Anomalous coronary artery found in the syncopal workup of an elderly man: A Case Report

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ABSTRACT

Syncope, defined as a transient loss of consciousness, is seen in 1% of all ER visits and urgent care clinics in the United States. Syncope is categorized as cardiogenic, neurologic, or psychogenic. Anomalies of the coronary arteries are rare, and anomalous coronary arteries present as syncope more often in the young than in the elderly and rarely occurs in patients 65 years and older. There are two major variants of coronary anomalies. In the first variant, the left main coronary artery arises from the right aortic sinus, and in the second variant the right coronary artery arises from the left aortic sinus. The risk of sudden death is higher in patients with the left coronary artery arising from the right aortic sinus. We present a case of an anomalous coronary artery discovered during the syncopal workup in a 66 year-old man as no such cases have been published in the United States. We will discuss the management of anomalous coronary arteries as well as a systematic approach to the diagnosis and management of syncope.

Case Report

A 66 year-old Caucasian male presented to our family medicine clinic after experiencing syncope. The day prior to his presentation, he reported driving home from work when he experienced chest heaviness, followed by a sensation that he was going to “pass out.” He slowed his car and pulled into a parking lot. Then he experienced a brief loss of consciousness. Upon awakening and after a few minutes, his chest heaviness subsided and completely resolved. He denied any seizure activity, tongue biting, urinary or fecal incontinence. His medical history was significant for hypertension, Parkinson’s disease, and erectile dysfunction. His medications included Carbidopa/Levodopa, Pramipexole, and Vardenafil.
On exam, he was alert, awake and oriented to person, place, and time. His vital signs included a heart rate of 85 bpm, supine blood pressure of 132/82, standing blood pressure of 118/78 and pulse oximetry 99% on room air. No carotid bruits or murmurs were noted on auscultation. He had a resting tremor of his left hand, and cog-wheel rigidity in the bilateral upper extremities. His gait was normal and his neurological exam showed normal strength, sensation to light touch, and deep tendon reflexes.

His EKG showed normal sinus rhythm with nonspecific T-wave flattening. His hemoglobin was 13.9 g/dl (normal: 14-18.0 g/dl), glucose 110 mg/dl (Normal 74- 160 mg/dl) and his sodium, potassium, and calcium levels were normal. His cardiac enzymes included Troponin I, 0.010 (normal: 0.020-0.060 ng/ml) and creatine phosphokinase 76 u/l (normal: 51 – 294 u/l). His chest x-ray showed no acute cardiopulmonary process.

He was admitted to our family medicine inpatient service with telemetry monitoring. Myocardial infarction was ruled out with serial cardiac enzymes. A dipyridamole exercise myocardial perfusion scan was performed and was within normal limits. A transthoracic echocardiogram noted an ejection fraction of 50%, inferior and apical hypokinesis, and impaired relaxation of the left ventricle with diastolic function. Cardiology was consulted and recommended that he undergo a cardiac catheterization.

His cardiac catheterization did not reveal any significant coronary artery disease but showed a left main coronary artery arising anomalously from the right coronary sinus. His coronary artery circulation was right dominant with the right coronary artery supplying the posterior descending artery. In addition, the inferolateral wall was supplied by several large-
caliber posterolateral branches arising from the right coronary artery. He was discharged home and continued on his home medications plus Aspirin and Propranolol. A Coronary Computed Tomography Angiogram was performed and showed an anomalous origin of the left main coronary arising off the right coronary sinus of Valsalva, running an oblique intra-arterial course anterior to the pulmonary artery and ascending aorta. See figures 1-3. He was referred to cardiothoracic surgery for further evaluation who did not recommend surgical repair because they felt surgical risks outweighed the benefits of surgery. The anomalous coronary artery in this patient was the benign variant and was discovered during the workup of syncope. Since he had lived until the age of 66 without prior cardiac symptoms, it is unlikely that his anomalous coronary artery was symptomatic or caused his syncope.

DISCUSSION

Work-up of Syncope

Syncope, defined as a transient loss of consciousness, is seen in 1% of all ER visits and urgent care clinics in the United States (1). Syncope is categorized as cardiogenic, neurologic, or psychogenic. In patients older than 45 years of age, neurocardiogenic, structural heart disease, and psychogenic manifestations are common causes of syncope (2, 3). Syncope presents with a wide variety of symptoms associated with a transient loss of consciousness. The diagnosis is often complicated by the fact that the patient is asymptomatic at the time of evaluation. While most often the cause of syncopal episodes are secondary to benign conditions such as neurocardiogenic reflex, the goal for a thorough and systematic evaluation is to identify serious and life threatening conditions. A systematic approach to a patient with syncope ensures a complete and thorough work up. Patients with a clinical or electrocardiographic history suggestive of arrhythmia,
severe anemia, electrolyte abnormalities, family history of sudden death, older age, and history of severe structural heart or coronary artery disease are considered high risk and hospital admission is recommended (4, 5). Patients younger than 50 years of age, no history of cardiovascular disease, normal electrocardiogram and cardiovascular exam are considered low risk and outpatient evaluation is recommended (4, 5).

