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CARDIOVASCULAR IMAGING

ORIGINAL ARTICLE

Vascular imaging findings with high-pitch low-dose dual-source CT in atypical Kawasaki disease

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PURPOSE
Determining the presence of aneurysms, thrombosis, and stenosis is very important for the diagnosis of atypical Kawasaki disease (AKD) and in the follow-up of AKD patients with aneurysms. We aimed to demonstrate high-pitch low-dose dual-source computed tomography (CT) angiography findings in pediatric patients with AKD.

METHODS
Over a 5-year period, high-pitch low-dose CT angiography was performed to determine vascular aneurysms or occlusions in 17 patients who had suspected AKD. The patients ranged from 2 months of age to 11.3 years, with a mean age of 3 years. The American Heart Association's criteria were used to diagnose AKD.

RESULTS
We did not detect any vascular problems in 6 of the patients, and they were not included in our study. Arterial aneurysms were present in 11 patients (aged 2 months to 11.3 years; mean age, 4.2 years; 7 males). In one patient, there was also a thrombus at an arterial aneurysm. Coronary artery aneurysms were detected in 7 patients and systemic artery aneurysms were detected in 7 patients. Three patients had both systemic and coronary aneurysms.

CONCLUSION
Our results suggest that high-pitch low-dose dual-source CT can detect all types of aneurysms, stenosis and occlusions of vessels in patients with AKD who were not previously diagnosed. This useful, easy, robust and fast technique may be preferred to diagnose AKD.

Kawasaki disease (KD) is an acute, febrile, self-limited vasculitis that affects small- and medium-sized arteries, with a predilection for the coronary arteries (1). The disease mainly affects infants and children younger than 5 years of age. The etiology of KD is unknown, and there are no specific diagnostic tests (2). The diagnosis of this disease is ideally made by clinical criteria according to the American Heart Association. Typical or classical Kawasaki (TKD) disease can be diagnosed if a fever lasts longer than 5 days and if a patient has 4 of the 5 clinical features. However, according to the Japanese guidelines, atypical or incomplete Kawasaki disease (AKD) is defined as the presence of 4 or fewer of the main findings of KD regardless of the presence or absence of coronary artery aneurysm (CAA).

In some cases with unexplained prolonged fever, the clinical features may be insufficient for the diagnosis of TKD. In this instance, AKD may be considered. Determining the presence of aneurysms in small- and medium-sized arteries is quite important in the diagnosis of AKD (3). CAAs are seen in up to 25% of cases, and systemic artery aneurysms (SAA) are seen in 2% (4). Especially in atypical cases, cardiac and other vascular complications can be more frequently seen because the diagnosis can be delayed. Therefore, early diagnosis and treatment is very important to prevent complications (5).

There are several imaging modalities for scanning vascular structures, including echocardiography, magnetic resonance imaging (MRI), computed tomography (CT) and ultrasonography (US). In the diagnosis of AKD, each of these imaging techniques has specific handicaps. High-pitch low-dose CT angiography can be very useful in screening for CAA and associated SAA in patients with AKD.

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Table. CT angiographic distribution of aneurysms in patients with atypical Kawasaki disease

Patient number	Coronary artery	Pulmonary artery	Axillary artery	Ulnar artery	Renal artery	Iliac artery	Femoral artery	Popliteal artery
1	+							
2	+					+	+	
3					+			
4	+							
5	+	+						
6	+		+	+				
7						+		+
8	+							
9	+							
10					+			
11			+*					

*The right axillary artery had thromboses.

In this article, the diagnosis and clinical features of AKD are presented with high-pitch low-dose dual-source CT angiography.

Methods

Patient selection

The study subjects seen at our institution between 2012 and 2017 with suspected AKD who did not have enough clinical features for a diagnosis of TKD according to the American Heart Association diagnostic guidelines were included in our study. We evaluated 17 consecutive patients who were referred to us by our center or an outside center at the pediatric cardiology clinic. High-pitch low-dose CT angiography was performed in all patients because systemic aneurysms can be present without CAA. Each CT angiography study was examined for aneurysms and occlusive disease. The age of the patients ranged from 2 months to 11.3 years, with a mean age of 3 years. Seven of the patients were male. Six patients did not have any aneurysms and were therefore excluded. This study was approved by our institutional ethics committee (the decision number of ethics committee approval: B.30.2.ATA.0.01.00/91), and informed consent was obtained from the families of all patients.

