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Coronary Artery-Left Ventricular Fistula and Takotsubo Cardiomyopathy – An Association or an Incidental Finding? A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Patient: Female, 68
Final Diagnosis: Takotsubo cardiomyopathy
Symptoms: Chest pain • shortness of breath
Medication: —
Clinical Procedure: Percutaneous coronary artery angiography
Specialty: Cardiology

Objective: Rare co-existence of disease or pathology

Background: A coronary artery-left ventricular fistula is an anomalous communication between the coronary arteries and the cardiac chambers and is a rare congenital coronary anomaly that is often small and asymptomatic. Takotsubo cardiomyopathy, on the other hand, is a syndrome characterized by transient regional systolic dysfunction of the left ventricle, mimicking myocardial infarction, but in the absence of angiographic evidence of obstructive coronary artery disease or acute plaque rupture. We present the case of an elderly woman who presented with Takotsubo cardiomyopathy and who was incidentally discovered to have an associated coronary artery-left ventricular fistula.

Case Report: We report the case of a 68-years-old woman with a family history of premature cardiac diseases who presented with ischemic chest pain and elevated troponin levels. Her EKG and troponins were suggestive of non-ST-elevation myocardial infarction (NSTEMI), for which she was initially treated medically and later underwent coronary angiography. Unexpectedly, the angiography revealed patent coronary arteries, and we discovered evidence of coronary artery to left ventricular fistula in the addition to angiographic evidence of Takotsubo cardiomyopathy. A working diagnosis of Takotsubo was made, for which she was treated medically with resulting improvement of her symptoms and later in the imaging findings.

Conclusions: This described case illustrates a rare association between coronary artery fistulas and Takotsubo cardiomyopathy. It is unclear if this association has played a role in the pathogenesis or perhaps is just an incidental finding. More similar cases are needed to expand the clinical presentation of both conditions and add to the literature.

MeSH Keywords: Arteriovenous Malformations • Fistula • Takotsubo Cardiomyopathy

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Background

A coronary artery-left ventricular fistula (CAF) is an anomalous connection between a coronary artery and any of the cardiac chambers or any of the great vessels. It is a rare and often congenital condition detected in approximately 0.2% of coronary angiographies [1]. Takotsubo cardiomyopathy (stress-induced cardiomyopathy) on the other hand is a syndrome of unknown etiology, characterized by transient regional systolic dysfunction of the left ventricle, mimicking myocardial infarction, but in the absence of angiographic evidence of obstructive coronary artery disease or acute plaque rupture. Takotsubo classically affects post-menopausal women, often following an antecedent acute emotional or physical stressor [2]. Patients with CAF are usually older and have a number of clinical presentations, including congestive heart failure, murmurs, atrial arrhythmia, and pulmonary hypertension [3], but it is unclear if Takotsubo cardiomyopathy is a presenting manifestation of the condition.

Case Report

A 68-year-old woman with a medical history including hypertension, dyslipidemia, smoking, and generalized anxiety disorder presented to our facility with chest pain and shortness of breath. The initial workup revealed an elevated troponin level, for which she was admitted with a likely diagnosis of acute coronary syndrome for further management. Home medication

included Carvedilol, Enalapril, Simvastatin, Alprazolam, and Paroxetine. Her mother had been diagnosed with heart disease in her 60s, and her father died of “enlarged heart” at age 36. She has no previous history of cardiac diseases.

On admission, blood pressure and heart rates were 112/73 mmHg and 96 beats/min, respectively. Physical examination of heart and lung was normal except for trace bilateral lower-extremity edema. Troponin level trended up to 1.28 (0–0.4 ng/mL), BNP was 51, BUN was 12 (8–21 mg/dL), and serum creatinine was 0.56 (0.8–1.3 mg/dL). A standard 12-lead EKG demonstrates poor R wave progression and borderline T wave changes, with no features of STEMI (Figure 1). The chest X-ray showed a normal cardiopulmonary process.

She was treated initially (based on a diagnosis of NSTEMI) with heparin drip and nitroglycerin infusion pending coronary angiography. Left heart catheterization and coronary angiography performed the following morning revealed patent coronary arteries, but, after injection of the left anterior descending artery, we noted a coronary artery-left ventricular fistula with small AV malformations, as shown by arrows in Figure 2A. In the same setting, a ventriculogram revealed typical features of Takotsubo syndrome with large focal wall motion disturbances, as shown below in Figure 2B. Transthoracic Echocardiogram also revealed mild reduction in the left ventricular function in addition to the typical regional motion abnormality and the apical ballooning of Takotsubo (Figure 3A, 3B). The patient was started on aspirin and statin (dose-titrated to

Figure 1. 12-lead EKG showing poor R wave progression and borderline T wave changes.

Figure 2. (A) Coronary angiography showing fistulae originating from the left anterior descending coronary artery and draining into the left ventricle, without significant coronary obstruction. (B) Ventriculogram showing regional wall motion abnormality with apical ballooning of left ventricle, a typical feature in Takotsubo.

