CLINICAL VIGNETTE

Recurrent Chest Pain and Shortness of Breath with Troponin Elevation in Setting of Apical Hypertrophic Cardiomyopathy with Coronary Cameral Fistula

James S. Lee, MD and Amir B. Rabbani, MD

Introduction

Patients frequently present to the hospital with symptoms of chest pain and positive troponins but at times are free of obstructive coronary artery disease. We present an 82 y/o female with an unusual cause for chest pain and shortness of breath with recurrent troponin elevation.

Case Report

The patient was an 82 y/o Hispanic female with type 2 diabetes, hypertension, and hyperlipidemia who presented to the emergency room with acute onset of epigastric discomfort radiating to the chest that was severe and associated with shortness of breath, abnormal ECG and mildly elevated troponins. She had a similar presentation 3 years prior and due to troponin elevation had undergone coronary angiography and told she had minimal non obstructive coronary artery disease. Patient and family did not wish to undergo repeat coronary angiography and thus was medically treated initially on therapeutically dosed enoxaparin and was also on aspirin, clopidogrel, beta blocker, ACE inhibitor, statin, with nitrates. Upon resolution of symptoms further assessment with lexiscan nuclear stress test revealed small reversible perfusion defect in distal inferolateral wall in setting of preserved LV function (EF 60%) and echocardiogram reported mild to moderate concentric left ventricular hypertrophy with no significant valvular lesions. Patient presented 2 years later, again with severe chest pain this time left sided with associated shortness of breath with ECG with right bundle branch block with T waves in V1-3 and biphasic ST segments and V4-6 (Figure 1) with recurrent mild troponin elevation (troponin I peak of 0.058). On day 2 patient's ECG appeared different with more prominent ST elevation without T wave inversion in V4-6 (Figure 2). Due to ECG changes, the patient was taken urgently to the cath lab and coronary angiography revealed no obstructive coronary artery disease (Figure 3). The patient was noted to have a left anterior descending artery to left ventricle coronary cameral fistula. Echocardiography was repeated and review of images revealed severe hypertrophy localized to the apex with decreased thickening noted during systole along this area (Figure 4). The patient was also diagnosed with heparin induced thrombocytopenia and treated with three months of dabigatran in combination with low dose aspirin along with more aggressive titration of beta-blocker, continued ACE inhibitor, and statin therapy.

Coronary Cameral Fistulas

Coronary cameral fistulas occur in less than 1% of the population and are usually congenital but can also occur from trauma and surgery and described as direct connections between coronary arteries with one of the four cardiac chambers (left atrium, right atrium, left ventricle, right ventricle).[1,2] They are usually classified as arterio-luminal involving communication with the cardiac chamber or arterio-sinusoidal involving communication through a sinusoidal network. A large fistula can cause a steal phenomenon resulting in ischemia due to reduction in myocardial blood flow distal to the site of the fistula connection.[3] The degree of shunting is dependent on the size of the fistula and differences in systemic resistance and resistance in the terminating chamber. More than 90% of the fistulas drain to the right side of the heart and are most commonly seen with the right coronary artery.[4] Patients can be asymptomatic but can also experience chest pain, shortness of breath, palpitations, tachycardia, and syncope. Patients can subsequently develop ischemic angina, congestive heart failure, myocardial infraction, pulmonary hypertension, endocarditis, arrhythmias, thrombosis, and rupture of the fistula.[5] Treatment usually involves percutaneous and surgical repair along with medical management based on size and hemodynamic significance of fistula and associated clinical status.[6]

Apical Hypertrophic Cardiomyopathy

Apical hypertrophic cardiomyopathy is a rare form of hypertrophic cardiomyopathy and is more common among Asian populations specifically in the Japanese.[7] In this condition, hypertrophy mainly involves the apex of the left ventricle.[8] As opposed to patients with hypertrophic cardiomyopathy who present with LVOT obstruction due to location of hypertrophy, midcavity ventricular obstruction can occur. Patients can present with symptoms of chest pain, shortness of breath, palpitations, tachycardia, and syncope which can also lead to ischemic angina, congestive heart failure, myocardial infarction, and arrhythmias.[9] T wave inversions in precordial leads on ECG and "spade-like" configuration with apical wall motion abnormalities are usually noted on diagnostic imaging.[10] Treatment strategies usually involve medical therapy and unlike other variants of hypertrophic cardiomyopathy the prognosis is relatively benign with lower risk of sudden cardiac death and rarely require ICD placement for primary prevention.[11]