Main points

- Kawasaki disease is a self-limited vasculitis of small- and medium-sized arteries.
- The disease mainly affects infants and children younger than 5 years of age.
- When clinical criteria are insufficient for the diagnosis of typical Kawasaki disease, atypical Kawasaki disease can be considered.
- High-pitch low-dose dual-source CT can detect all types of aneurysms, as well as stenosis and occlusion of vessels in patients with atypical Kawasaki disease.

CT protocol

All high-pitch low-dose CT examinations were performed on a dual source CT system (Definition Flash, Siemens Healthcare). The scans were performed with free-breathing, in a craniocaudal direction. CT parameters were as follows: 0.28 s gantry rotation time, 128×0.6 mm slice acquisition by z-flying focal spot technique, weight adapted setting for tube current (50 effective mAs for patients <5 kg body weight, 80 effective mAs for patients 5–10 kg body weight, 100 effective mAs for patients >10 kg body weight) at 80 kV tube voltage, 411 mm/s table speeds. The high pitch was 3.4 for CT examinations.

Contrast agent (Iopromide, 350 mg I/mL, Ultravist, Bayer HealthCare) was injected via the peripheral vein at a volume of 1.5 mL/kg body weight with a chaser saline of 1.0 mL/kg body weight. After the contrast material and saline were injected, the scan was started immediately without delay. Vac-lok cushions were used for the immobilization of patients. A reconstruction of the images was conducted with a slice thickness of 0.75 mm and increment of 0.5 mm.

We evaluated a broad range of anatomic areas on the CT. The examination was usually focused on clinical symptoms of the patients as well as the coronary arteries such as the branches of the abdominal aorta and extremity arteries. Generally positive findings were detected at symptomatic areas and in the different arteries. Each of the scans was finished in 1–1.5 s without any complications.

All of the images were assessed in consensus by two radiologists who were blinded to the information about the patients and who had more than 4 years of experience at a workstation (Syngo Via, Siemens Healthcare).

Results

Multiple CAA and several SAA were found in 11 patients (age range, 2 months to 11.3 years; mean age, 4.2 years; median age, 26 months; 7 males), and AKD was diagnosed in these patients (Table). CAA was present in 4 patients without SAA (36%) (Fig. 1). SAA was present in 4 patients without CAA (36%). Three patients had both SAA and CAA (27%).

Two patients had sterile pyuria, proteinuria and flank pain. CT angiography

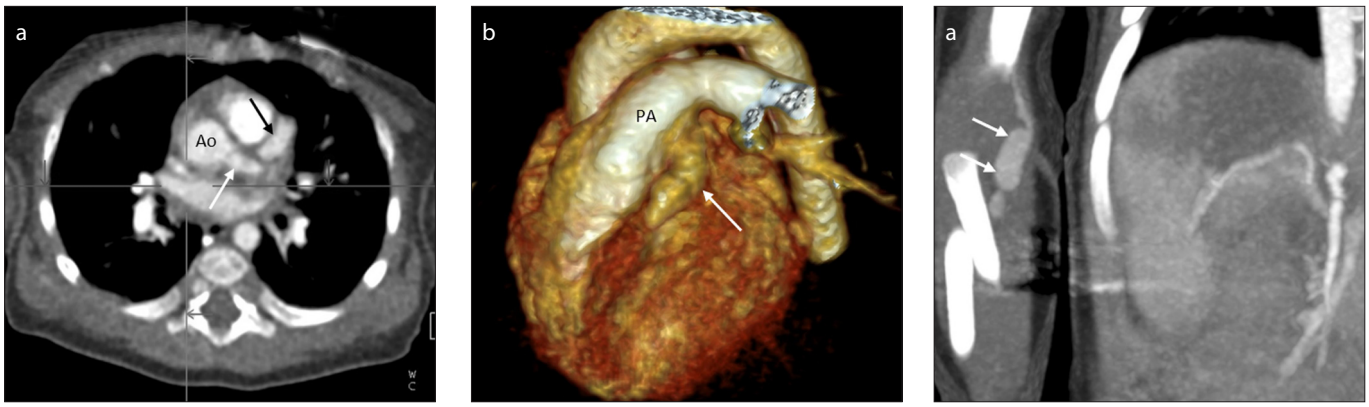

Figure 1. a, b. Left main coronary artery (LMCA) and left anterior descending (LAD) artery aneurysms in a 6-month-old male with unexplained fever. Axial MIP CT angiogram **(a)** shows LMCA *(white arrow)* and LAD *(black arrow)* aneurysms (Ao, aorta). Anterior 3D volume-rendered CT image **(b)** demonstrates an LMCA aneurysm *(arrow)* (PA, pulmonary artery).

