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Coronary Artery Fistula Complicating the Evaluation of Kawasaki Disease

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SUMMARY. **Two patients clinically diagnosed with Kawasaki disease were found to have a coronary artery to pulmonary artery fistula. The dilemma of deciding the etiology of coronary artery dilatation in these patients is discussed along with management.**

KEY WORDS: **Kawasaki disease — Coronary artery fistula**

One of the echocardiographic criteria for coronary artery involvement in Kawasaki disease is a large lumen diameter [5]. Coronary artery dilatation may also be the result of other coronary artery lesions, such as fistulae, anomalous origin, or single coronary artery. We describe two patients with acute Kawasaki disease and coronary artery dilatation seen on echocardiography. However, subsequent coronary angiography demonstrated a coronary artery fistula in both. These cases illustrate the diagnostic and management dilemma in patients with coexisting coronary artery dilatation, coronary artery fistulae, and Kawasaki disease, and the role of serial echocardiography in evaluating these patients.

Case Reports

Case 1

A 3-year-old white female child presented with a 3-week history of fever, a 7-day history of a diffuse erythematous maculopapular rash, stomatitis, cervical adenopathy, and desquamation of the feet and genital area. There was no history of conjunctivitis. The platelet count was 601,000/mm^3 and the antistreptolysin titer (ASO) was negative. The chest x-ray and electrocardiogram were normal. A diagnosis of Kawasaki disease was made and aspirin therapy begun. Gammaglobulin was not started since the patient presented before investigative studies showing its efficacy had been published. The initial echocardiogram (before the development of color flow mapping) showed left ventricular and left atrial enlargement. The left coronary artery (LCA) was enlarged (4 mm) but without discrete aneurysms. Cardiac catheterization 6 weeks later showed no coronary artery aneurysms but did demonstrate a dilated proximal left anterior descending coronary artery (LAD). In addition, a fistula between the LAD and the pulmonary artery was visualized. The fistula was hemodynamically insignificant, and the patient was followed without further intervention. Follow-up echocardiography 2 years later showed a normal-sized LCA (3 mm) without aneurysms.

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Case 2

A 19-month-old male infant presented with a 5-day history of fever and conjunctivitis, a 4-day history of an erythematous rash, and a 1-day history of erythema of the oral mucous membranes and palms. Laboratory studies were significant for a platelet count of 608,000/mm^3 and an erythrocyte sedimentation rate of 50 mm/h. The ASO was negative. A diagnosis of Kawasaki disease was made; intravenous gammaglobulin and high-dose aspirin were begun. The electrocardiogram was normal. The initial echocardiogram showed enlarged chambers. The LAD was dilated (4 mm) but without discrete aneurysms. Dipyridamole was begun. Symptoms and signs resolved within the first days of anti-inflammatory therapy. Peripheral desquamation occurred 2 weeks later. Serial echocardiograms showed persistent enlargement of the LAD. A selective LCA angiogram, performed 4 weeks after the diagnosis, showed a dilated proximal LCA and LAD. In addition, a fistula between the proximal LAD and the pulmonary artery was visualized. The LCA distal to the fistula was normal (Fig. 1). No aneurysms were noted. Dipyridamole was discontinued, and the aspirin dose was lowered (5 mg/kg/day) and, 6 weeks later, discontinued. A follow-up echocardiogram 6 months later still showed a slightly enlarged left coronary artery without discrete aneurysms. Color-flow Doppler mapping failed to reveal the fistulous tract in the initial or follow-up studies, despite the knowledge of its presence.

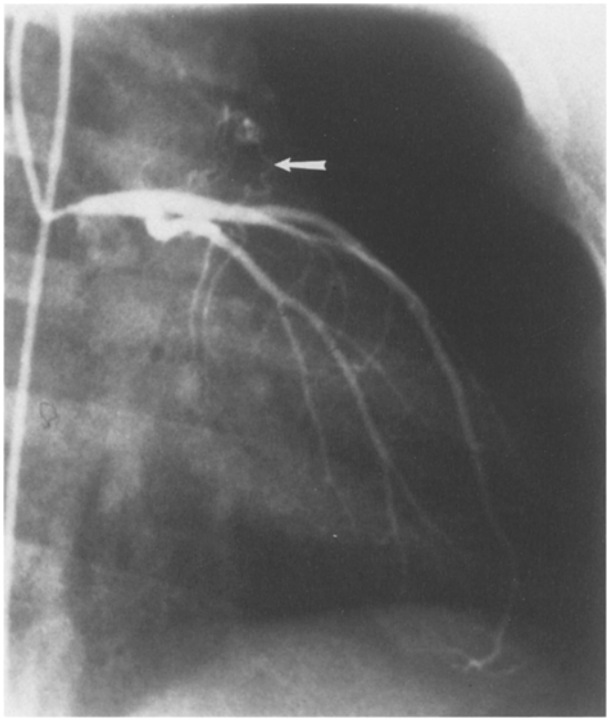

Fig. 1. Selective LCA angiogram showing a dilated proximal LCA, with a coronary to pulmonary artery fistula (*arrow*) arising from the LAD.

Discussion

In the initial evaluation for coronary artery involvement in Kawasaki disease, the current standard of practice is to obtain an echocardiogram. Previous studies [1, 3, 4, 7] have shown that the initial echocardiogram is a very sensitive test for the presence of coronary artery aneurysms. The preference for a noninvasive screening examination is obvious, but echocardiography may fail to show coronary artery stenosis in patients after the acute phase of Kawasaki disease [6] and may not demonstrate lesions of the distal coronary arteries [3]. Therefore, some investigators [1, 4, 6] suggest performing angiography when echocardiography demonstrates coronary involvement, which includes aneurysms (saccular or fusiform), dilatation (>3 mm in a child <5 years old, >4 mm in a child >5 years old), irregularity, or stenoses [5, 6]. However, the differential diagnosis of coronary artery dilatation also includes coronary arteriovenous fistula, anomalous origin of the coronary artery, and single coronary artery [2].

We describe two cases in which angiography was performed to further investigate coronary artery enlargement seen on echocardiography and attributed to Kawasaki disease. Echocardiography did not demonstrate other coronary artery lesions; however, in both cases, coronary angiography demonstrated a dilated LAD and a small coronary artery fistula. This presented a diagnostic dilemma. In both cases the absence of lumen irregularities, aneurysms, or abnormal tapering in the segment proximal to the fistula made the diagnosis of coronary involvement secondary to Kawasaki disease less likely. It is believed that coronary dilatation from Kawasaki disease may be more transient and resolve as inflammation subsides. Coronary artery dilatation secondary to a fistula would more likely persist or even progress. In case 1, since follow-up echocardiography (2 years after patient presentation) has shown normalization of coronary artery size, initial coronary artery dilatation may indeed have been due to involvement from Kawasaki disease rather than from the coronary artery fistula. In case 2, the follow-up is not long enough to make a conclusion. Follow-up serial echocardiography and possibly coronary angiography will be necessary to distinguish between these pathogenic mechanisms.

These cases serve to illustrate that patients with coronary arteries appearing suspicious on echocardiography should undergo coronary angiography not only to define the extent and nature of coronary involvement in Kawasaki disease but also to demonstrate other rare causes of coronary artery dilatation, which may alter clinical management.

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