

# Atypical Kawasaki Disease Presenting with Hemiparesis and Aphasia: A Case Report

Ali Nikkhah, MD

Department of Pediatric Neurology,  
Mazandaran University of Medical  
Sciences, Sari, Iran

## Correspondence:

Ali Nikkhah, MD;  
Department of Pediatric Neurology,  
Mazandaran University of Medical  
Sciences, Sari, Iran  
**Tel/Fax:** +98 11 32353061  
**Email:** alinik52@yahoo.com  
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## Abstract

Kawasaki disease (KD) is an inflammatory vasculitis. KD is classified into two groups based on clinical characteristics criteria, namely classic and incomplete. Cerebral vascular abnormality, especially arterial ischemic stroke (AIS) is very rare and unusual in KD. Here, we report a 4-year-old boy who was referred to our tertiary pediatric center with abrupt right hemiparesis and aphasia. At admission time, he had febrile illness and was toxic. On physical examination, we found unilateral left submandibular lymphadenopathy. On neurologic examination, we obtained right sided hemiparesis with hemiparetic gait and aphasia. His deep tendon reflexes (DTRs) of right extremities were exaggerated and his sensory system was intact. Based on these features, some differential diagnoses were suggested, such as acute encephalitis with focal signs, brain abscess, cerebral vasculitis, hemorrhagic insults, and ischemic stroke, etc. After a complete evaluation, especially brain MRI and MRA, our diagnosis was arterial ischemic stroke (AIS) following atypical KD. Based on these findings, we administered intravenous immunoglobulin (IVIG 2 gm/kg) and oral high dose aspirin (100 mg/kg/d). He responded to these anti-inflammatory treatments dramatically.

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## What's Known

- Kawasaki disease (KD) is an inflammatory disease of small- and medium-sized arteries in infants and young children.
- Coronary arteries involvement is the most common complication, especially in atypical form of KD disease.

## What's New

- Cerebral arteries involvement, especially as arterial ischemic stroke (AIS) is very rare and unexpected complication in KD that is presented in our patient with incomplete (atypical) form of this disease.

## Introduction

Kawasaki disease (KD) is an acute inflammatory disorder of blood vessels that involves small and medium-sized arteries in patients and almost always affects children younger than 5 years of age.<sup>1,2</sup> Involvement of some organs, such as the heart (coronary aneurysms) is common and expected in KD. However, cerebral vascular involvement is very rare and unexpected. KD is a clinical diagnosis and physicians should be able to differentiate it from other similar vasculopathies in children.<sup>2</sup> Based on the characteristics of clinical manifestations, KD is classified into two groups of classic (typical) and incomplete (atypical). Main criteria are prolonged fever (five days or more), oral cavity erythematous changes (cracked lips, strawberry tongue), bilateral conjunctival involvement, skin rash (polymorphous rash involving trunk, extremities, and perineal regions), erythematous changes in the hands and feet, and cervical lymphadenopathy (usually unilateral).<sup>3-5</sup> In atypical (incomplete) KD, patients have fever (even less than five days) with only two or three of the cardinal clinical manifestations.<sup>6</sup> Most

pediatricians believe that KD is a challenging disorder and its diagnosis is difficult particularly in young children with atypical features. We could only find one similar Chinese case report in the literature. Here, we report an atypical (incomplete) KD who presented with fever for 3 days and sudden onset of hemiparesis and aphasia (arterial ischemic stroke with middle cerebral artery occlusion).

### Case Presentation

A 4-year-old male child was referred to our neurology ward of tertiary pediatric center with sudden onset of right hemiparesis and aphasia. He had fever (up to 38 °C) for 3 days. At admission time, he was ill and toxic. His axillary temperature was 38.5 °C. His other vital signs included heart rate of 112/min, respiratory rate of 28/min and blood pressure of 90/60 mmHg. On physical examination, we found unilateral cervical lymphadenopathy (1×1 cm, left submandibular). There was no evidence for tonsillitis and/or pharyngitis. No skin rash was found. On neurologic examination, we obtained right-sided hemiparesis with hemiparetic gait and aphasia. Deep tendon reflexes (DTRs) of the right upper and lower extremities were brisk but DTRs of the left extremities were normal. The sensory system was intact. Initial white blood cell count (WBC) 12600/mm<sup>3</sup> (Neut 66%, Lymph 32%, Eos 2%), hemoglobin (Hb) level 11.4 mg/dl, platelet 465000/mm<sup>3</sup>, C-reactive protein (CRP) 110 mg/dl, and erythrocyte sedimentation rate (ESR) was 92 mm/h. His biochemistry panel was unremarkable. Antistreptolysin O (ASO) titer and liver function tests were normal. Antinuclear antibody (ANA) was negative. Furthermore, his urine and blood cultures were negative. We did not find neck rigidity and meningismus in this patient. We did not perform lumbar puncture and CSF analysis due to focal neurologic deficit. However, we started parenteral antibiotic (cefotaxime 200 mg/kg/day and vancomycin 60 mg/kg/day) based on pediatric infectious consult. We also planned neuroimaging study, including brain magnetic resonance imaging (MRI), magnetic resonance angiography (MRA), and magnetic resonance venography (MRV) with the impression of acute cerebral vascular insult. The next day, our patient not only remained febrile but also became irritable. On the 3<sup>rd</sup> day of admission time, his temperature was 39.5 °C and we found obvious cracked red lips (figure 1). We then repeated CBC, ESR, and CRP. With an impression of incomplete KD, we requested pediatric cardiologist consult. The results of

these laboratory studies were similar to those at the admission time, except for CRP and ESR that were 145 mg/dl and 98 mm/h, respectively. A pediatric cardiologist performed transthoracic echocardiography and reported some bright spots on the right coronary artery with the suggestion of thrombi or microaneurysms. This case report is compiled after obtaining informed consent from the patient's parents.

