CASE OF THE MONTH

A Cardiac Arrest Survivor

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CASE HISTORY

A 55-year-old man presented seeking a second opinion after recovery from a cardiac arrest. He had a history of esophagitis, hiatal hernia, and dyslipidemia. Otherwise, the patient had enjoyed overall good health and had no known hypertension. He exercised regularly, including playing racquetball on a regular basis. A year and a half ago, his primary care provider evaluated him for an "abnormal" electrocardiogram (ECG), and he underwent a nuclear stress test with apparent "normal results." The patient reportedly had a normal catheterization about 6 to 7 years before this visit as part of an evaluation for chest pain. He described the results as "normal." Approximately 3 months before the cardiac arrest, he had one episode of transient syncope lasting 10 to 20 sec while playing racquetball. He did not seek medical care at the time.

On the day of the cardiac arrest, he played racquetball as usual. Later that night, when going to sleep at home, he "felt ill" and within 1 to 2 min lost consciousness. His wife started CPR and called 911; paramedics administered intravenous epinephrine and DC shock with restoration of sinus rhythm. The patient was taken to a local hospital where an ECG was performed (Figure 1) and he was placed on a hypothermia protocol. He subsequently went back into cardiac arrest with ventricular fibrillation (VF), requiring multiple electrical shocks to resuscitate. A review of hospital records showed a mild troponin elevation of 1.8 ng/mL.

Figure 1 and Videos 1-5 are from the previous hospitalization records.

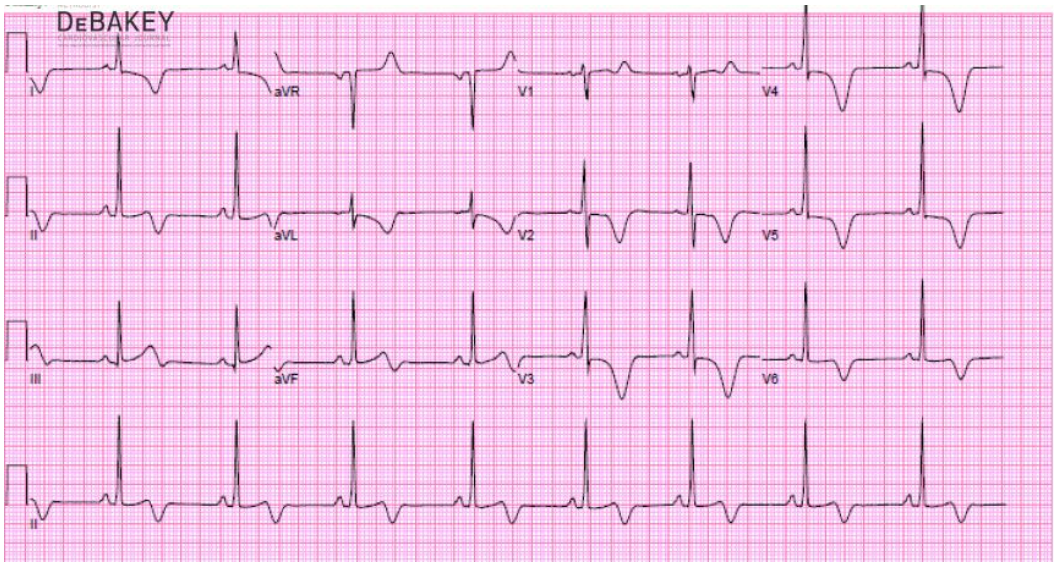

Figure 1. Post-cardiac-arrest electrocardiogram at the outside hospital.

QUESTION 1: What do you suspect?

a) Myocardial ischemia (i.e., acute coronary syndrome [ACS])
b) Left ventricular hypertrophy with strain
c) Hypertrophic cardiomyopathy, apical variant
d) Nonspecific ST-T changes

ANSWER

C: Hypertrophic cardiomyopathy, apical variant

Explanation: The ECG shows diffuse, deep T wave inversion which could be due to ischemia, but in the presence of increased QRS voltage is more likely to be due to apical hypertrophic cardiomyopathy.

