THE EYE OF A PROBLEM

Daniel T. Yoo[1,3], Binita R. Chacko[2,3], Shaheeda Ahmed[1,3]

1. *Schulich Heart Program, Sunnybrook Health Sciences Centre, University of Toronto, Toronto, Ontario, Canada.*
2. *Department of Medical Imaging, Sunnybrook Health Sciences Centre, University of Toronto, Toronto, Ontario, Canada.*
3. *Temerty Faculty of Medicine, University of Toronto, Toronto, Canada*

Clinical Presentation

A 68-year-old male with ischemic cardiomyopathy and coronary artery bypass surgery 5 years prior, with grafts to the left anterior descending (LAD) and left circumflex (LCx) artery, presented with exertional dyspnea.

Imaging Findings

A transthoracic echocardiogram (TTE) revealed stable left ventricular function (EF 40%). A 1.6 cm round echolucent structure with a central bright echodensity was seen lateral to the mitral annulus *(image A,B)*, which was previously absent on pre-operative TTE. Based on its location near the atrioventricular groove, a LCx aneurysm was suspected. However, it did not opacify with ultrasound enhancing agents (UEA) *(image C)*.

Cardiac CT angiogram revealed marked 10 mm circumferential soft tissue thickening along most of the native LCx (*image D,E*). The small vessel lumen contained intimal calcification as seen on TTE. Short segments of the proximal LAD and right coronary artery were also abnormally thickened. Bypass grafts were patent.

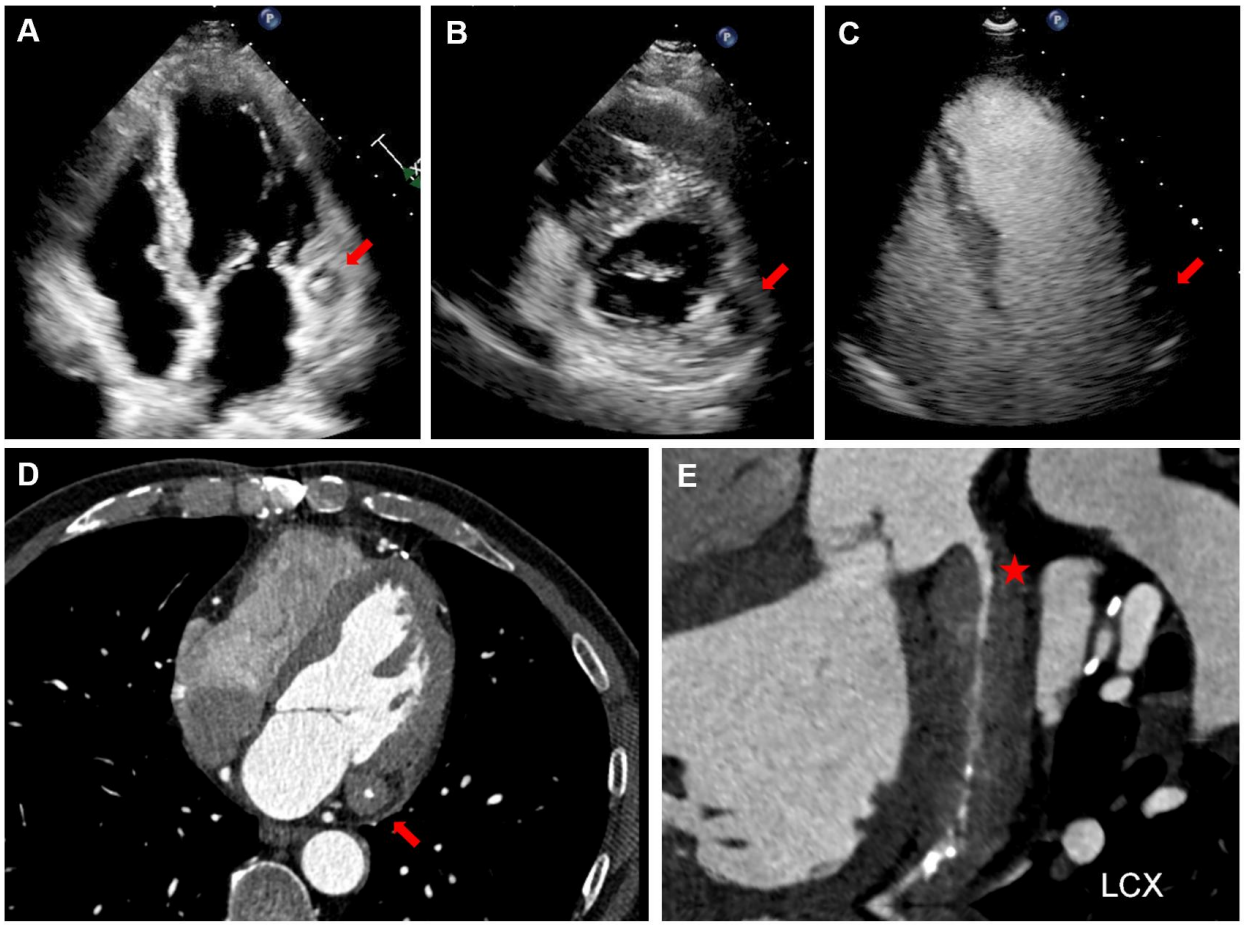
The imaging appearance favoured coronary vasculitis (CV) or IgG4-related disease. Erdheim-Chester disease and low-grade lymphoma were less likely.

Clinically, the patient lacked suggestive systemic features. ESR and total IgG level were mildly elevated, which were both non-specific. Systemic vascular imaging with CT and PET are pending.

Summary/Discussion

CV can be present in up to 50% of primary systemic vasculitis, manifesting as coronary stenosis, occlusion, aneurysm or rupture[1]. Systemic vasculitis involving coronary arteries include Takayasu's arteritis, giant cell arteritis, polyarteritis nodosa, ANCA-associated vasculitis, Behcet's disease and, in children, Kawasaki's disease[2]. Isolated CV, while very rare, has been described previously[3,4]. CV can be mimicked by periarteritis with perivascular thickening from IgG4-related disease and Erdheim-Chester disease[2]. IgG4-related disease can present with lymphadenopathy, salivary gland enlargement, retroperitoneal fibrosis and autoimmune pancreatitis[5]. Erdheim-Chester disease is a rare histiocytic disorder with multiple systemic manifestations.[6]

This case highlights the ability of imaging an abnormal coronary artery on routine TTE, and the importance of anatomic landmarks to formulate a differential diagnosis. As the structure was near the mitral annulus, possibilities included an abnormal LCx, dilated coronary sinus, annular abscess, caseous calcification, granulomatous disease and neoplasm. Echocardiographic contrast (agitated saline or UEA) may help narrow the differential, but complementary imaging is often needed.



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