

Coronary artery aneurysms

Christopher Welles, MD; Scott A. Jorgensen, MD; Alexander J. Towbin, MD; and Richard Towbin, MD

CASE SUMMARY

A 9-year-old girl with known systemic lupus erythematosus (SLE) presents with increasing chest pain. The pain is pleuritic in nature and a pulmonary etiology is suspected.

IMAGING FINDINGS

An initial noncontrast CT scan of the chest showed no significant findings within the lungs to explain the patient's symptoms. However, a soft-tissue abnormality containing a thin rim of calcification was identified within the pre-aortic space (Figure 1). This abnormality was indeterminate, but possible etiologies included vascular or lymphatic abnormalities.

The next day, an ECG-gated, contrast-enhanced CT of the heart was performed. This image showed focal dilatation of the proximal right coronary artery (RCA), just distal to the origin of the artery (Figures 2 and 3) corresponding to the soft-tissue abnormality identified on the non-contrast CT scan. The focal dilatation within the proximal right coronary artery contained a small amount of nonocclusive mural thrombus, as well as a thin rim of calcification (Figure 4). A few smaller areas of focal dilatation were identified within the distal right coronary artery (Figure 3).

Within the proximal left anterior descending artery, there was a small focal dilation as well as a more fusiform dilation located just distally (Figure 5).

DIAGNOSIS

Multiple coronary artery aneurysms secondary to systemic lupus erythematosus

DISCUSSION

Coronary artery aneurysms are a rare complication of a variety of disease processes. Kawasaki disease is the most common cause of coronary artery aneurysms worldwide, particularly in children. Other etiologies include trauma, coronary atherosclerosis, SLE, polyarteritis nodosa, Takayasu's arteritis, scleroderma, rheumatic fever, bacterial endocarditis, and complications from percutaneous intervention.¹

A coronary artery aneurysm is defined as >50% increase in arterial diameter when compared to the adjacent arterial segment;² they are often asymptomatic. When symptoms are present, they are usually related to



FIGURE 1. Axial image from the noncontrast CT of the chest shows a soft-tissue abnormality within the preaortic space (arrow) with a small amount of rim calcification.

myocardial ischemia, such as angina, myocardial infarction, congestive heart failure, or sudden death. Complications from coronary artery aneurysms include thrombosis, embolism, AV fistualization or rupture.² SLE is a rather uncommon cause of coronary artery aneurysms. Patients with SLE frequently have coronary artery manifestations, with atherosclerosis being much more common than aneurysms. Interestingly, clinical disease activity and serological markers do not seem to correlate well with coronary artery vasculitis or aneurysm.³

The right coronary artery is most commonly affected.¹ Catheter angiography historically was used in detect-





FIGURE 2. Axial image from the contrast-enhanced cardiac CT shows focal dilatation of the proximal right coronary artery (arrow), just distal to the RCA origin. This corresponds to the soft-tissue abnormality identified on the non-contrast CT of the chest.



FIGURE 5. Oblique MIP obtained from the cardiac CT shows a small focal area of dilatation within the proximal left anterior descending artery (closed arrow), with a more fusiform area of dilatation just distally (open arrow).

ing coronary artery aneurysms, but due to invasiveness and morbidity, as well as technological advances in other modalities, it is performed less commonly now. Echocardiography is commonly used as a screening exam, with excellent visualization of the proximal coronary arteries.⁴ It can also give valuable data regarding cardiac function to evaluate for complications. With the evolution of multidetector CT, coronary CT angiography (CTA) is becoming more common. Studies have shown CTA to be highly sensitive



FIGURE 3. Sagittal maximum intensity projection (MIP) obtained from the cardiac CT shows the focal area of dilatation within the proximal RCA (closed arrow). Additionally, there are two small focal dilatations (open arrows) within the distal RCA near the posterior interventricular septum.

and specific—and more sensitive than echocardiography for the detection of and demonstration of fusiform and distal aneurysms.² CTA findings include concentric wall thickening with dilatation of the vessel. Often, a thin rim of calcification can be seen. Coronary stenosis and occlusion are important complications. Hemopericardium can be a clue to aneurysm rupture or impending rupture.

Treatment often varies depending on the etiology of the aneurysm.² Many patients are placed on aspirin to decrease the changes of thrombosis. Serial exams with echocardiography and/or CT angiography are often performed to evaluate anatomy and function. If stenosis or thrombosis occurs, coronary stenting can be done, though stenosis from vasculitis can be more difficult to stent than stenosis from atherosclerosis. Coronary artery bypass graft surgery is also performed, often with resection of aneurysms.¹⁴

CONCLUSION

Systemic lupus erythematosus is an uncommon cause of coronary artery aneurysms, with Kawasaki disease being a much more common cause



FIGURE 4. Coronal image from the cardiac CT shows the proximal RCA dilatation (arrow). This image shows mural thrombus within the dilatation, as well as a thin calcified rim corresponding to the calcification seen on the noncontrast CT.

in children. However, children with known SLE who present with chest pain should be evaluated for coronary artery disease, including aneurysms, with echocardiograms or contrast enhanced CT. Treatment varies but can help prevent life threatening complications.

REFERENCES

 Ebina T, Ishikawa Y, Uchida K et al. A case of giant coronary artery aneurysm and literature review. *Journal of Cardiology* 2009; 53:293-300.
Johnson P, Fishman E. CT angiography of coronary artery aneurysms: Detection, definition, causes, and treatment. *AJR*. 2010; 195:928-934.
Karrar A, Sequeira W, Block J. Coronary artery disease in systemic lupus erythematosus: A review of the literature. *Semin Arth Rheum*. 2001;30:436-443.

4. Newburger J, Burns J. Kawasaki disease. *Vasc Med.* 1999;4:187-202.

Prepared by Dr. Welles while a Radiology Resident at the Department of Radiology, Mayo Clinic, Rochester, MN; Dr. Jorgensen and Dr. Richard Towbin, while at the Department of Radiology, Phoenix Children's Hospital, Phoenix, AZ; and Dr. Alexander Towbin while at the Department of Radiology, Cincinnati Children's Hospital, Cincinnati, OH.