Figure 3. Transthoracic echocardiography in apical 4-chamber views; Images A and B demonstrating left ventricular end-diastolic volume (LVEDV) and end-systolic volume (LVESV) at the time of presentation. Note the apical ballooning and the reduced function. Images C and D demonstrating LVEDV and LVESV at 4 weeks after discharge as a follow up. Note the improvement in systolic function.

high intensity). Carvedilol and enalapril were continued. Her symptoms completely resolved and she was discharged home on medical therapy in good condition.

At 1 month after discharge, the patient remained asymptomatic. Transthoracic echocardiography performed at follow-up revealed improvement of echocardiographic findings of Takotsubo, albeit with some residual wall motion changes (Figures 3C, 3D).

Discussion

Takotsubo (TM), or stress-induced cardiomyopathy, is a syndrome characterized by reversible transient ventricular systolic dysfunction, mimicking myocardial infarction, but without any angiographic evidence of obstructive coronary artery disease or acute plaque rupture [4]. Classically, TM is usually precipitated by an acute emotional and/or physiologic stress, but in up to 28.5% of the patients no inciting event was discovered in the International Takotsubo Registry study [5].

The precise pathogenesis is unknown; however, sympathetic or catecholamine-mediated myocardial injury and vascular dysfunction is the widely accepted theory [6–8]. The hallmark is radiological evidence of wall motion abnormalities that extend beyond a single coronary territory, identified by echocardiography, angiography, or other imaging studies. This condition is usually associated with recovery of systolic function in the acute period. Nevertheless, patients with Takotsubo should be treated with ACE inhibitors and beta blockers acutely [9].

A coronary artery-left ventricular fistula is an anomalous communication between the coronary arteries and the cardiac chambers and is a rare congenital coronary anomaly found in approximately 0.2% of coronary angiographies [10]. They are often due to developmental anomalies; however, they may also be acquired from conditions such as trauma or invasive cardiac procedures such as coronary angiography, endomyocardial biopsy, or pacemaker implantation [11]. Most of the fistulae drain into the right heart chambers and rarely into the left atrium and ventricle [12].

Most coronary artery fistulae are small and asymptomatic without compromise to the myocardial blood flow. Spontaneous closure often occurs in children but is less frequent in adults [13]. Persistent lesions can lead to coronary steal with a hemodynamic compromise to the affected myocardium. If untreated, hemodynamically significant fistulae result in clinical symptoms or sequelae in up to 63% of adult patients [14]. Symptoms and sequelae include chronic myocardial ischemia and angina, heart failure, cardiomyopathy, myocardial infarction, pulmonary hypertension, endocarditis, rhythm abnormalities, thrombosis of the fistula and associated aneurysm, and, rarely, rupture [15]. Despite the spectrum in clinical presentation of coronary artery

fistulas, it is unclear if TM is associated with these anomalies either as a presenting feature or sequelae. Our search of the literature revealed that the association was reported in 1 case in the year 2012 in South Korea [16].

The clinical diagnosis of a coronary artery-left ventricular fistula is often difficult given the non-specific clinical presentation, laboratory results, and ECG manifestations. Two-dimensional and color Doppler echocardiography are helpful in detection of the entrance and termination site of the shunt, which is characterized by a continuous turbulent systolic and diastolic flow pattern [17]. Fistulae can also be detected non-invasively using 64-slice multi-detector computed tomography [18]. Coronary angiography can be reliably used to detect the size and anatomical features of the fistulae.

Treatment is usually not required for clinically silent hemodynamically insignificant fistulae. Large hemodynamically significant fistulae, on the other hand, require closure, with ligation or coiling during cardiac catheterization being the method of choice. Other factors in treatment the decision include the clinical presentation of the patient and the age at diagnosis. Since most small fistulae tend to get larger with age, elective closure is recommended in younger patients with hemodynamically significant fistulae, irrespective of symptoms [19].

In our described case, despite having a known anxiety disorder, no known emotional nor physiological stressor was identified on presentation, likely marching with the 28.5% of patients who lack identified inciting event, as in the ITR study [5]. It was unclear to us if the concomitant presence of the coronary artery-left ventricular fistula plays a part in the etiology of TM or the resulting incomplete resolution of the segmental wall motion abnormality later detected in follow-up echocardiography.

Conclusions

Although coronary artery-left ventricular fistulae are rare anomalies and are often hemodynamically insignificant, large ones can develop into pathophysiologically significant lesions resulting in clinical symptoms and sequelae. Despite the varieties in clinical features of this rare anomaly, TM is very rarely included as a presenting feature or as an associated condition. A possible mechanism is a steal phenomenon induced by the fistula, leading to regional wall ischemia and resulting in a TM-like picture. More similar cases are needed to further understand the nature of this association.

Conflict of interests

None.

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