Case Report

Kawasaki disease masquerading as anomalous origin of left coronary artery from the pulmonary artery

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Summary

Although myocardial ischaemia/infarction is rare in childhood, it is a well-described complication of both Kawasaki disease (KD) and anomalous origin of the left coronary artery from the pulmonary artery (AOLCA). We describe a case of Kawasaki disease appearing as an AOLCA in a 2-year-old boy with myocardial infarction.

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Acute myocardial infarction (MI) in children is well described, albeit rare in practice.[3,9] Both Kawasaki disease (KD) and anomalous origin of the left coronary artery from the pulmonary artery (AOLCA) can result in MI. The clinical features of KD complicated by coronary thrombosis and myocardial ischaemia/infarction and those of AOLCA are similar.

Case report

A 2-year-old boy of mixed racial origin was admitted to hospital with a 1-day history of cough, shortness of breath, vomiting and loss of appetite. There was no previous history of fever, rash, red eyes, or mucosal lesions. The only medical history of note was that he had been on tuberculosis prophylaxis (under 2 years of age: rifampicin, isoniazid, pyrazinamide) for the past 6 months due to an adult contact. No documentation of active tuberculosis was ever demonstrated.

Clinical examination showed an acutely ill-looking, miserable boy. He had a temperature of 38°C (this was the only recorded fever during his entire hospital admission). Heart rate was 168/min, blood pressure 95/45 mmHg and respiratory rate 72/min. He was pale and had good peripheral perfusion. There was no lymphadenopathy, oedema, conjunctival congestion, skin rash or desquamation. The growth parameters were all on the 25th percentile. Cardiovascular examination revealed a jugular venous pressure (JVP) of 4 cm, the apex beat was in the 5th intercostal space lateral to the midclavicular line, and heart sounds were normal, with a gallop rhythm and a systolic murmur at the apex radiating to the axilla. There was intercostal recession with wheezing and crepitations heard in both lung fields. The liver was palpable 6 cm below the costal margin in the right midclavicular line.

The chest radiograph showed an enlarged heart with a cardiothoracic ratio of 0.58, with evidence of pulmonary oedema. ECG showed possible evidence of a MI (Fig. 1). Echocardiography revealed left atrial and ventricular dilatation, a shortening fraction of 29% with an ejection fraction of 55%. The right coronary artery was not dilated and showed a normal colour Doppler flow pattern, but the origin of the left coronary artery was uncertain.

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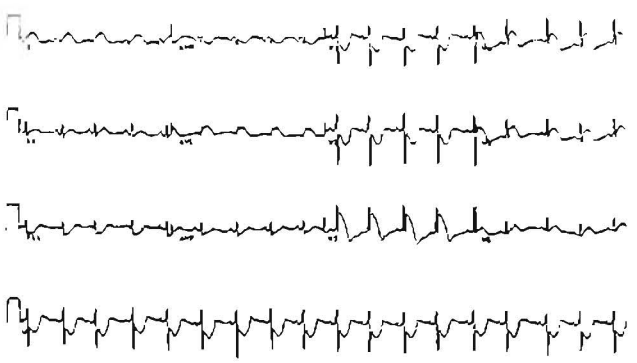

Fig. 1. Twelve-lead ECG. Leads SI, SII, aVL and V3-V6 show picture of hyperacute infarction (non-Q wave), while changes in leads V1 and V2 may be indicative of a posterior infarction.

The white cell count was 14.6 × 10⁹/l (with a normal differential count for age), the haemoglobin concentration 9.6 g/dl, and the platelet count 421 × 10⁹/l. The clotting profile, including antithrombin III and protein C levels, were normal. The erythrocyte sedimentation rate was 18 mm/1st h and the C-reactive protein was repeatedly negative. Antistreptolysin 0 titre and complement levels were normal. Collagen and metabolic screens were negative. Serial cardiac iso-enzymes are shown in Table I.

TABLE I. SERIAL CARDIAC ENZYMES

Day	CK (U/l)	MB fraction (%)	AST (U/l)	ALT (U/l)	LDH (U/l)
1	73	—	128	64	509
2	47	—	96	—	388
10	94	—	54	29	285
18	103	24.2	57	19	358
23	107	20.9	52	21	353
30	84	—	57	—	305
37	68	—	51	—	334
42	101	—	—	—	—
59	68	—	51	—	334
74	78	—	—	—	—
85	105	—	44	—	229
173	—	—	41	20	333

CK = creatinine kinase; AST = aspartate transaminase; ALT = alanine transaminase; LDH = lactate dehydrogenase; MB = muscle brain.

Blood, urine, and stool cultures were negative for bacteria and viruses and blood antibody tests were negative for any recent viral infection.

A presumptive diagnosis of AOLCA from the pulmonary artery was made and radio-isotope scan (pyrophosphate and resting MIBI scanning) was requested as work-up to angiography. This could not exclude an area of infarction in the antero-apical region.

