Critical Ostial Left Main and Right Coronary Artery Stenosis Secondary to Takayasu Arteritis in a Young Female Simulating Pulmonary Embolism at Presentation

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Abstract: A 28-year-old Caucasian female presented to the Emergency Department (ED) with sudden-onset chest pain. Thirty-six hours earlier, she was diagnosed with a pulmonary embolism, for which anticoagulation was started. Evaluation was significant for atrial fibrillation, elevated cardiac biomarkers, and echocardiography demonstrating a new wall motion abnormality. Symptoms resolved with spontaneous conversion to normal sinus rhythm and the patient was initiated on intravenous heparin and eptifibatide, followed by admission. Shortly after admission, the patient experienced recurrent chest pain with dynamic electrocardiographic (ECG) changes that prompted emergent cardiac catheterization. Prior to angiography, the patient developed pulseless cardiac arrest for which advanced cardiac lifesaving (ACLS) techniques were initiated. Coronary angiography showed critical ostial stenosis of the left main coronary artery. Despite transient periods of hemodynamic stability after successful stenting, the patient decompensated and was pronounced dead. Postmortem findings were consistent with Takayasu arteritis (TA). TA involving the coronary arteries is rare, presenting in fewer than 5% of cases. Diagnosis relies heavily on clinical suspicion. For our patient, dynamic ECG changes with wall motion abnormalities visible on echocardiography prompted diagnostic/therapeutic cardiac catheterization and stenting. Management involves high-dose systemic steroid therapy. However, due to recurrence of disease and adverse effects of prolonged steroid use, additional disease-modifying agents such as methotrexate, azathioprine, or cyclophosphamide may be used. With appropriate therapy, short-term prognosis is favorable. This case underlines the importance of having a high clinical suspicion for TA in the young female population with ischemic symptoms in order to allow early diagnosis in hopes of preventing further complications.

Key words: Takayasu arteritis, pulmonary embolism

Case report
A previously healthy 28-year-old Caucasian woman presented to the emergency department with pleuritic chest pain and tachycardia. Deep vein thrombosis (DVT) risk factors included oral contraceptives and recent airline
travel. Initial labs showed a white blood cell (WBC) count of 13.7 x10^9/L and D-dimer of 1.40 ng/mL. Computed tomography (CT) pulmonary angiography revealed filling defects in distal segmental and subsegmental pulmonary arteries of the left lower lobe consistent with pulmonary embolism. Bedside echocardiogram in the emergency department (ED) showed no evidence of right heart strain. The patient was discharged on enoxaparin 1 mg/kg subcutaneously twice daily and warfarin with close interval follow-up planned. The patient returned to the ED 36 hours later for sudden onset of chest pain, palpitations, and feeling faint. Initial electrocardiogram (ECG) showed atrial fibrillation with rapid ventricular response and associated ischemic ST-segment depressions that improved after spontaneously converting to sinus rhythm. Initial exam was remarkable for tachycardia and a 2/6 systolic murmur at apex radiating to axilla. Laboratory assessment revealed a troponin of 0.54 ng/mL, white blood cell count of 13.2 x10^9/L, erythrocyte sedimentation rate (ESR) of 76 mm/hr, and C-reactive protein (CRP) of 75 mg/L. A pain-free ECG showed no significant ST-segment abnormality. Repeat echocardiogram showed new moderate aortic and mitral regurgitation and severe hypokinesis of the posterior–lateral wall. The patient was admitted to the cardiac care unit (CCU) and started on intravenous heparin and eptifibatide with plans for early cardiac catheterization. Four hours post admission, the patient had recurrent chest pain associated with hypotension and ST-segment changes in the lateral and inferior leads and ST elevation in aVR on ECG (Figure 1). The patient was emergently transferred to the cardiac catheterization lab; however, she developed a pulseless cardiac arrest and advanced cardiac life support techniques were initiated. Coronary angiography demonstrated a 99% ostial left main coronary stenosis (Figure 2). A stent was successfully placed in the left main while cardiopulmonary resuscitation was ongoing (Figure 3). Despite transient periods of hemodynamic recovery, the patient decompensated and died. Postmortem evaluation revealed severe arteritis involving diffuse segments of her ascending and descending aorta, pulmonary arteries (Figure 4 and Figure 5), and coronary arteries consistent with Takayasu arteritis (TA).