Figure 2. Bilateral renal artery aneurysms in a 4-month-old male with fever and sterile pyuria. Coronal MIP CT angiogram shows bilateral renal artery aneurysms *(arrows)* (Ao, aorta).

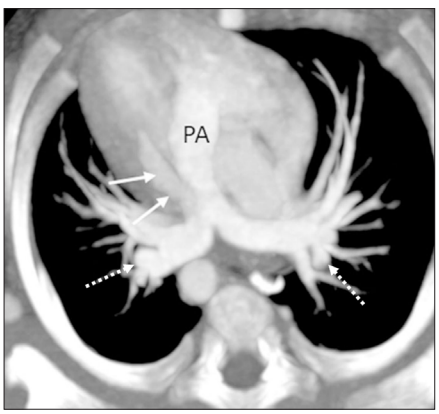

Figure 3. Bilateral pulmonary artery aneurysms accompanying a coronary artery aneurysm in an 8-month-old female with fever, cervical lymphadenopathy (>1.5 cm) and bilateral conjunctival infection. Axial MIP CT angiogram shows bilateral pulmonary artery aneurysms *(dashed arrows)*, and an RCA aneurysm (solid arrows) (PA, pulmonary artery).

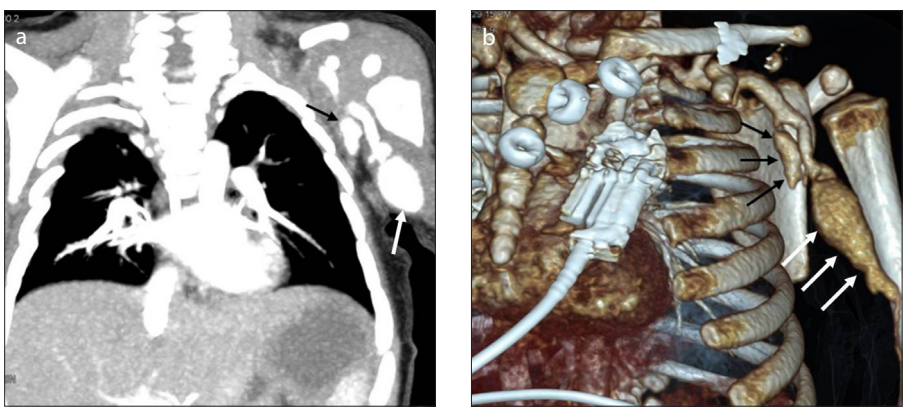

Figure 4. a, b. Left axillary artery aneurysms without thromboses in a 5-year-old male patient with peripheral gangrenous findings and fever. Coronal CT angiogram **(a)** demonstrates left axillary *(black arrow)* and brachial *(white arrow)* artery aneurysms without thromboses. Anterior 3D volume-rendered CT image **(b)** shows left axillary *(black arrows)* and brachial *(white arrows)* artery aneurysms.

demonstrated the presence of renal artery aneurysms without CAA (18%) (Fig. 2). Pulmonary artery aneurysm was present in addition to CAA in one patient (9%) (Fig. 3).

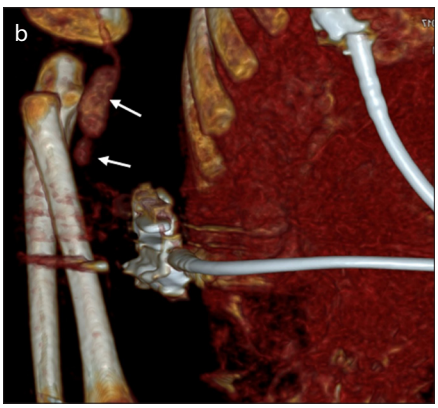

Figure 5. a, b. An ulnar artery aneurysm in a 5-month-old female patient with fever and rash. Coronal MIP CT angiogram **(a)** shows a right ulnar artery aneurysm *(arrows)*. Anterior 3D volume-rendered CT **(b)** shows a right ulnar artery aneurysm *(arrows)*.

