CLINICAL VIGNETTE

Coronary Artery Vasculitis and Aneurysm Formation due to Lupus Erythematosus

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Introduction

Cardiovascular manifestations of systemic lupus erythematosus (SLE) are not uncommon. They include accelerated atherosclerosis, pericarditis, myocarditis and valvulitis.[1,2] However, involvement of major coronary arteries has been reported infrequently. Even more unusual is coronary artery vasculitis and aneurysm formation due to SLE. There have only been 15 cases of SLE-associated coronary artery vasculitis with aneurysm formation reported previously in English literature.[3]

We discuss a 27-year old woman with SLE who presented with chest pain in the setting of clinical and serologic evidence of SLE flare, who was found to have coronary vasculitis with aneurysms on cardiac computed tomography (cardiac CT).

Case Report

A 27-year-old woman was initially diagnosed with SLE at the age of 7 years after developing a malar rash, alopecia, oral ulcers and synovitis. She had positive serologic markers for antinuclear, anti-double stranded DNA, anti-Smith, and anti-SSA/SSB. She was followed closely by rheumatology on maintenance therapy with rituximab, hydroxychloroquine and prednisone.

She reported new-onset, intermittent, left sided chest pressure and palpitations over the week before her routine rheumatology follow up. She also reported a new rash on her face, upper chest, back, and arms. She denied any previous complaints consistent with cardiac disease. There were no changes to her immunosuppressant medications and she reported adherence to treatment. Additionally, she had no previous history of tobacco, alcohol, or illicit drug use. Given the interval development of new symptoms, she was transferred directly to the emergency department for further evaluation of her chest pain.

On examination, in the emergency department she was afebrile with a blood pressure of 108/63; heart rate of 67 beats/min; and respiratory rate of 18/min. She had one small oral ulcer on the right palate and a papular erythematous rash with central scabbing on her face, upper chest, back and bilateral upper extremities. Heart sounds were regular without murmurs, rubs, or gallops. Lungs were clear to auscultation. The rest of her examination was normal.

Laboratory tests were notable for pancytopenia, with white blood cell count of 2.3 x 1000 per mm3, hemoglobin of 10.6 g/dL, platelet count of 88 x 1000 per mm3, and low levels of complement 3 and 4 (34 mg/dL and 2.2 mg/dL respectively). The troponin-I level was elevated and peaked at 1.75 ng/mL. Electrocardiography findings showed normal sinus rhythm with no Q waves or ST segment changes. A chest radiograph was normal. Transthoracic echocardiogram showed a normal ejection fraction of 60-65% and a small pericardial effusion without evidence of tamponade. A computed tomographic pulmonary angiogram showed normal pulmonary arteries, cardiomegaly, and a right-sided aorta.

A cardiac CT was performed to further evaluate the cause of her chest pressure and troponin elevation and showed multiple coronary artery aneurysms involving the left anterior descending artery, first diagonal branch, right coronary artery, and ectasia of the left circumflex artery. The left anterior descending artery had multifocal aneurysms from the origin involving both proximal and mid segments, the largest measuring 1cm. There was also aneurysmal dilatation of the first diagonal branch. The right coronary artery had aneurysmal dilatation involving the proximal and mid portions. The left circumflex artery appeared mildly ectatic. There was also vessel wall thickening and adjacent soft tissue thickening consistent with coronary vasculitis. There was no coronary artery calcifications or obstruction. Given clinical and serologic evidence of an active SLE flare, in addition to a troponin elevation and cardiac CT findings, her presentation was consistent with coronary artery vasculitis with aneurysm formation.

She was treated with parenteral pulse methylprednisolone 1 gram/day for 3 days and IV cyclophosphamide 1 gram. Her chest pressure subsequently resolved, and she was discharged with a plan for aggressive management of her SLE. She was seen for follow-up 3 weeks after discharge in rheumatology clinic and reported doing well with no further episodes of chest pressure.

Discussion

Coronary artery involvement has been reported in, Kawasaki disease and Takayasu arteritis. However, beyond these disorders, vasculitic involvement of coronary arteries is uncommon.[4] Only a few case reports have previously documented coronary vasculitis and aneurysm formation in SLE. The pathogenesis of SLE-associated coronary aneurysm formation is incompletely understood but is thought to be caused by

inflammation and necrosis of the tunica media of the vessel wall.[5,6] The large majority of cases were diagnosed either angiographically or at autopsy. The most common findings reported in SLE coronary vasculitis using coronary angiography include formation of aneurysms with diffuse irregularities and stenosis or occlusion of vessels.[7,8] Our patient's cardiac CT results appeared to be consistent with these angiographic findings. There has been only one other case report that describes the appearance of SLE-associated coronary vasculitis and aneurysm formation on cardiac computed tomography in English literature.[3] Given the rarity of SLE vasculitis there is no diagnostic gold standard and the role of non-invasive imaging has not yet been defined. We believe this is the second case where cardiac CT was used to identify SLE coronary vasculitis.

Conclusion

Non-invasive imaging, in particular cardiac CT, can be used successfully in identifying SLE-associated vasculitis, as evidenced by our case. It can also prevent complications associated with angiography and help differentiate between other cardiac manifestations of SLE, such as accelerated atherosclerotic disease. Moreover, early and non-invasive identification of this process using cardiac CT can help guide appropriate therapy. In our case, cardiac CT findings showing multiple coronary artery aneurysm was essential in the diagnosis, management, and treatment of SLE vasculitis. The availability, non-invasive nature, and diagnostic utility of Cardiac CT imaging supports its use as potentially the gold standard for the diagnosis of coronary artery vasculitis.

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