Discussion

Coronary cameral fistulas and apical hypertrophic cardiomyopathy are both rare conditions. The presence of both conditions in patients has been described in case reports.[5] Since the presenting symptoms can overlap with so many of the commonly treated cardiac conditions, it is important to recognize the findings for these conditions when patient undergo diagnostic testing with ECG, echocardiography, cardiac computed tomography and magnetic resonance imaging and invasive coronary angiography with catheterization. Treatment measures have not been standardized for these conditions since they are rare and usually individualized. Understanding the hemodynamic significance of the presence of a coronary cameral fistula in conjunction with apical hypertrophic cardiomyopathy allowed us to understand the mechanism for our patient's recurrent symptoms and troponin elevation. As the fistula was very small, aggressive titration of the beta blocker led to marked reduction in symptoms. No surgical or percutaneous interventions were required. Long term prognosis for patients with both conditions is poorly defined but for patients with apical hypertrophic cardiomyopathy in the largest reported North American series was reported to be benign with no reported sudden cardiac death during a fifteen year follow up with long term cardiovascular mortality of 1.9% and annual mortality of 0.1%.[9]

Conclusion

This case demonstrates an unusual and rare diagnosis as the etiology for chest pain and shortness of breath with recurrent troponin elevation. It also emphasizes the importance of a proper differential diagnosis for troponin elevation and to carefully assess the diagnostic studies for any abnormalities that can suggest the etiology of the patient's presentation. It is common for patients to not receive an accurate diagnosis when obstructive coronary artery disease is not found.

Figures

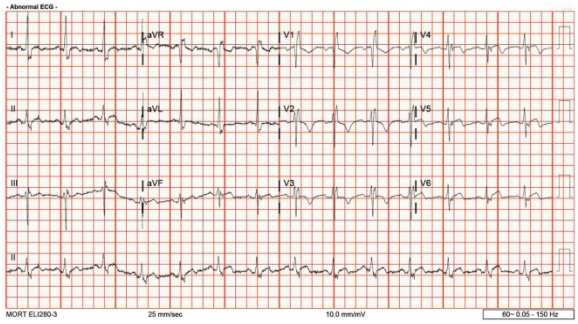

Figure 1: ECG revealing abnormal ST and T wave changes in precordial leads.

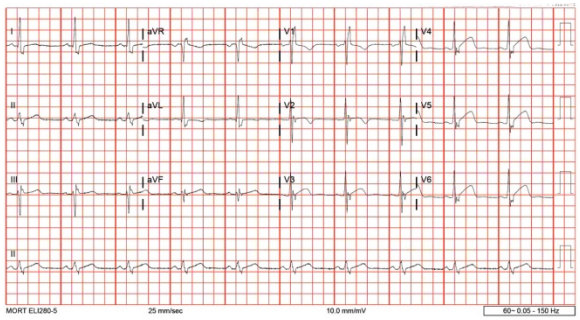

Figure 2: ECG revealing dynamic ST and T wave changes in precordial leads.

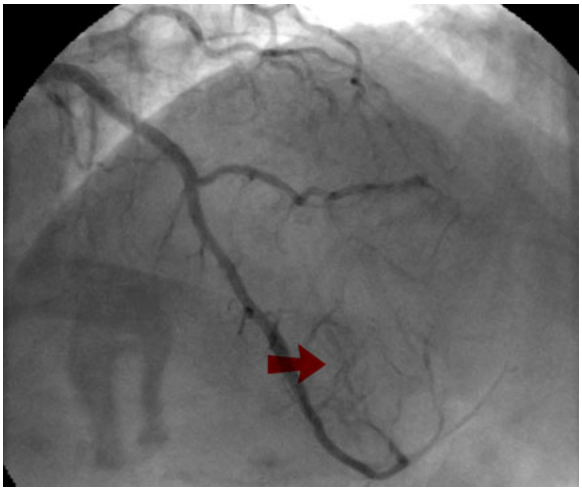

Figure 3: Coronary angiogram with presence of distal left anterior descending artery to left ventricle coronary cameral fistula (arrow).