Axillary artery aneurysms were found in two patients (18%). In one patient with axillary artery aneurysm, CAA was also present. In the other patient, the axillary artery aneurysm was the only arterial aneurysm that was identified on CT (Fig. 4). This axillary artery was thrombosed, and peripheral gangrenous features were found in this patient.

One patient had an ulnar artery aneurysm with CAA and axillary aneurysm (Fig. 5).

CT angiography showed iliac artery aneurysms in two patients (18%) (Fig. 6). In one patient, a femoral artery aneurysm was accompanied by iliac artery aneurysm and CAA (Fig. 7), and in another patient, a popliteal artery aneurysm was found along with an iliac artery aneurysm (9%) (Fig. 8).

Right iliac artery aneurysm, the largest of these aneurysms, was measured 25 mm in diameter. One of the CAAs was the smallest aneurysm, with a diameter of 4 mm.

The effective radiation dose was measured as 1.2 to 4.3 mGy depending on the patient's body weight.

Discussion

The diagnosis of TKD is easy, and treatment can be started without loss of time, while AKD has an atypical clinical presentation, and its diagnosis is very difficult. When treatment is delayed, the consequences may be disastrous. Thus, in patients with AKD, determining the presence of vascular aneurysms is crucial (4, 6). Coronary arteries are the most common location for aneurysms in patients with KD, but SAAs can also be seen on rare occurrences.

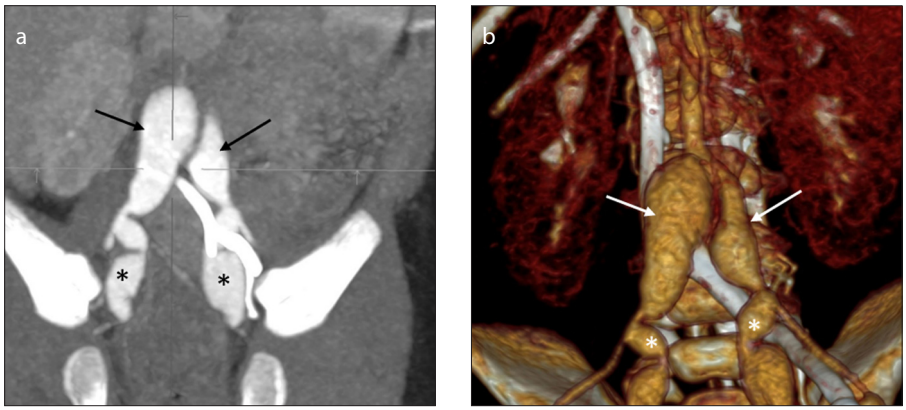

Figure 6. a, b. Bilateral common iliac artery and internal iliac artery aneurysms in a 7-year-old male patient with fever and cervical lymphadenopathy (>2 cm). Coronal MIP CT angiogram **(a)** shows bilateral common iliac arteries *(black arrows)* and bilateral internal iliac artery aneurysms *(asterisks)*. Anterior 3D volume-rendered CT image **(b)** shows bilateral common iliac arteries *(white arrows)* and bilateral internal iliac artery aneurysms *(asterisks)*.

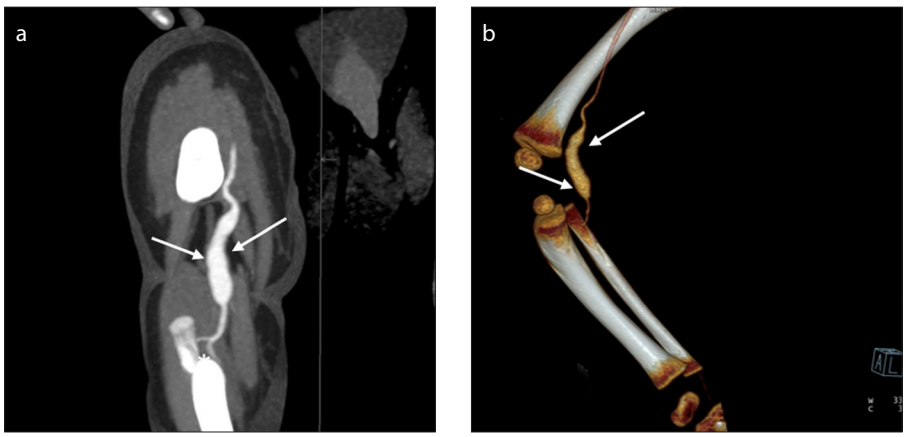

Figure 7. a, b. A right popliteal artery aneurysm in a 6-month-old male patient with fever and erythema of the feet. Coronal MIP CT angiogram **(a)** demonstrates a right popliteal artery aneurysm *(arrows)*. Lateral 3D volume-rendered CT image **(b)** shows a right popliteal artery aneurysm *(arrows)*.

