Polymorphic Ventricular Tachycardia in a Patient with Coronary Arteriovenous Fistulae with Mild Coronary Artery Stenoses

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Torsade de Pointes can be triggered by several mechanisms. Cardiac ischemia is one of the etiologies of Torsade de Pointes. About 50% of patients are asymptomatic in the presence of a coronary arteriovenous fistula; others may have congestive heart failure, infective endocarditis, myocardial ischemia, or rupture of an aneurysmal fistula. They often present with angina because of coronary artery steal phenomenon. We report a 79-year-old female with repeated attacks of Torsade de Pointes even after correcting possible causes of Torsade de Pointes. Coronary angiography revealed two coronary arteriovenous fistulae, one of them arising from the proximal left anterior descending artery and draining into the pulmonary artery and the other from the right coronary artery and also draining into the pulmonary artery. There were coexisting atherosclerotic lesions at the middle and distal left anterior descending artery and the obtuse marginal branch of left circumflex artery. After fistulectomy and coronary artery bypass graft surgery, the patient remained asymptomatic for 2 years.

Key Words: Coronary arteriovenous fistulae • Polymorphic ventricular tachycardia

INTRODUCTION

Coronary arteriovenous fistula (CAVF) is an uncommon congenital anomaly. If the fistula is small, myocardial blood flow will not be compromised, and the patient remains asymptomatic. However, ischemic events can still be induced by “coronary artery steal phenomenon” if fistulae coexist with coronary artery disease. Long QT syndrome with Torsade de Pointes (TdP) can be due to many etiologies, including electrolyte imbalance, medications, structural heart disease, HIV infection, stroke, brain injuries, and eating disorders. We report an aged female who had repeated attacks of TdP associated with stenoses of coronary arteries, and two coronary fistulae, one from the proximal left anterior descending artery (LAD) and the other from the right coronary artery (RCA). TdP and chest pain resolved after fistulectomy and coronary artery bypass graft surgery. The patient remained uneventful during subsequent follow-up.

CASE REPORT

This patient was a 79-year-old female with a history of hypertension, type 2 diabetes mellitus, and atrial fibrillation with frequent long pause, and had had a VVIR permanent pacemaker implanted about 2 months prior to this episode. She complained of dizzy spells and chest tightness one day before admission. Dizziness was preceded by chest tightness and diaphoresis, which could be aggravated by exertion and relieved by resting. The patient’s consciousness was clear on arrival at the emergency room, with stable vital signs. Electrocardiography (ECG) showed pacemaker rhythm and chest X-ray re-
vealed cardiomegaly and pulmonary congestion. Cardiac enzymes, biochemical profile, and hemogram were within normal limits except her potassium was 2.9 mEq/L. Her chest tightness was resolved after giving oxygen, aspirin loading, nitroglycerine infusion and potassium supplement; yet subsequent sudden onset loss of consciousness occurred at the emergency room. The ECG at that time showed polymorphic ventricular tachycardia with changing QRS morphology implicating TdP. The patient regained her consciousness and pacemaker rhythm after 200 joules of defibrillation shock.

After being admitted to cardiac care unit, her heart rate was 81 bpm, and blood pressure 129/77 mmHg and physical examination revealed a GrII/IV continuous murmur at the left upper sternal border. ECG showed atrial fibrillation with moderate ventricular response with T-wave inverted at leads II, III, aVF, V3-V6, and with prolonged QT interval; the corrected QT interval (QTc) measured 642 msec (Figure 1A). TdP repeatedly attacked in spite of intravenous lidocaine plus potassium chloride and MgSO4 supplement. There were no offending medications that could attribute to her ventricular tachyarrhythmia, and her potassium level was 4.6 mEq/L, magnesium 2.6 mg/dL, calcium 7.7 mg/dL, phosphate 2.0 mg/dL after appropriate treatment, but QTc remained at 666 msec. We increased pacing rate up to 80 bpm but in vain (Figure 1B). Serial cardiac enzymes showed her CK was 171 U/L, CKMB 7.9 U/L, and troponin-I 1.20 ng/mL. It was postulated that her long QT and TdP could be due to myocardial ischemia. An emergent coronary angiography showed 40% stenosis of the middle LAD, 58% stenosis of the distal LAD (Figure 2A) and a CAVF arising from the proximal LAD and draining into the pulmonary artery (Figure 2B); there was also 48% stenosis of the left circumflex artery (LCX) obtuse marginal branch (Figure 2B) and a fistula arising from the proximal RCA and draining into the pulmonary artery (Figure 2C). Shunting measurement could not be obtained due to unstable hemodynamics secondary to frequent TdP by echocardiography initially but right-side cardiac catheterization revealed step-up of oxygen saturation and pulmonary-to-systemic flow ratios (Qp/Qs) was 1.26. Percutaneous coronary intervention (PCI) was carried out in an attempt to smooth out lesions of the LAD, with result of residual stenoses of LAD being around 30% with thrombolysis in myocardial infarction (TIMI) flow grade 3.