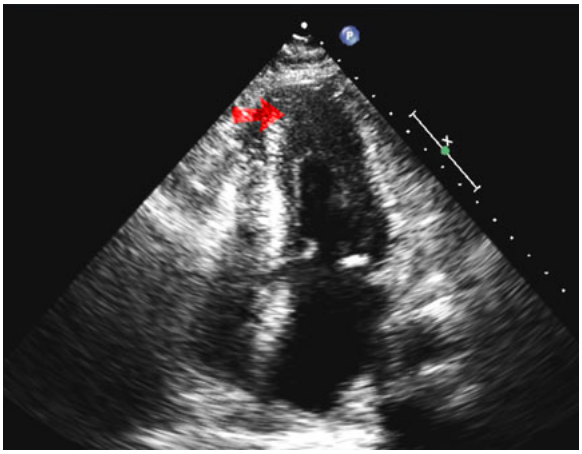

Figure 4: Echocardiogram revealing severe left ventricular apical hypertrophy (arrow).

REFERENCES

1. **Spaedy TJ, Wilensky RL**. Coronary artery fistulas: clinical implications. *ACC Current Journal Review*. 1994; 3:24.
2. **Sandhu JS, Uretsky BF, Zerbe TR, Goldsmith AS, Reddy PS, Kormos RL, Griffith BP, Hardesty RL**. Coronary artery fistula in the heart transplant patient. A potential complication of endomyocardial biopsy. *Circulation*. 1989 Feb;79(2):350-6. PubMed PMID: 2644055.
3. **Schamroth C**. Coronary artery fistula. *J Am Coll Cardiol*. 2009 Feb 10;53(6):523. doi: 10.1016/j.jacc.2008.06.055. PubMed PMID: 19195610.
4. **Levin DC, Fellows KE, Abrams HL**. Hemodynamically significant primary anomalies of the coronary arteries. Angiographic aspects. *Circulation*. 1978 Jul;58(1):25-34. Review. PubMed PMID: 348342.
5. **Said SA, Schiphorst RH, Derksen R, Wagenaar L**. Coronary-cameral fistulas in adults (first of two parts). *World J Cardiol*. 2013 Sep 26;5(9):329-36. doi: 10.4330/wjc.v5.i9.329. Review. PubMed PMID: 24109496; PubMed Central PMCID: PMC3783985.
6. **Chia BL, Chan AL, Tan LK, Ng RA, Chiang SP**. Coronary artery-left ventricular fistula. *Cardiology*. 1981;68(3):167-79. PubMed PMID: 7317886.
7. **Alyan O, Ozeke O, Golbasi Z**. Coronary artery-left ventricular fistulae associated with apical hypertrophic cardiomyopathy. *Eur J Echocardiogr*. 2006 Aug;7(4):326-9. Epub 2005 Jul 1. PubMed PMID: 15994129.
8. **Yamaguchi H, Ishimura T, Nishiyama S, Nagasaki F, Nakanishi S, Takatsu F, Nishijo T, Umeda T, Machii K**. Hypertrophic nonobstructive cardiomyopathy with giant negative T waves (apical hypertrophy): ventriculographic and echocardiographic features in 30 patients. *Am J Cardiol*. 1979 Sep;44(3):401-12. PubMed PMID: 573056.
9. **Eriksson MJ, Sonnenberg B, Woo A, Rakowski P, Parker TG, Wigle ED, Rakowski H**. Long-term outcome in patients with apical hypertrophic cardiomyopathy. *J Am Coll Cardiol*. 2002 Feb 20;39(4):638-45. PubMed PMID: 11849863.
10. **Sakamoto T, Tei C, Murayama M, Ichiyasu H, Hada Y**. Giant T wave inversion as a manifestation of asymmetrical apical hypertrophy (AAH) of the left ventricle. Echocardiographic and ultrasono-cardiotomographic study. *Jpn Heart J*. 1976 Sep;17(5):611-29. PubMed PMID: 136532.
11. **Yusuf SW, Bathina JD, Banchs J, Mouhayar EN, Daher IN**. Apical hypertrophic cardiomyopathy. *World J Cardiol*. 2011 Jul 26;3(7):256-9. doi: 10.4330/wjc.v3.i7.256. PubMed PMID: 21860706; PubMed Central PMCID: PMC3158873.

Submitted November 27, 2017