CASE CONTINUED

The patient was emergently taken to the catheterization laboratory (Videos 1-4).

Video 1: Left anterior descending artery catheterization
https://youtu.be/kdFB5tk17fU

Video 2: Left circumflex artery catheterization
https://youtu.be/-gm_yBs8jd8

Video 3: Right coronary artery catheterization
https://youtu.be/MqxZFTJSXHI

Video 4: Ventriculography
https://youtu.be/0xp3XvCH6rk

QUESTION 2: What would you do next?

a) Perform a measurement of functional flow reserve (FFR) in the circumflex artery and right coronary artery
b) Perform percutaneous coronary intervention (PCI) in the circumflex artery lesion
c) Perform PCI in the RCA lesion
d) All of the above

ANSWER: D: All of the above

Given the ECG findings, choices A-C are all appropriate. The coronary angiogram showed a moderate lesion in the proximal RCA and a moderate-to-severe lesion in the left circumflex artery. Nevertheless, because of the acuity of the presentation and suspicion of an acute ischemic presentation, PCI was performed on the left circumflex artery, which was interpreted as site of the culprit lesion. The left ventricular (LV) angiogram showed a hyperdynamic ventricle with apical trapping and an apical aneurysm.

CASE CONTINUED

Next, an echocardiogram was performed. Video 5 shows a parasternal long axis view.

Video 5. Echocardiography
https://youtu.be/LKfsY6ftBsM

QUESTION 3: What would you do next?

a) Discharge on appropriate medications for PCI, refer patient to cardiac rehab and reassess in 90 days for secondary ICD prevention (per ACC/AHA guidelines for cardiac arrest during ACS
b) Automatic implantable cardioverter-defibrillator (AICD) before discharge
c) Elective PCI of RCA within 30 days
d) Functional assessment of RCA lesion at a later date

ANSWER

B: Automatic implantable cardioverter-defibrillator (AICD) before discharge

The LV angiogram (Video 4) and the echocardiogram showed typical features of apical hypertrophic cardiomyopathy with apical trapping and a small apical aneurysm (best seen in the angiogram). The ECG findings were quite consistent with this diagnosis. The mild troponin elevation suggested an acute injury, but it is unlikely that this would account for the small apical aneurysm seen in the angiogram. Thus, our choice would be an AICD before discharge given that the possible ischemic event did not appear sufficient to explain the multiple episodes of VF. However, the patient was discharged from the facility without an AICD and with instructions to come back for follow-up.

Since discharge (3 weeks before this clinic visit), the patient has been doing well. He exhibits no symptoms of ischemia, heart failure, or low forward perfusion symptoms. He walks slowly 1 to 2 miles a day.

OUR CLINICAL EVALUATION

Physical examination. When the patient presented at our clinic, his blood pressure was 110/70 mm Hg and his heart rate was 70 bpm. He was afebrile and in no distress. The examination was unremarkable except for a soft early systolic murmur at the left sternal border that did not change with maneuvers.

Imaging. Upon review of the available records (catheterization and echocardiogram), we requested cardiac magnetic resonance imaging (CMR) to better assess the extent of the hypertrophy and the apical aneurysm, and look for perfusion defects and scarring that would place the patient at higher risk for subsequent electrical instability (Video 6, Figure 2).

Video 6. Cardiac magnetic resonance imaging (CMR) shows findings diagnostic of apical HCM with cavity obstruction and a small apical aneurysm.
https://youtu.be/dFsycAQ69vQ