Brain MRI revealed high signal abnormalities in the left basal ganglia and external capsule that was compatible with arterial ischemic stroke (AIS) due to emboli or thrombosis (figure 2). In MRA, we found near complete obliteration of the left middle cerebral artery by thrombi/emboli that strongly suggested AIS (figure 3). Brain MRV was normal.

Based on these findings, we administered intravenous immunoglobulin (IVIG 2 gm/kg) and oral high-dose aspirin (100 mg/kg/d) followed by the discontinuation of antibiotics. Two days later, the child was afebrile and after 5 days, we changed oral aspirin to 5 mg/kg/d. On the 12<sup>th</sup> and the 26<sup>th</sup> day of follow-up, his aphasia and hemiparesis were respectively eliminated. We tapered off aspirin on the 60<sup>th</sup> day of the follow-up. Now (six months after admission time), he is completely healthy with normal echocardiogram and brain MRA except mild gliosis in MRI after AIS due to atypical KD.



Figure 1: Cracked red lips in our patient (with permission).

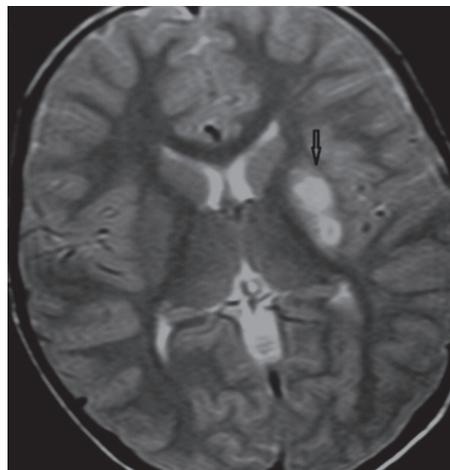


Figure 2: T<sub>2</sub>W MR shows hyperintensities in the left basal ganglia and external capsule (arrow).



**Figure 3:** Magnetic resonance angiography (MRA) shows occlusion of the left middle cerebral artery (red arrow) that is compatible with arterial ischemic stroke (AIS).

### Discussion

KD is an acute febrile inflammatory vasculopathy with unknown mechanism that involves small and medium-sized arteries and can involve any organ with this vascular condition.<sup>1,2</sup> In typical (classic) KD, children have fever for five days or more with at least four out of the five clinical manifestations.<sup>3</sup> These features include the involvement of oral cavity and lips (erythematous fissured lips and strawberry tongue), skin involvement (maculopopular eruptions), bilateral non-purulent conjunctivitis, changes at extremities after 2-3 weeks, and cervical lymphadenopathy.<sup>3-5</sup> In atypical (incomplete) KD, patients have fever (even less than five days) with only two or three of the cardinal clinical manifestations.<sup>6</sup> Atypical KD is more common in infants and is unusual in preschool children.<sup>7,8</sup> With due attention to lack of specific diagnostic test for KD, it is difficult to propose this disease, especially in atypical forms. Some laboratory findings in KD include marked elevated ESR, increased CRP, hypoalbuminemia, elevated hepatic transaminases and leukocytosis.<sup>8</sup> Inasmuch as KD is a systemic vasculitis, involvement of multiple organs is expected. Coronary abnormalities such as thrombi or aneurysms are the most common organ involvement in KD,<sup>9,10</sup> whereas musculoskeletal and gastrointestinal involvement are very rare.<sup>11</sup> We found an interesting case report in the literature about parapharyngeal abscess (PPA)-like lesion in a 3 years old boy in Asia.<sup>12</sup> PPA-like lesion is extremely rare in KD.<sup>12</sup> On the contrary, cerebral arteries abnormality in KD is very rare and we only found one case report about KD complicated with arterial ischemic stroke (AIS) in medical literature (Chinese article).<sup>13</sup> We did

not find another reasonable diagnosis that could explain our patient's condition. All infectious disorders that mimic KD were excluded. Appropriate parenteral antibiotics were ineffective and our patient was afebrile after the administration of IVIG and high-dose aspirin. Therefore, our final diagnosis was incomplete KD. Because of hemiparesis and aphasia, we evaluated the patient to detect unilateral cerebral lesions and found the left middle cerebral artery obliteration leading to the right hemiparesis and aphasia. Other investigations (lipid profile, coagulation profile, metabolic panel, and hematologic tests) were normal.

### Conclusion

KD is an age-specific vasculitis with unknown etiology in infants and young children. All pediatricians should keep in mind that KD could present with different features and diagnosis of atypical KD is difficult. Physicians, especially pediatricians, have to notice the rare and unusual manifestations of KD particularly in infants and young children with atypical features of this disorder. We can prevent potentially dangerous complications (coronary aneurysms and arterial ischemic stroke) with early diagnosis and treatment.

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**Conflict of Interest:** None declared.

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