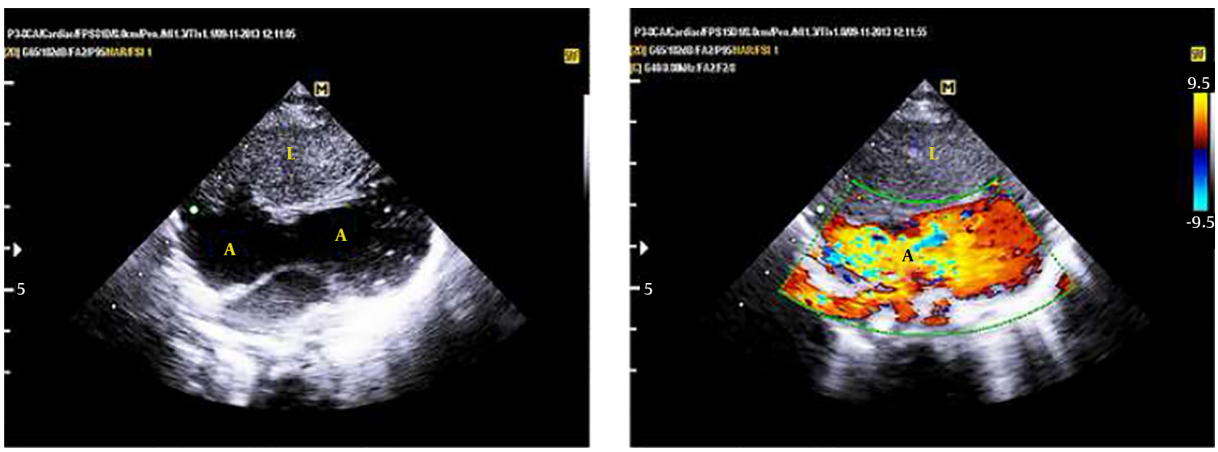


Figure 2. Subcostal two-dimensional echocardiogram on the left shows the huge CGRAA, with an oblique lie from right and anterior to left and posterior of the heart. The right image shows the appearance of blood flow within the aneurysm by lowering the Nyquist frequency to about 10 cycles/second.

3. Discussion

We performed a literature review on congenital right atrial aneurysm from 2001 to 2014 (Table 1). Fifteen cases, with age ranging from 20 weeks of gestation to 75 years, are reported from eight countries. However, none of them had the unusual shape and geometry of CGRAA seen in our case. Furthermore, in none of these cases, the coronary artery was attached to the aneurysm.

Congenital giant right atrial aneurysm is a very rare cardiac anomaly and right atrial aneurysms are rarer than the left (3, 4). Yildirim et al. reported 18 cases up to 2006, whereas Chatrath et al. reported 60 cases up to 1998 (4, 5). Klisiewicz et al. reported only seven cases up to 2004 (6). About half-of the reported cases were asymptomatic (7-9). Clinical presentation included arrhythmia, thrombosis, heart failure and incidental cardiomegaly on chest X-ray (10-13). In a recent review by Harder et al., all cases underwent resection of aneurysm (13).

Normal P wave on Electrocardiography (ECG), despite huge enlargement of right atrium (RA) (1), can be explained by the absence of myocardium in the aneurysmal wall.

We report on an infant with an unusual-shaped CGRAA and imperforated anus, leading to death because of injury to the posterior descending coronary artery (PDA) that was

Table 1. Literature Review on Congenital Right Atrial Aneurysm From 2001 to 2014

	Authors	Year	Country	Number of Cases	Type of Article	Age at Diagnosis	Gender	Age at Operation	Treatment	Outcome	Presentation	Duration of Follow-Up
1	Harder et al. (13)	2014	USA	5	Case series and review	26 months	Female	26 months	Resection	good	Supraventricular tachycardia	4 m
						20 weeks (prenatal)	Not defined	4 months	Resection	good	-	3 years of age
						Prenatal	Not defined	15 months	Resection	good	-	4 years
						18 weeks (prenatal)	Not defined	6 weeks	Resection	good	-	2 years
						5	Male	5 years	Resection	good	Incidental finding of cardiomegaly	10 years
2	Uppu et al. (12)	2013	USA	1	Case report	2	Male	2 years	Resection (surgical reduction)	good	Incidental finding of massive cardiomegaly	Not defined
3	Narain et al. (11)	2012	India	1	Case report	18	Male	18 years	Medical treatment	good	Right heart failure	Not defined
4	Zaqout et al. (14)	2011	Belgium	1	Case report	7 days	Male	Not defined	Resection	good	Cardiac murmur	Not defined
5	Lee et al. (15)	2011	Korea	1	Case report	2 weeks	Not defined	2 weeks	Resection	good	Referral case	Not defined
6	Zhao-xia et al. (16)	2010	China	1	Case report	38	Male	38 years	Resection	good	Paroxysmal chest pain	Not defined
7	Agematsu et al. (17)	2009	Japan	1	Case report	31	Male	31 years	Resection	good	Cardiomegaly on chest X-Ray(CXR)	Not defined
8	Yoon et al. (8)	2009		1	Case report	69	Female	-	-	good	Incidental finding of cardiomegaly on CXR	Not defined
9	Yildirim et al. (4)	2006	Turkey	1 (authors state 18 cases were reported to 2006)	Case report	42	Female		Medical treatment (anticoagulation therapy)	good	Palpitation, dyspnea and precordial pain	Not defined
10	Klisiewicz et al. (6)	2004	Poland	1 (authors state 7 cases were reported to 2004)	Case report	30	Female	30 years	Resection	good	Abnormal cardiac silhouette	Not defined
11	Chatrath et al. (5)	2001	USA	1 (authors state 60 cases were reported to 1998)	Case report	5	Male	5 years	Resection	good	Incidental finding of cardiomegaly	1 year

attached to the aneurysmal wall. This case indicates the importance of careful preoperative delineation of coronary arteries in patients with CGRAA with a retro-left ventricular extension. Resection of aneurysm is not the optimal treatment for all cases of CGRAA. Closure of the neck of the aneurysm from the inside of the right atrium is recommended in cases with this special anatomy, as in our case.

3.1. Conclusion

This case was the first reported case of CGRAA associated with imperforated anus and retro-left ventricular extension of the CGRAA with posterior descending coronary artery embedded in the wall of the aneurysm. Our patient died due to damage to PDA. Careful delineation of coronary artery anatomy by either coronary computerized tomography (CT) angiography or conventional coronary angiography, is mandatory in these cases. Simple closure of the neck from inside RA seems to be a safer surgical approach.

Supplementary Material

Supplementary material(s) is available here.

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Footnotes

Authors' Contribution: Both authors contributed to this manuscript during all phases.

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