Aortic angiography showed only the right coronary artery arising from the aorta, but no fistulas or retrograde flow to the left coronary artery could be demonstrated (Fig. 2). Repeat angiography 3 weeks later showed no left coronary artery arising from the pulmonary artery, but both coronary arteries arising normally from the aorta and no evidence of aneurysmal dilatation (Fig. 3).

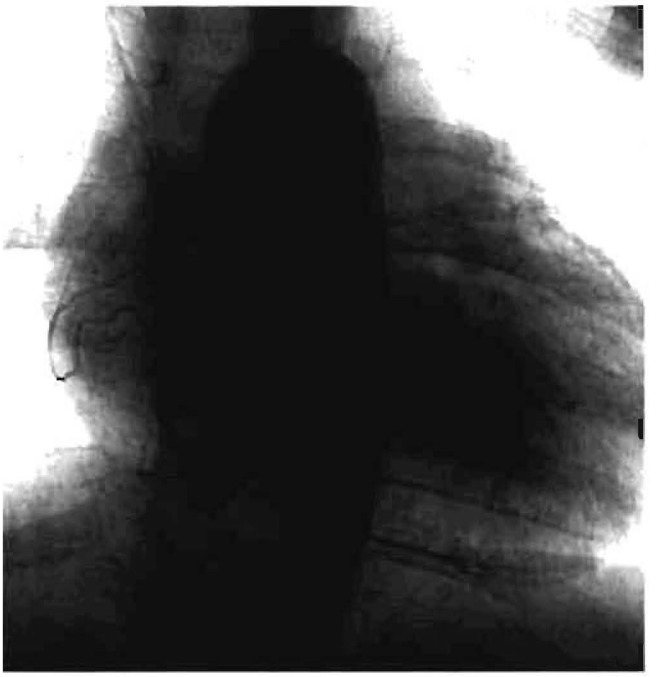

Fig. 2. First aortogram. Normal right coronary artery (RCA), with absence of left coronary artery.

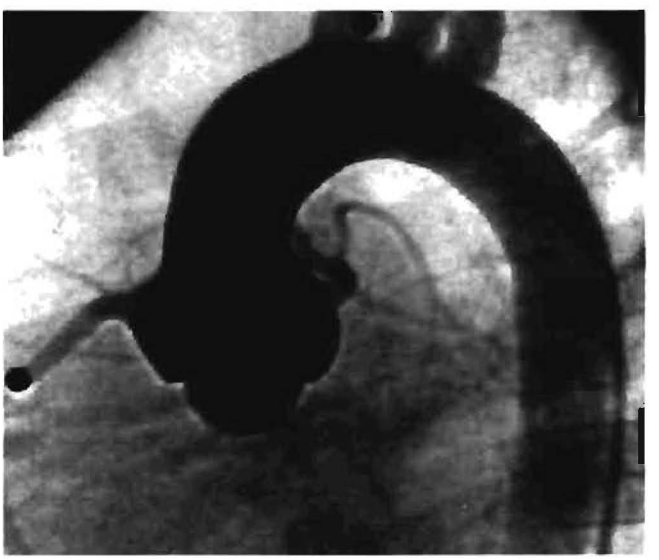

Fig. 3. Repeat aortogram. Normal origin and distribution of left and right coronary arteries (LCA and RCA).

Clinical course

Antifailure therapy in the form of digoxin and furosemide was commenced. The heart failure resolved speedily, but the ECG changes of infarction remained. The boy had intravenous gammaglobulin (IVIG) (2 g/kg) 7 days after admission and aspirin anticoagulation was commenced. He currently remains asymptomatic on the antiplatelet and antifailure therapy. The ECG has normalised, but a repeat echocardiogram at 5 months after presentation shows a clear 5 mm aneurysmal dilatation of the proximal left coronary artery with no stenotic flow pattern (Fig. 4).

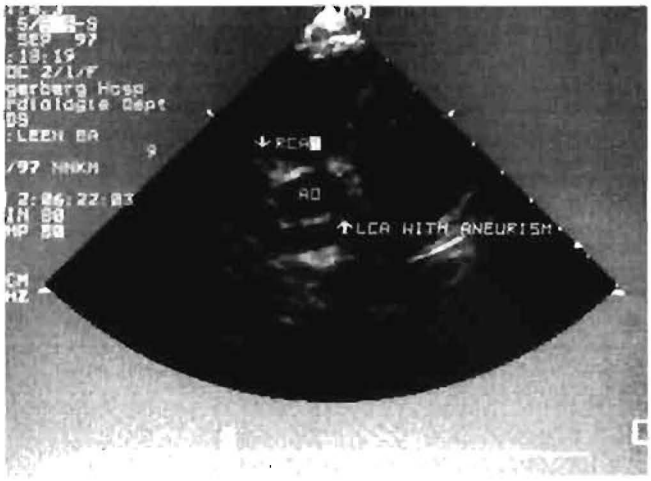

Fig. 4. Short-axis view of aorta (AO) and coronary arteries showing 5 mm aneurysm at origin of left coronary artery (LCA) with normal size right coronary artery (RCA).

