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Case Report

Coronary Artery Fistula Associated with Mitral Valve Regurgitation in a Young Symptomatic Patient: A Case Report

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Abstract

Congenital coronary artery fistulas are rare cardiac defects. A fistula associated with other cardiac anomalies, like valvular heart disease, is an extremely rare condition. We report a young symptomatic patient who presented with a continuous murmur heard along second right intercostal space and a systolic murmur at the apex on clinical examination. Chest x-ray showed left ventricular prominence and transthoracic echocardiography with doppler studies showed right coronary fistula draining into the right atrium and moderate mitral regurgitation. She is being followed up with medical management at the outpatient department. We recommend coronary angiography with cardiac catheterization, and if patient will give her consent, surgical repair of the fistula with possible mitral valve surgery is recommended. We conclude that diagnosis of coronary artery fistula should be considered when patient presents with continuous murmur. A non-invasive test, like transthoracic echocardiography with Doppler studies, can demonstrate dilated coronary arteries and their receiving chambers or vessels.

Keywords: Coronary Artery Fistula; Coronary Artery Malformation; Coronary Artery Anomaly; Congenital Heart Disease; Continuous Murmur

Introduction

A coronary artery fistula (CAF) is an abnormal communication between a coronary artery and a cardiac chamber, great vessel, or other vascular structure [1]. The incidence of CAF is estimated at 1 in 50,000 live births, and it is detected in approximately 0.2% of the adult population during coronary angiography [2]. The most common origin is the right coronary artery (RCA) and a single termination is found in the majority of patients.3 The right ventricle is the most common site of drainage (45%) followed by the right atrium (25%), pulmonary artery (15–20%) and coronary sinus (7%) [3]. These can be symptomatic or asymptomatic because the hemodynamic consequences of the fistula vary and depend on the shunt dimensions.4 In general, symptoms of angina, palpitations and cardiac failure may occur in patients aged over 30 years, with the cardinal clinical finding of a continuous murmur similar to a patent ductus arteriosus (PDA) [3] but the diagnosis of CAF may be suggested by the finding of a continuous murmur in a precordial location, which is atypical for PDA [1].

Doppler echocardiography is currently used as the noninvasive method to establish the diagnosis and evaluate management of CAF [3]. Magnetic resonance imaging has become an alternative method to evaluate anatomy, flow and function [3]. In most instances the diagnosis is made during heart catheterization for coronary or congenital heart disease [5]. Cardiac catheterization is usually performed in order to confirm anatomy and plan surgical treatment [3].

Gurbuz., *et al*. reported a case of CAF with mitral valve stenosis and they mentioned that reports of the coincidence of mitral stenosis and CAF are rare in the literature. A fistula opening into the right atrium is rare in patients with coronary artery anomalies and mitral valve disease.[2] Now, for its rarity, we report a case of a symptomatic young patient with concomitant mitral valve regurgitation and CAF, with the fistula draining into the right atrium.

Case

A 25 years old female reported dyspnea and easy fatigability for 1 year already. There was no cough, fever, chest pain, orthopnea or edema. Symptoms progressed and she also noted palpitations, so she sought consult and was given unrecalled medications. 2D echocardiography was advised and she was told to have a 'hole in the heart'. Symptoms persisted, thus, she was referred to Philippine Heart Center. Medical or surgical history was unremarkable, as well as, family history. On physical examination, she had dynamic precordium, displaced apex beat, continuous murmur at second right intercostal space radiating to the mid right parasternal border area and grade 3/6 systolic murmur at apex radiating to the back.

We proceeded with diagnostic evaluation. 12-lead electrocardiogram (ECG) was normal. Chest x-ray revealed an enlarged heart with prominent aorta, prominent main pulmonary artery and increased pulmonary vascularity (Figure 1A). On lateral view, it showed LV prominence (Figure 1B). 2-dimensional transthoracic echocardiography (TTE) with Doppler studies showed a dilated RCA draining into the right atrium, with noted continuous turbulent flow most likely the fistulous connection, and a mosaic color flow display across the mitral valve during systole denoting mitral regurgitation. (Figure 2. A, B and C).

She was managed medically on an outpatient basis. Further diagnostic tests were discussed and if patient will give her consent, surgical repair of the CAF and possible mitral valve surgery is being contemplated.

