Correspondence

Incomplete Kawasaki disease with a giant coronary artery aneurysm; an unexpected finding in an adult male

Incomplete Kawasaki disease is a rare entity, especially in adults. Many patients are misdiagnosed or diagnosed retrospectively, with its subtle and atypical presentation.

A 38-year-old male presented with three days of a sharp left-sided chest pain, worse with inspiration and lying down. He denied fever, cough, or shortness of breath, but recently recovered from an episode of bronchitis. A non-smoker with no known history of coronary artery disease, he was transferred to our facility for an initial electrocardiogram (ECG), which showed diffuse ST elevation and high troponin. Physical exam showed a heart rate of 67 bpm, blood pressure 91/50 mm Hg, and a temperature 102.6 degrees F. Jugular veins were distended, with an estimated central venous pressure of 13 cm. Heart sounds were normal, without murmurs, gallops, or extra sounds. The rest of the physical exam was unremarkable.

The laboratory work showed white blood cells at 15,400, hemoglobin 11.2 g/dl, albumin 2.9 gm/dl (3.7–5.1 mg/dl), ESR 78 mm/hr, CRP 190 mg/L (0–10 mg/L), troponin-I 0.14 ng/ml (0–0.04 ng/ml). ECG showed diffuse concave ST elevation with no reciprocal ST-segment changes. Chest X-ray revealed a mildly enlarged cardiac silhouette. Transthoracic echocardiogram showed large global pericardial effusion but no tamponade, normal ejection fraction, and no wall motion abnormalities (Fig. 1).

The diagnosis of myopericarditis was established based on the nature of the chest pain, the presence of diffuse ST elevation on EKG, elevated troponin, and pericardial effusion. An inflammatory or infective process was suspected in view of the fever, elevated white blood cell count, ESR, and CRP. A cardiothoracic surgeon was consulted for placement of a pericardial window for diagnostic

Fig. 1. Echocardiogram Images, (A) Parasternal long axis view showing moderate pericardial effusion behind the posterior LV wall, (B) Parasternal short axis view showing global pericardial effusion, note no collapse of the right ventricle during diastole, (C) and (D) tricuspid and mitral inflow velocities respectively with no significant respiratory variations.
and therapeutic purposes. The patient's chest pain was thought to be out of proportion to the level of pericardial effusion; thus, prior to the procedure, a computed tomography (CT) of the chest was done, revealing a large right-sided mass, measuring 9 cm × 8.3 cm × 10 cm, contiguous with the right atria and pericardial sac, suggestive of coronary aneurysm, along with a large pericardial effusion (Fig. 2).

One likely reason the echocardiogram failed to identify such a mass was the presence of large circumferential effusion and close proximity of the mass to the low pressure right atrium, where most effusions tend to accumulate. Regarding the finding of a large cardiac tumor of unknown etiology, median sternotomy was performed, which revealed the presence of a tense pericardium. On incision of the pericardium, a moderate amount of serosanguinous fluid drained. There was evidence of chronic pericarditis with fibrinous exudate. Diagnosis of a large right coronary artery (RCA) aneurysm was made intraoperatively.

The aneurysm measured 10 cm × 10 cm, so cardiopulmonary bypass was established with aortic and right common femoral vein cannulation. The aorta was cross-clamped, such that cardioplegia could be administered. The wall of the RCA aneurysm was excised, and the inflow and outflow were ligated by oversewing within the aneurysmal sac using 5–0 proline. Following this reversed saphenous vein graft was anastomosed proximally to the ascending aorta and distally to the posterior descending branch of the RCA. Right or left internal mammary artery would not have reached the target in an in-situ form, but free graft was a viable option.

The aneurysm was filled with a large thrombus. No aneurysm was found on the other coronary arteries. The pathology report showed fibrosis with acute and chronic inflammation and granulation tissue formation. No malignant cells or infectious organisms were seen.

Coronary artery aneurysm (CAA) is defined as a localized luminal dilation measuring at least 1.5 times the diameter of a normal adjacent reference segment [1]. Although there is no consensus for a giant coronary aneurysm (GCAA), a size over 20 mm is generally considered [2]. GCAAs are extremely rare, with a prevalence of 0.02–0.2%. The right coronary artery is the most affected, and involved in 40–70% of cases [3]. CAA is usually an incidental finding on coronary angiogram, most are asymptomatic, even though patients can present with angina pectoris, fistula formation, or sudden death [4].

The presence of a coronary artery aneurysm with acute inflammatory pericarditis in this patient raised the possibility of vasculitis, particularly with Kawasaki disease and Takayasu’s arteritis. The former has a predilection for the coronary arteries, compared to...
Takayasu’s, which tends to affect the aorta and its major branches. Our patient’s CT showed a normal aorta and upper thoracic branches, making Takayasu’s arteritis unlikely. As stated, Kawasaki disease rarely presents in adults [5], and our patient did not fulfill the diagnostic criteria for typical or complete disease; however, occasionally an atypical form can occur. Adults, more than children, tend to present with atypical or incomplete Kawasaki disease [6].

Incomplete Kawasaki’s is not well-established in adults, although scarce publications exist; thus, the American Heart Association suggested, in a piece released in 2017, using the same diagnostic criteria as those for children. Our patient met the diagnosis on these criteria (Fig. 3), but probably presented in the subacute phase of the disease, which could explain the absence of persistent fever; he otherwise had a leucocyte count >15,000, anemia with Hb of 11.2 g/dl, and hypoalbuminemia (<3 gm/dl). Two echocardiographic criteria were met, which included coronary aneurysm, a body surface area adjusted-Z score, and a right coronary artery diameter >2.5.

The criteria for incomplete Kawasaki disease has witnessed a less restrictive approach to diagnose and manage these patients. It permits early use of IVIG, even in those who do not fully meet diagnostic criteria, as the risk of undertreatment outweighs that of overtreatment, while IVIG can shorten the course of the disease and its complications, namely aneurysm.

To our knowledge, this is the largest coronary artery aneurysm to be reported in an adult with incomplete Kawasaki disease; this disease should be considered in adults presenting with pericarditis and coronary artery aneurysm. It is not typically thought of in adults, due to its atypical presentation, but can result in serious complications.

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Declaration of Competing Interest

The authors report no relationships that could be construed as a conflict of interest.

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