Discussion

Vasculitis syndromes are characterised by inflammation of the blood vessels. The clinical picture depends on the size and distribution of the vessels affected. When the coronary arteries are involved, coronary arteritis results. It may therefore be a manifestation of several multisystem diseases.[9] The necrotising vasculitides may be divided into polyarteritis (including infantile polyarteritis nodosa (PAN) and Kawasaki disease), allergic vasculitis, giant cell arteritis, and vasculitis associated with collagen diseases. Each has clinical features that allows classification into subgroups, but the vasculitis present in one patient may have features of more than one subgroup, making exact classification difficult and at times somewhat arbitrary.

KD or acute febrile mucocutaneous lymph node syndrome was first described by Dr Tomisaku Kawasaki in 1967.[7] It has a worldwide distribution, with 20% of untreated cases developing coronary artery lesions.[4,7] The coronary arteritis can lead to aneurysm formation, myocardial thrombosis with infarction, and dysrhythmias.[4] The aetiology and pathogenesis of this disease still remains an enigma.

Pathologically, KD is an acute systemic inflammatory disease with systemic vasculitis[7] mainly affecting children under 5 years of age.[4] There are as yet no diagnostic tests, the diagnosis being based on the presence of 5 out of 6 principal symptoms.[7] Atypical or subclinical cases occur, which do not meet the diagnostic criteria, as coronary aneurysms have been found on echocardiography or at autopsy.[4] The incidence of this form of disease has been reported to be as high as 18.5% in patients with acquired coronary artery disease.[11] These cases have been shown to have a mild clinical course by Japanese researchers, with their American counterparts showing severe residual coronary artery lesions.[7]

The association of KD and coronary aneurysms was first described between 1974 and 1975. Coronary aneurysms are neither exclusive nor pathognomonic of KD, as they have also been described with atherosclerosis, congenital arteriovenous fistulas, PAN, trauma and infection.[3] Although exceptional, coronary aneurysms have also been described with Takayasu's arteritis in association with the aortitis.[10]

A giant thrombus of a coronary artery affected by arteritis will have the same clinical presentation as an AOLCA, namely clinical features of heart failure, systolic murmur, cardiomegaly on radiography, and ECG changes indicative of myocardial ischaemia/infarction. What confounded the diagnosis in our patient was the absence of the principal features governing the diagnosis of KD; we therefore assume it to be a subclinical or atypical presentation. The second angiogram showing both coronary arteries arising from the aorta can only mean that a giant thrombus was occluding the left coronary artery completely, with ECG and angiographic features suggestive of AOLCA. A case of a giant coronary thrombus in a coronary artery complicating KD was described previously by Kadar *et al.*[6]

In our patient the diagnosis of infantile PAN was considered, but this distinction is now only of academic value. Landing and Larson[8] compared the clinical and pathological (both gross and microscopic) features of both the vascular lesions and their patterns of distribution, and they concluded that there was no evidence for a separation of infantile PAN from fatal cases of KD. Fatal cases of KD were first described as having the autopsy findings of infantile PAN.[10]

The clinical presentation of our patient, namely congestive heart failure, a systolic murmur, cardiomegaly and pulmonary oedema on chest radiography, and ECG changes of MI, is classically that of AOLCA.[1,2] Although rare, with an incidence of 0.025 - 0.05%,[11] the first paediatric case was described by Kossof in 1911.[11] Aortic angiography is still the described diagnostic test for this entity.[1,12]

In our case the angiographic findings were misleading, although we should have looked for the presence of a left-to-right shunt initially, as this has important prognostic implications.[12] The flow of blood from the AOLCA to the pulmonary artery is a prerequisite for surgery, if ligation is considered, in order to prevent fatal consequences.

Although the clinical presentation of our patient was typically that of AOLCA, the clinical course, later demonstration of both coronary arteries arising from the aorta and coronary aneurysm formation, is in favour of KD with a subclinical or atypical presentation.

Conclusion

We present a case of a 2-year-old boy presenting in cardiac failure with an ischaemic pattern on ECG, and showing only one coronary artery (the right) arising from the aorta on initial aortic angiography, the latter being the gold standard for diagnosing AOLCA. The ischaemic pattern on ECG remained for many months and follow-up echocardiogram showed an aneurysm on the left coronary artery. We therefore assume his primary disease to be KD, presenting atypically or subclinically. Careful evaluation of aortic angiography and pulmonary artery injection is necessary to confirm the diagnosis of AOLCA.

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