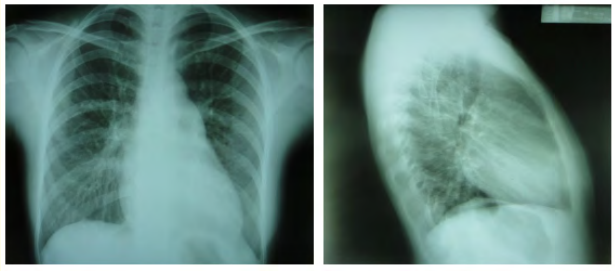

Figure 1: A. Enlarged heart with a CT ratio of 0.52 but right atrium is not enlarged and right cardiac border is not prominent with the lateral border of the cardiac silhouette measuring 2 cm from the lateral border of the spine. The aorta and MPA are prominent. The lungs are hypervascular. **B.** Right ventricle is not prominent. Hoffman-rigler measurement is 1.6cm indicative of LV prominence.

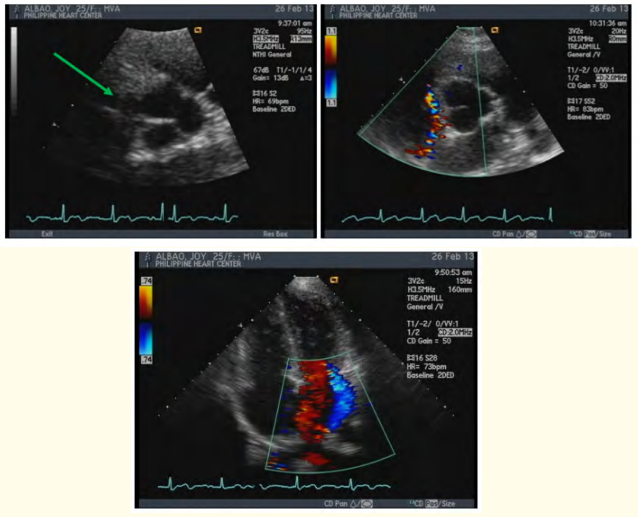

Figure 2: A. Dilated right coronary artery (green arrow). **B.** Continuous turbulence noted from the right coronary artery to superior aspect of the right atrium denoting the fistulous connection. **C.** Mosaic color flow display noted across the mitral valve during systole denoting mitral regurgitation.

Discussion

Congenital CAFs, a subgroup of anomalies of the coronary arteries, are an extremely rare cardiac defect [6]. It is an infrequent but potentially important abnormality that can affect any age group [1]. In an article by Chu., et al. the ages of the patients, who underwent surgical treatment for CAF, ranged from 4 months to 50 years [7]. Our case is a 25 years old female which falls near the mean age of this report, which is 22.7 years. In general, symptoms are rare under the age of 20 years [3]. The pathophysiologic mechanisms resulting in symptoms include cardiac volume overload due to the shunting of blood and reduction of the myocardial blood supply due to "coronary steal."1 Dyspnea and chest pain represented a frequent (71%) clinical symptom in CAFs in adults while in the pediatric age group, the majority was silent (79%) and dyspnea and chest pain accounted for only 8% of the symptoms [8]. Liu and his colleages reviewed fourteen patients with congenital CAF and only one was asymptomatic, but diagnosis was suspected from clinical presentation of a continuous murmur. In the symptomatic patients, angina and exertional dyspnea were the most common symptoms [9]. Also, Hong and his colleagues reviewed fifteen cases of congenital CAF, and twelve patients were symptomatic at the time of the diagnosis [10]. Parga., *et al*. mentioned the cardinal clinical finding of continuous murmur, usually heard at the middle left or right sternal border or even at the lower sternal border. Even Chu and his colleagues mentioned that continuous heart murmurs was heard in all the cases they reviewed except one. Our patient presented with dyspnea and easy fatigability since she was 24 years old, consistent with these reports (3, 8, 9, 10) and she had a continuous murmur at the 2nd right intercostal space, radiating to the mid right parasternal border area which is also consistent with these reports (3, 7).