At the initial evaluation, a detailed medical history and physical examination including a careful medication review, orthostatic vital signs, and bilateral brachial blood pressure measurements should be obtained. The history and physical examination is crucial as studies have shown a diagnostic utility of 75-85% in patients with syncope (6). Initial diagnostic testing typically includes electrocardiogram in all patients and complete blood count, electrolytes, and glucose measurement as indicated (7). The electrocardiogram serves as a noninvasive tool in the detection of cardiac etiologies, e.g. arrhythmia, myocardial ischemia, or ventricular hypertrophy (8). The initial findings from the history, physical, and laboratory evaluation will help to narrow the focus of evaluation into cardiogenic, neurologic, and psychogenic. See figure 4.

If cardiac ischemia is suspected based on the history and electrocardiogram, cardiac enzymes should be obtained and an evaluation for cardiac ischemia should be initiated, e.g. cardiac stress testing or cardiac catheterization. In patients with history of cardiac disease or abnormal findings in the initial evaluation, echocardiography and a Holter monitor should be obtained. Echocardiography may identify structural heart disease as well as identify left ventricular systolic dysfunction (9). A Holter monitor may identify arrhythmias, e.g. supraventricular tachycardia or paroxysmal atrial fibrillation. Electrophysiological studies should be considered in patients with probable arrhythmia induced syncope. In the absence of structural or cardiac arrhythmias, tilt table testing might be useful to assess neurocardiogenic causes.
Neurogenic and psychogenic causes of syncope should be considered in the absence of any cardiogenic cause or based on initial evaluation. A non-contrasted computed tomography of the brain is often an initial evaluation of non-cardiogenic cause for syncope followed by carotid duplex ultrasonography. Initial testing will help distinguish primary versus secondary autonomic failure in the neurological evaluation of syncope. In the absence of neurological findings, it is important to consider psychogenic causes such as factitious disorders (6). A neurology or psychiatry consultation may be helpful if the etiology is still unclear. See figure 3 for an algorithmic approach to syncope in the adult patient. We suspected a cardiogenic cause of syncope in our patient based on his abnormal electrocardiogram and echocardiogram which prompted additional cardiac testing and led to the diagnosis of anomalous origin of the coronary artery.

Anomalous Origin of the Coronary Artery

Anomalies of the coronary arteries are rare with an incidence of 0.2-1%, and anomalous coronary arteries present as syncope more often in the young than in the elderly and rarely occurs in patients 65 years and older (2, 3). There are two major variants of coronary anomalies. In the first variant, the left main coronary artery arises from the right aortic sinus, and in the second variant the right coronary artery arises from the left aortic sinus. The risk of sudden death is higher in patients with the left coronary artery arising from the right aortic sinus (2). A recent case series of 151 adult patients with anomalous coronary arteries found that the mean reported age was 41 with 17% presenting with life threatening conditions, e.g. ventricular arrhythmia, syncope, or sudden death (10). Coronary artery with an anomalous origin represents a rare and yet important cause of syncope. The left coronary artery or the left main artery typically arises from left sinus of Valsalva and branches into the left anterior
descending and left circumflex arteries. The right coronary artery usually arises from the right sinus of Valsalva. Rarely, the left coronary artery originates from the right sinus of Valsalva or the right coronary artery originates from the left sinus of Valsalva. The incidence of coronary artery anomalies was reported as high as 5.6% in the general population (11).

The majority of coronary anomalies such as split right coronary artery or ectopic right coronary artery from the right cusp are of no clinical significance. Coronary arteries with anomalous origin can result in episodic or obligatory myocardial ischemia, and have been implicated in chest pain, syncope, myocardial ischemia, malignant ventricular arrhythmia and sudden cardiac death. An anomalous left coronary artery from the pulmonary artery leads to obligatory ischemia with 95% mortality rate during the first year of infancy. In comparison, Anomalous Aortic Origin of a Coronary Artery may only result in episodic clinical symptoms and may not be discovered until adulthood.