Discussion
TA is idiopathic, chronic, large-vessel vasculitis with characteristic clinical features of asymmetrically diminished or absent pulses associated with limb ischemia, vascular bruits, hypertension, and aortic regurgitation, often occurring in females of child-bearing age.1-3 Cardiac complications occur mainly due to uncontrolled hypertension and aortic regurgitation. Less than 5% of cases involve the coronary arteries, most often at ostial regions.1,2 Diagnosis is made based on clinical suspicion supported by imaging studies such as magnetic resonance imaging/magnetic resonance angiography and CT angiography.1,3 Elevations in ESR and CRP may be useful, however, they are neither specific nor sensitive, as levels of these biomarkers are normal in up to 50% of patients with active disease. In addition, there is a discrepancy between radiological progression of disease and elevations of these biomarkers.4 Delays
in diagnosis are common due to the rare nature of this disease and limited diagnostic markers. In the case of our patient, she sought medical attention for a myriad of nonspecific systemic symptoms including fatigue, malaise, and intermittent ear pain for about a year, perhaps suggesting longer-standing unrecognized disease. Patients usually respond well to high-dose systemic steroid therapy.\textsuperscript{2,4} For patients with glucocorticoid-resistant disease or recurrence of disease while on steroid tapering, additional disease-modifying agents such as methotrexate, azathioprine, or cyclophosphamide may be used.\textsuperscript{1,2,4,12} Anti-tumor necrosis factor therapy has been studied with promising results in patients with recurring or refractory disease.\textsuperscript{5} Other aspects of treatment consist of repair of vascular abnormalities by endovascular procedures and vascular bypass surgery. Although there often is good initial response to these interventional therapies, long-term patency is not durable.\textsuperscript{2,4,6-8} Overall, with treatment, short-term prognosis is favorable.\textsuperscript{1,2} From a cardiac standpoint, long-term mortality is commonly due to congestive heart failure.\textsuperscript{1,6,12}

Our case is unique in that the patient’s presentation and initial imaging mimicked a pulmonary embolism, perhaps obscuring her significant coronary artery pathology, ultimately leading to rapid disease progression and deterioration. Although in many cases of TA, radiographic studies showed pathological involvement of pulmonary arteries, pulmonary symptoms are generally uncommon.\textsuperscript{9,10} The patient’s disease progression also was rather rapid. A study of TA natural history demonstrated a cumulative survival rate at 5 and 10 years after the onset of 91.0 ± 3.3% and 84.0 ± 5.6%, respectively.\textsuperscript{11}

Conclusion
This case underlines the importance of having a high clinical suspicion for TA in the young female population with ischemic symptoms in order to allow early diagnosis in hopes of preventing further complications. Although rare, Takayasu arteritis involving the coronary arteries should be considered in any young woman presenting with cardiac ischemia.

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References

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Figure 1. Electrocardiogram during recurrence of chest pain and hypotension indicating dynamic ST-segment changes in inferior and lateral leads and ST elevation in aVR.
Figure 2. Coronary angiogram showing left main ostial stenosis prior to stent placement (arrow). Video link.
Figure 3. Coronary angiogram after stent placement showing adequate coronary blood flow. Video link.
Figure 4. Gross specimen of patient heart: thickening of ascending aorta (arrow) as well as pulmonary artery thickening (arrowhead).
Figure 5. Histological specimen of aorta: intimal wall thickening with fibrointimal proliferation with luminal narrowing (arrow). Medial lymphocytic infiltration and inflammation (arrowhead).