The diagnosis may be established non-invasively by echocardiography, demonstrating the dilated coronary artery and the fistula (including its entry site into the chamber or vessel) [3]. But according to Bauer., *et al*. in most instances the diagnosis is made during heart catheterization for coronary or congenital heart disease. Likewise, Said and his colleagues stated that the diagnostic modalities for CAF, from the literature between 1993 and 2004, were mainly cardiac catheterization and coronary angiography. However, the CAF in our case was demonstrated by TTE, as a dilated RCA draining into the superior aspect of the right atrium (RCA fistula).

Several reports reviewing congenital CAF cases have different views regarding the origin and drainage of CAF. Bauer., *et al*. showed that there was equal affectation of the right and left coronary artery (LCA). According to Tirilomis and his colleagues, CAF originated mostly from the left coronary system (proximal left descending artery, left main stem, circumflex artery) which was also the same with the reports of Liu., *et al*. and Abdelmoneim., *et al*, showing the left anterior descending coronary artery (LAD) as the most commonly involved. In contrast, the findings of Parga., *et al*. revealed that 55% to 65% of congenital CAFs arise from the RCA. The right ventricle is the most common site of drainage (45%) followed by the right atrium (25%), pulmonary artery (15–20%) and coronary sinus (7%) [3]. This report regarding drainage of CAF was also affirmed by the article of Chu., *et al*. which mentioned that all the drainage sites of the CAF were on the right side of the heart (right atrium, right ventricle, pulmonary artery) as well as these other reports (9, 10, 11). Our finding of RCA fistula proves that RCA is still the most common origin of CAF, as mentioned by Parga., *et al*. Also, its drainage to the right atrium is consistent with these previous reports (3, 7, 9, 10).

Meanwhile, Chu., *et al*. noted that only one of the patients with CAF had associated cardiac disease while the report of Hong., *et al*. showed that six patients with CAF had associated cardiac anomalies. Gurbuz and his colleagues reported a rare case of CAF and mitral valve stenosis in a patient with dyspnea and fatigue before valve replacement and surgical radiofrequency ablation. They mentioned that CAF can be diagnosed more frequently if coronary angiography is performed simultaneously with cardiac catheterization to evaluate valve functions or nonatherosclerotic myocardial ischemia in each valvular heart disease case. Our patient also presented with a grade 3/6 systolic murmur at the apex radiating to the back. On TTE, there was presence of moderate mitral regurgitation. Hence, it is a rare finding to see patients with CAF and associated cardiac disease, like in our case, RCA fistula with mitral regurgitation, which was confirmed by TTE.

The management of CAF is complicated and recommendations are based on anecdotal cases or very small retrospective series [12]. Correction of CAF is indicated if the patients are symptomatic or if other secondary complications develop [12]. Early surgical treatment for coronary artery fistulas is safe and effective. The risk of operative correction appears to be considerably less than the po-

tential for development of serious and potentially fatal complications, even in asymptomatic patients.10 The report of Hong and his colleagues showed that out of fifteen CAF patients, for whom six had associated cardiac anomalies, all received surgical correction. All patients had stable condition and were asymptomatic during a mean postoperative follow-up of 13.3 years [10]. Also in the case reported by Gurbuz., *et al*. the patient with CAF and mitral stenosis had coronary angiogram one month after surgical repair which showed normal coronary anatomy and occluded fistula. In this case, our recommendation is for coronary angiography which is to be performed simultaneously with cardiac catheterization to confirm the anatomy, evaluate the valves and to plan the surgical treatment. We recommend surgical repair of the CAF with possible mitral valve surgery, since patient is symptomatic and it is proven from literatures that the surgical risk is less than the adverse events from the procedure itself.

Conclusion

In patients presenting with continuous murmur, especially if symptomatic, a diagnosis of coronary artery fistula should be considered. Non-invasive diagnostic tests, particularly two-dimentional transthoracic echocardiography with Doppler studies, can demonstrate a dilated coronary artery, with flow in the fistula and identification of its receiving chamber, vessel or other vascular structures. From the previous reports cited, our recommendation is to proceed with coronary angiography with cardiac catheterization to confirm the anatomy, evaluate the valves and to plan the surgical treatment, if the patient will give her consent. Surgical repair of the RCA fistula and possible mitral valve surgery is recommended since correction of CAF is indicated if patients are symptomatic and knowing that early surgical treatment for CAF is safe and effective.

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