The subsequent course of coronary artery can be inter-arterial (between aorta and pulmonary artery), retro-aortic, pre-pulmonic or septal (beneath right ventricular outflow tract), all of which predispose to dynamic myocardial ischemia or sudden cardiac death in otherwise young healthy individuals. The increased risk of sudden death may be due to external compression from great arteries, especially after exercise as exercise leads to expansion of aortic root and pulmonary trunk. In addition, the aberrant coronary artery may take an intramural course or have slit like ostium or acute angulation with a bend, further reducing luminal diameter. According to the Sudden Cardiac Death committee of the American Heart Association, coronary anomalies may have contributed to 19% of sudden cardiac death in athletes, (12) only second to sudden death caused by hypertrophic cardiomyopathy (13).
The diagnosis of anomalous origin of a coronary artery was often established post-mortem. In patients with suspected anomalous coronary arteries, echocardiography may establish the diagnosis but the predictive value remains controversial (14, 15). Coronary angiography is generally useful to establish the presence of coronary anomalies. However, the exact course of the aberrant coronary artery may not be easily visualized and determined. Coronary Computed Tomography Angiogram provides excellent depiction of coronary ostia and anatomic course of the artery (16, 17). Coronary magnetic resonance angiography is also being increasingly used because of excellent anatomic delineation and avoidance of radiation exposure, especially in the young adults with suspected coronary anomalies (18).

The presence of anomalous course of coronary artery between the aorta and pulmonary artery in the young adults remains the greatest risk for an adverse event with or without clinical symptoms (19). Surgical intervention is indicated when left coronary arteries arise from the right sinus and courses between the aorta and pulmonary artery. Surgical repair is also indicated with documented myocardial ischemia when coursing between the aorta and pulmonary artery or intramurally. With the right coronary artery arising from the left sinus, surgical repair is also indicated when there is unexplained inducible ischemia in the right coronary artery territory or when coursing between the aorta and pulmonary arteries (20). Surgical revascularization with coronary artery bypass grafting, unroofing (marsupialization) or coronary reimplantation has favorable results, however coronary artery bypass grafting is deemed less desirable due to the presence of competitive flow. Coronary artery stenting has also shown good short-term results and could be the only option in critically ill patients (21, 22).

CONCLUSION

Anomalous coronary artery, while a rare cause of syncope, can easily be missed without a proper systematic approach. This case report describes an anomalous coronary artery discovered
during the syncopal workup in an elderly man. Syncope is a common cause for emergency room visits. The appropriate approach to the evaluation includes a complete history and physical examination and electrocardiogram in all patients. Consider complete blood count, glucose measurement, and electrolytes as indicated. Additional testing may include echocardiogram, Holter monitor, cardiac ischemia valuation, tilt table testing, and electrophysiologic study. Echocardiography is a noninvasive tool that helps assess ventricular function and wall motion abnormalities.
REFERENCES

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A. The normal origins of the coronary arteries are shown, with the left coronary artery (LCA) arising from the left sinus of Valsalva.

B. The anomalous origin of the LCA from the right sinus of Valsalva is depicted showing two variants. The "benign" variant (dashed lines) runs in the septum anterior to the pulmonary artery (PA), while the "malignant" version runs posterior to the PA and between the annuli of the aorta and pulmonary artery, creating a risk for compression of the LCA and compromise of blood flow to the myocardium.
Figure 2. Cardiac computed tomography scan three-dimensional reconstruction. Pulmonary artery was selectively removed from volume rendering images to demonstrate an inter-arterial course of left main coming off right coronary cusp with right coronary artery.

Figure 3. Cardiac computed tomography scan. Left main artery and right coronary artery originated from right coronary cusp. Left main artery coursed between aorta and pulmonary artery was clearly seen.

Figure 4. Algorithmic approach to the workup of syncope in adults

- History and physical examination
  - Includes careful medication review, orthostatic vital signs, and bilateral brachial blood pressure measurements
- Electrocardiogram in all patients and complete blood count, electrolytes, and glucose measurement as indicated

- Suspect cardionic
  - Suspect cardiac ischemia
    - Obtain cardiac enzymes, consider cardiac stress testing or cardiac catheterization
  - Suspect structural disease
    - Obtain computed tomography angiography or magnetic resonance angiography
- Suspect neurogenic or psychogenic
  - Consider computed tomography of brain, carotid artery duplex ultrasonography. Consider neurology or psychiatry evaluation
  - Obtain echocardiogram and Holter monitor

Positive ECG  Positive Holter