The patient remained in unstable hemodynamics after PCI, and subsequent echocardiography revealed pericardial effusion which may be due to wire perforation during PCI. A surgeon was called upon for an emergent operation, which proved 2 coronary fistulae arising from the LAD and RCA and draining into the main pulmonary artery. About 350 ml pericardial effusion was drained out, with no active bleeder found except some ecchymoses over the distal LAD. The two coronary arteriovenous fistulae were successfully ligated after coronary artery bypass graft surgery. TdP resolved with QTc shortened to 459 msec about 1 month after the patient’s surgery (Figure 1C). The patient remained chest pain-free and had no more attacks of TdP in the following 2 years.

**DISCUSSION**

CAVF is an uncommon, abnormal communication between the epicardial coronary artery and vena cava, subpulmonary veins, pulmonary artery, mediastinal vessels, coronary sinus or a cardiac chamber, occurring as an incidental finding in 0.1% to 0.2% of routine cardiac angiographic studies. Incidence of fistulae arising from the RCA is around 50%, from the left coronary artery 42%, from both vessels 5% and the remaining 3% not specified.1 41% of fistulae drain into the right ventricle, 26% into the right atrium, 17% into the pulmonary artery, 7% into the coronary sinus, 5% into the left atrium, 3% into the left ventricle, and 1% into the superior vena cava.1 Around 50% of patients are asymptomatic when CAVF is found during evaluation of a cardiac murmur or on a coronary angiography. In the others, CAVF was found to be due to symptoms like congestive heart failure, infective endocarditis, myocardial ischemia, or rupture of an aneurysmal fistula. There is a tendency to occur in early infancy or at > 40 years of age. Physical examinations may reveal a continuous murmur. Selective coronary arteriography is the gold standard for diagnosis. Other diagnostic modalities such as transthoracic echocardiography, transesophageal echocardiography, or multidetector computed tomography have also been reported.2 If the shunt is large enough, the step-up of oxygen saturation can be measured by right-sided cardiac catheterization.

Among patients with atherosclerotic coronary artery
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Figure 1. (A) ECG after 1st ventricular tachycardia episode showed atrial fibrillation with moderate ventricular response, T-wave inverted at leads II, III, aVF, V3-V6, and prolonged QT interval, with corrected QT interval measured 642 msec. (B) Long QT interval with Torsade de Pointes repeatedly attacked in spite of intravenous lidocaine, potassium, MgSO4 supplement and pacing rate increasing to 80 bpm. (C) Torsade de Pointes resolved with corrected QT interval shortened to 459 msec about 1 month after the patient's surgery.
disease, CAVF can lead to certain particular symptoms. In this case, the CAVF arose from the proximal LAD, and there were two discrete lesions with around 40%-60% stenosis. Our proposed theory is a "coronary steal phenomenon", with shunting from the high-pressure coronary artery into the lower-resistance pulmonary system. If the patient only had CAVF or insignificant coronary artery diseases (CAD) with her myocardial blood flow remaining uncompromised, she would be asymptomatic. We propose that the effect of coronary steal phenomenon in conjunction with CAD caused myocardial ischemia distal to the fistula.

In our case, hypokalemia was found initially, but long QT interval with TdP repeatedly attacked despite supplement of potassium and MgSO4. According to the patient’s ischemic ECG change, positive troponin-I level and the findings on coronary angiography, the ischemia-induced long QT syndrome and Torsade de Pointes are the most probable causes, since her TdP resolved and QT interval shortened after fistulectomy and coronary artery bypass graft surgery.

