vasospasm and provides for immediate percutaneous or surgical therapy.

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Anomalous left coronary artery origin from the opposite sinus of Valsalva: Evidence of intermittent ischemia

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Although a rare anomaly, anomalous aortic origin of a coronary artery that courses between the great vessels (AAOCA) is the second leading cardiovascular cause of death in young athletes.¹⁻³ Common presenting symptoms include chest pain, dizziness, and syncope during or just after exertion; however, patients may also have no symptoms, which makes the diagnosis challenging.¹⁻⁴ After diagnosis, most patients undergo provocative ischemia testing with

an exercise stress test. We present the case of a 13-year-old boy with anomalous left coronary artery arising from the opposite sinus of Valsalva (ALCA), with evidence of ischemia on stress electrocardiography (ECG) on one occasion and a normal stress ECG several days later.

CLINICAL SUMMARY

A 13-year-old boy came to our Exercise Physiology Laboratory for evaluation of exercise-induced asthma. On a submaximal exercise stress test, significant ST-segment depression was noted in the inferior and lateral ECG leads but resolved during recovery (Figure 1). The boy had no symptoms during the test. In the past, he had reported chest tightness with exercise, which had been attributed to asthma. He returned the next day for a clinic visit. Results of physical examination were unremarkable, and his resting ECG was normal. His transthoracic echocardiogram was indicative of ALCA. Cardiac magnetic resonance imaging confirmed the diagnosis,

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FIGURE 1. Electrocardiogram from submaximal exercise stress test on treadmill shows inferior and lateral lead ST-segment depression.

FIGURE 2. Magnetic resonance imaging (off-axis axial) of anomalous left coronary artery (arrow) coursing intramurally between aorta (AO) and pulmonary artery (PA).

showing that the left main coronary coursed tangentially between the aorta and pulmonary artery (Figure 2). One week after his initial exercise stress test, the patient underwent a maximal test with nuclear perfusion. Results of both the exercise and nuclear perfusion tests were normal, and he remained free of symptoms (Figure 3). The patient was referred for surgical repair.

Surgical intervention was performed through a standard median sternotomy with a modified unroofing procedure.

The anomalous coronary orifice was noted to be slitlike, arising above the affected coronary commissure. The postoperative course was unremarkable, and the patient was discharged home 4 days after surgery. A transthoracic echocardiogram performed before discharge demonstrated normal function without aortic or mitral regurgitation. Follow-up exercise testing with nuclear perfusion at 3 postoperative months revealed no evidence of ischemia.

DISCUSSION

This unusual case documents the intermittent nature of myocardial ischemia in AAOCA with laboratory testing. Although this intermittence has been suspected from pathologic findings, ECG evidence has been lacking.¹

AAOCA is a rare congenital anomaly associated with an increased risk of sudden death. Although ALCA is less common than anomalous right coronary artery (ARCA), it carries a higher risk of sudden death.¹⁻³ Most physicians agree that surgery is indicated if a patient of any age with AAOCA shows signs or symptoms of myocardial ischemia. What remains controversial is how to manage the cases of children and young adults who have no symptoms.

On the basis of recent data from Maron and colleagues,³ we calculated the cumulative risk of sudden death from AAOCA during the 20-year period from ages 15 to 35 years, when the vast majority of athletic sudden deaths occur. Assuming that the sudden death risk rate is constant during the 20-year period (which may overestimate the risk because data suggest that sudden deaths cluster before 22 years of age³), a sudden death rate of 0.6 deaths/100,000 people an incidence of 0.2% of AAOCA in the population, that 15% of the deaths are due to coronary abnormalities, that the ratio of ALCA to ARCA is 1 to 6, and that the ratio of deaths from these defects is 4 to 1, then the cumulative risk of death from ALCA is 6.3% and that from ARCA is 0.2%. These calculations tend to support the strategy of

FIGURE 3. Electrocardiogram from maximal exercise stress test on bicycle ergometer 1 week after previous exercise test shows no evidence of ischemia.

operating on all patients diagnosed with ALCA while calling into question the utility of this approach for patients with asymptomatic ARCA.

For those who have no symptoms, an exercise stress test is usually performed to demonstrate evidence of ischemia. These data help guide clinical decision making regarding whether to proceed with surgical intervention. As recommended in the most recent Bethesda guidelines, physicians also use postoperative exercise stress testing before allowing a patient to return to exercise.⁵ Our concern is that ischemia is intermittent in AAOCA. Our patient with ALCA clearly had evidence of ischemia on one exercise test and a normal result just 1 week later. Symptoms of ischemia were not reliable, because the patient was free of symptoms during both tests. Our patient could by chance have had two normal test results and never had his ALCA diagnosed, possibly until a sudden catastrophic event occurred. Indeed, more accurate measures of ischemia are needed in this population. Until

then, use of exercise tests as the sole indicator of ischemia for both preoperative and postoperative decision making should be questioned.

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Primary thoracic cavity gastrointestinal stromal tumor

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Gastrointestinal stromal tumors (GISTs) are mesenchymal tumors that mostly arise in the gastrointestinal tract. Other locations such as the omentum, the mesentery, and the retroperitoneum have also been reported. Here we report a rare case of primary thoracic cavity GIST. Although this rare tumor may be difficult to distinguish clinically from more common mass lesions of the thoracic cavity, a correct diagnosis is necessary because treatment is specific for GIST.

CLINICAL SUMMARY

First Operation

A 62-year-old female patient was hospitalized with a 6-month history of common cold symptoms in September 2007. Her medical record and family history were unremark-

able. The x-ray film showed a dense shadow in the middle and lower lobes of the right lung (Figure 1, A). Computed tomographic scan of the chest revealed a huge heterogeneous mass in the right thoracic cavity, the mediastinum and the heart were pushed to the left, and no mediastinal lymphadenopathy was found (Figure 1, B). There were no other abnormalities by physical examination or laboratory testing.

The patient was then scheduled for surgery because of suspected lung neoplasm. At thoracotomy, there was an 800-mL reactive benign pleural effusion. The tumor was attached to the diaphragm through a 2-cm pedicle and invaded the right lower lobe of the lung but did not involve the esophagus or other mediastinal structures. After the adhesion had been separated from the lung, it was resected en bloc with a portion of the diaphragm.

The tumor was covered with a pseudocapsule except for the site where the tumor was attached to the diaphragm with a gray-white mass measuring 20 × 15 × 10 cm. Microscopically, the tumor was predominantly composed of spindle-shaped cells with a moderate degree of nuclear pleomorphism. Immunohistochemical studies revealed the tumor cells positive for alpha smooth muscle actin and CD34, focally positive for desmin. Considering the origin, morphologic features, and immunohistochemistry of the tumor, leiomyosarcoma was finally reported.

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