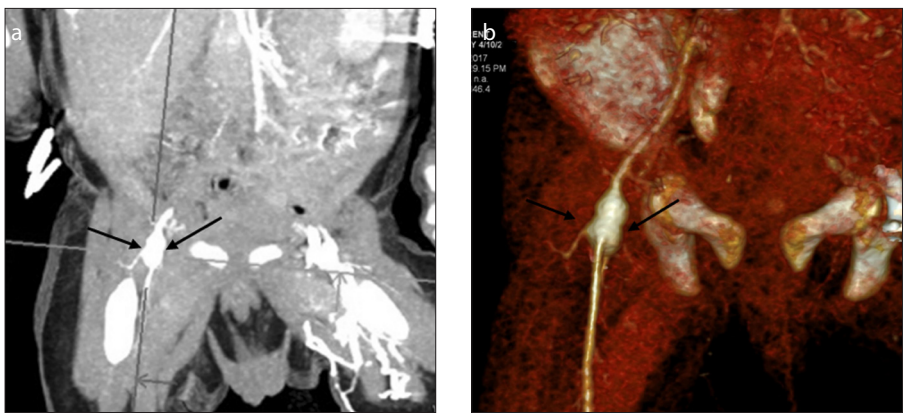

Figure 8. a, b. A right femoral artery aneurysm in a 4-month-old male patient with fever and bilateral conjunctival injection. Coronal MIP CT angiogram **(a)** shows a right femoral artery aneurysm *(arrows)*. Anterior 3D volume-rendered CT image **(b)** shows a right femoral artery aneurysm *(arrows)*.

Peripheral arteries, not including abdominal and thoracic arteries, can be demonstrated with ultrasonography (3, 7). All of these peripheral arteries can be scanned with magnetic resonance angiography (MRA). However, the difficulties of showing the coronary artery by MRA are known. Contrary to CT angiography, MRA has low resolution in small aneurysms. In addition, MRA requires anesthesia and more time in young children (8). Invasive catheter angiography can detect both systemic and coronary arteries. However, invasive angiography has some disadvantages such as its invasiveness, possible complications, requirement of anesthesia and radiation exposure (9, 10).

High-pitch low-dose CT angiography is an impressive alternative imaging modality for patients with AKD. CT angiography is free-breathing, does not require anesthesia and does not depend on the user (8, 11). This technique can detect aneurysms that are missed by echocardiography (22) and can also detect more distal aneurysms that are identified with ultrasonography (13) along with SAA in any location in young children. Further, it is a noninvasive technique, and CT angiography can detect vascular aneurysms, occlusions and stenoses previously identified by invasive angiography. In addition, with this modality, the wall of the vessel can be assessed in addition to its lumen. Traditional CT angiography, unlike low dose CT angiography, is more harmful especially for young children. During routine pediatric body CT examinations, the radiation burden is 4.4–8.5 mSv (14). In our study, we used the SAFIR denoising method on work stations that maintain spatial resolution and retain diagnostic quality images. With this technique, the effective radiation dose can be decreased (1.4–4.3 mSv, main 1.9 mSv), and high-resolution images can be obtained easily and quickly (15).

Advice on high-pitch low-dose CT angiography mentions not only the diagnosis of KD but also discusses the follow-up for patients with KD. According to the American Heart Association and the Japanese Circulation Society, patients without aneurysms should be assessed with electrocardiography and echocardiography for cardiovascular risk for 5 years after disease onset. Patients with aneurysms can be examined with CT angiography (3, 16). In the Dietz et

al. (17) study, the authors suggested that high-pitch low-dose CT angiography can be used at both early and late stages of monitoring for the development of stenosis.

Our study has several limitations. First, the sample size was small due to the relatively rare incidence of the disease. Second, we were not able to compare our results with other modalities because we wanted to uncover the efficiency of CT angiography. Although the radiation dose is reduced by high pitch, this imaging technique still requires radiation.

In conclusion, high-pitch low-dose CT angiography can be considered to be a noninvasive, robust and safer diagnostic imaging modality, given that this technique shows aneurysms at any location in AKD.

Conflict of interest disclosure

The authors declared no conflicts of interest.

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