It is generally acknowledged that early afterdepolarizations (EADs) can induce TdP. EADs are single or multiple oscillations of the transmembrane voltage or depolarization and failure of normal depolarization. Prolongation of the action potential duration and the QT interval and repolarization failure may occur by activation of delayed sodium current, an increased inward calcium current and decreased outward potassium current. Sodium influx may precede potassium eflux during ischemia, and the activation of prolong depolarization due to inward sodium current causes net potassium loss. Hypoxia increases the activity and open probability of persistent sodium current in rat ventricular myocytes. Otherwise, a product of ischemic metabolite lysophos-

**Figure 2.** (A) Coronary angiogram in the right anterior oblique cranial projection showed a 40% stenosis of the middle LAD (arrow) and a 58% stenosis of the distal LAD (bold arrow) with TIMI - 2 flow. (B) Coronary angiogram in the right anterior oblique caudal projection showed a fistula from the proximal LAD into pulmonary artery (bold arrow) and a 48% stenosis of the obtuse marginal branch of the LCX (arrow). (C) Coronary angiogram in the right anterior oblique cranial projection showed a fistula from the proximal right coronary artery into pulmonary artery (bold arrow) with TIMI - 2 flow.
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Undrovinas AI, Fleidervish IA, Makielksi JC. Inward sodium current at resting potentials in single cardiac myocytes induced by the ischemic metabolite lysophosphatidylcholine. Circ Res 1992; 71:1231-41.


Phosphatidylcholine changes the sodium channel kinetics, and then causes lengthening of repolarization by a non-nativcation of sodium channel. All the previous findings suggest that myocardial ischemia may increase inward sodium current and decrease outward potassium current, which induces EADs, QT prolongation and precipitates TdP. Furthermore, the numbers of α1-adrenergic receptors, the increased efficiency of effector-receptor coupling, and the release of norepinephrine from local nerve endings are increased by ischemia and all of them augment the net α1-adrenergic activation. EADs are facilitated by α1-adrenergic activation. Kenigsberg et al. reported QTc prolongation developed during coronary angioplasty and early ischemia in a series of all 74 patients undergoing serial ECGs. QTc prolongation may occur during the early phase of ischemia.

The key word coronary arteriovenous fistulae or coronary fistulae combined with Torsade de Pointes or polymorphic ventricular tachycardia was searched by Medline, but no related case report was found. This case may be the first reporting CAVF-induced TdP. CAVF with coronary steal phenomenon is an uncommon etiology of myocardial ischemia, and myocardial ischemia is not causative of TdP frequently. The inference of the extremely uncommon appearance with regard to CAVF-induced TdP is reasonable.

The natural history of CAVF is variable; some have long periods of stability and others have sudden onset or gradual progression of symptoms. Spontaneous closure has been reported in children but is uncommon in adults. There is a general consensus that repairing the fistula is recommended for symptomatic patients, and for those asymptomatic patients at risk for future complications including coronary steals, aneurysms, and large shunts (Qp/Qs greater than 1.5:1). Otherwise, the prognosis among asymptomatic patients is good and a conservative follow-up is recommended. The need for endocarditis prophylaxis among untreated patients remains controversial. Although our patient’s Qp/Qs was 1.26, it is reasonable to assume that her myocardial ischemic was induced by steal phenomenon of CAVF combined with coronary artery stenoses.

Surgical closure usually has low mortality and morbidity, and the long-term outcomes are excellent. The potential efficacy and safety of transcatheter closure make it an alternative method other than surgery. The use of implantable coils is currently considered as the method of choice in the majority of cases. Several factors must be taken into account to choose between surgical and percutaneous treatment of CAVF, including age, concomitant cardiac defects, coronary atherosclerosis, the anatomy of the fistula, and the expertise of the operator. Transcatheter occlusion may be safely performed in older patients with a higher surgical risk and a favorable anatomy, and surgery should be considered for patients having associated congenital defects or considering coronary artery bypass and with a complex anatomy.

CONCLUSION

CAVF can cause coronary steal phenomenon, which may cause myocardial ischemia in the presence of significant or insignificant CAD and induce long QT syndrome and TdP. Closure of CAVF by either surgical method or transcatheter occlusion should be applied in such symptomatic patients.

REFERENCES

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**Errata**

This is hereby to express our sincere apology to the readers.

The third word of the first sentence of the discussion section on page 33 was misspelled in the March issue, 2009 (Vol. 25 / No. 1). The correct spelling should be “venomous” instead of “enomous”.


The title on page 7 is incorrect in the March issue, 2009 (Vol. 25 / No. 1). It should read as follows: “NT-ProBNP but not High Sensitivity CRP Independently Predicts Abnormal Exercise Duke Score in Well-Controlled Hypertension and Pre-Hypertension – A study of Subjects Undergoing Health Evaluation”.


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