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

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Conflict of interest: None declared

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 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.
Despite the spectrum in clinical presentation of coronary artery fistula and associated aneurysm, and, rarely, rupture [15]. Hypertension, endocarditis, rhythm abnormalities, thrombosis of the entrance and termination site of the shunt, which can develop into pathophysiologically significant lesions requiring closure is recommended in younger patients with hemodynamically significant fistulae. Large hemodynamically significant fistulae, on the other hand, require closure, with ligation or coil during cardiac catheterization being the method of choice. Other factors in treatment decision include the etiology of TM or the resulting incomplete resolution of the segmental wall motion abnormality later detected in follow-up echocardiography.

Treatment is usually not required for clinically silent hemodynamically insignificant fistulae. Large hemodynamically significant fistulae, on the other hand, require closure, with ligation or coil during cardiac catheterization being the method of choice. Other factors in treatment 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 variegated clinical presentation 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.
References:

1. Sommer RJ, Hijazi ZM, Rhodes J, John F: Pathophysiology of congenital heart disease in the adult: part I: Shunt lesions. Circulation, 2008; 117: 1090–99
2. Castillo Rivera AM, Ruiz-Bailén M, Rucabado Aguilar L: Takotsubo cardiomyopathy – a clinical review. Med Sci Monit, 2011; 17: RA135–47
3. Luo L, Kebede S, Wu S, Stouffer GA: Coronary artery fistulae. Am J Med Sci, 2006; 332: 79–84
4. Pilgrim TM, Wyss TR: Takotsubo cardiomyopathy or transient left ventricular apical ballooning syndrome: A systematic review. Int J Cardiol, 2008; 124: 283–92
5. Templin C, Ghadri JR, Diekmann J et al. Clinical features and outcomes of Takotsubo (stress) cardiomyopathy. N Engl J Med, 2015; 373: 929–38
6. Haghi D, Athanasiadis A, Papavassiliu T et al: Right ventricular involvement in Takotsubo cardiomyopathy. Eur Heart J, 2006; 27: 2433–39
7. Parodi G: Incidence, clinical findings, and outcome of women with left ventricular apical ballooning syndrome. Am J Cardiol, 2007; 99: 182–85
8. Abraham J, Mudd JD, Kapur N et al: Stress cardiomyopathy after intravenous administration of catecholamines and beta-receptor agonists. J Am Coll Cardiol, 2009; 53: 1320–25
9. Palecek T, Kuchynka P, Linhart A: Treatment of Takotsubo cardiomyopathy. Curr Pharm Des, 2010; 16(26): 2905–9
10. Yamanaka Q, Hobbs RE: Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet Cardiovasc Diagn, 1990; 21: 28–40
11. Friesen CH, Howlett JG, Ross DB: Traumatic coronary artery fistula management. The Annals of Thoracic Surgery. UNITED STATES: Elsevier Inc., 2000; 69: 1973–82
12. Patel S: Normal and anomalous anatomy of the coronary arteries. Semin Roentgenol, 2008; 43: 100–12
13. Cotton JL: Diagnosis of a left coronary artery to right ventricular fistula with progression to spontaneous closure. J Am Soc Echocardiogr, 2000; 13: 225–28
14. Geller CM, Dimitrova KR, Hoffman DM, Tranbaugh RF: Congenital coronary artery fistulae: A rare cause of heart failure in adults. J Cardiothorac Surg, 2014; 9: 87
15. Challoumas D, Pericleous A, Dimitrakaki I et al: Coronary arteriovenous fistulae: A review. Int J Angiol, 2014; 23: 1–10
16. Jo U, Hwang H, Kim H et al: Takotsubo cardiomyopathy in a patient with coronary artery – left ventricular fistulae. Int J Cardiol, 2012; 157: e5–6
17. Vitarelli A, De Curtis G, Conde Y et al: Assessment of congenital coronary artery fistulas by transesophageal color Doppler echocardiography. Am J Med, 2002; 113: 127–33
18. Natarajan A, Khokhar AA, Kirk P et al: Coronary-pulmonary artery fistula: Value of 64-MDCT imaging. QJM, 2013; 106: 91–92
19. Armsby LR, Keane JF, Sherwood MC et al: Management of coronary artery fistulae. Patient selection and results of transcatheter closure. J Am Coll Cardiol, 2002; 39: 1026–32