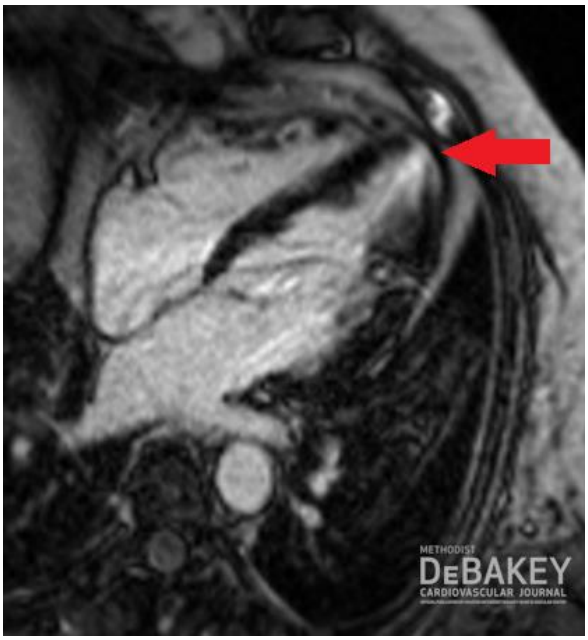

Figure 2. CMR reveals delayed gadolinium enhancement in the apex consistent with scarring. In addition a perfusion defect was documented at the apex after vasodilator stimulation with adenosine.

Next we performed repeat coronary angiography to assess FFR in the RCA territory. FFR was normal at 0.83, and we decided against PCI. However, we felt that the patient had a high risk for subsequent cardiac arrest episodes, so we decided to implant an AICD, which was done without complications. Four months after AICD placement, the patient is doing well.

DISCUSSION

Apical aneurysm in hypertrophic cardiomyopathy (HCM) is a relatively under-diagnosed finding with reported diagnosed incidences of 2% to 3% in patients with apical HCM.[1] For a long time, HCM with apical variant was considered a relatively benign disease, but within the last few years, data is coming to light that demonstrates otherwise.[2]

An apical aneurysm is a discrete, thin-walled akinetic or dyskinetic segment of the most distal portion of the LV chamber. Diagnosis is often made by echocardiography, but in many cases, ultrasound contrast is needed for more accurate detection. Left ventricular angiography and CMR can provide a definitive diagnosis when echocardiography is equivocal.[3]

In a recent review by Rowin et al., apical aneurysms were identified in 4.8% of 1940 consecutive patients with HCM. A higher percentage of patients with apical aneurysm developed ventricular arrhythmias, including ventricular tachycardia (VT) and ventricular fibrillation (VF). One third of patients with an ICD implanted required appropriate shock therapy for VT or VF. Furthermore, thromboembolic events were noted in around 5%; however, the sample was too small to clearly define a role for chronic anticoagulation.[2]

Sudden cardiac death risk calculators for HCM do not account for an LV apical aneurysm. However, given recent data, we believe that this finding should be considered as a stand-alone sudden cardiac death (SCD) risk factor. Furthermore, detection of scar within the aneurysm and reduced perfusion are likely to be markers for higher SCD risk given that, in the population of patients with HCM, these findings are associated with higher incidence of VT and/or VF.

In view of our patient's prolonged cardiac arrest and resuscitative process (both before and after hypothermia), we did not think that a peak troponin of 1.8 ng/mL pointed toward ACS as the cause of his cardiac arrest cardiac arrest; a primary arrhythmic event was a more likely cause.

TAKE-HOME POINTS

1. In patients with apical HCM, the presence of an apical aneurysm denotes a higher risk for life-threatening arrhythmias and SCD.
2. Ventricular arrhythmias are not uncommon and probably arise from scar substrates.
3. Primary prevention ICD should be highly considered and discussed with the patient. Because of an association with higher thromboembolic events, anticoagulation could be considered. However, more data are needed to establish this as an official recommendation.
4. Aggressive detection of apical involvement/aneurysm is needed in patients with HCM. Echocardiography and CMR are preferred imaging modalities. Clinicians must have a high suspicion of this disease entity, and an ECG showing diffuse and deep T wave inversion is helpful in raising suspicion for this diagnosis.

REFERENCES

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3. Rowin EJ, Maron BJ, Haas TS, et al. Hypertrophic cardiomyopathy with left ventricular apical aneurysm: Implications for risk stratification and management. J Am Coll Cardiol. 2017;69:761-73.