Clinical Case Report

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Kawasaki disease complicating bilateral facial nerve palsy and giant coronary artery aneurysms

A case report

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Abstract

Rationale: Kawasaki disease (KD), which is also known as mucocutaneous lymphnode syndrome, is a vasculitic disease and involves multi-system disorder with various clinical manifestations. KD is specific predilection for the coronary arteries and is the most common cause of childhood-acquired heart disease in developed countries. KD is rarely complicated by cranial nerve VII palsy.

Patient concerns: This report described a 7-month-old infant who suffered from bilateral infranuclear facial nerve palsy (FNP) and multiple coronary artery aneurysms (CAAs) in parallel with KD. The patient had an intermittent fever for 18 days and had a medical history of bilateral conjunctival injection, strawberry tongue, reddened lips, and perianal excoriation. Physical examinations revealed fever (38.5°C), fingertips desquamation of the skin, and left cervical lymphadenopathies.

Diagnosis: The diagnosis of KD is based on the presence of clinical features of persistent fever (≥5 days) together with polymorphous exanthema, cervical lymphadenopathy, non-purulent conjunctival injection, changes of the lips, oral cavity, and extremities. An echocardiogram has showed a beaded sample dilatation of all coronary arteries, in addition to aneurysms of the middle of the right coronary artery (6.2 mm in diameter; 14.5 *Z* score), and the left coronary artery (5.4 mm in diameter; 9.4 *Z* score). The physical examinations revealed incomplete closure of both eyes and bilateral drooping of the mouth, suggesting a bilateral infranuclear FNP.

Interventions: The patient received intravenous immunoglobulin (IVIG) (2 g/kg) with high-dose aspirin according to the clinical guidelines.

Outcomes: Her fever finally resolved after 2 days' IVIG. All inflammatory indexes returned to normal or near-normal levels prior to discharge. However, the echocardiogram remained unchanged and the patient's facial nerve palsies had not recovered.

Lessons: FNP in KD is uncommon. Yet, it may be a marker of disease progression. One should be aware of the diagnosis of KD when children suffer from high fever, FNP, and even with incomplete clinical features.

Abbreviations: CAAs = coronary artery aneurysms, FNP = facial nerve palsy, IVIG = intravenous immunoglobulin, KD = Kawasaki disease.

Keywords: coronary artery aneurysms, facial nerve palsy, Kawasaki disease

1. Introduction

Kawasaki disease (KD), as a multisystem vasculitic disorder, was first described by Kawasaki,[1] in which coronary artery lesion is the most important complication. Prolonged fever for at least 5 days and presence of at least four in five main clinical criteria are required for KD diagnosis.[2] Several neurological complications have been reported in patients with KD including lethargy, aseptic meningitis, cerebral infarction, facial nerve palsy, and acute demyelinating lesions of the upper thoracic spine.[3] KD accompanied with FNP was first reported in 1974 by Murayama and these patients may be associated with severe clinical progression and high incidence of coronary artery disease.[4]

We describe a 7-month-old girl, who presented with bilateral infranuclear FNP, which the diagnosis was delayed until 18 days after the illness.

2. Case report

A 7-month-old girl, developmentally normal, was admitted to our hospital with an 18-day of intermittent fever, 4-day of drooping of the left side of her mouth, and 1-day of keeping a poker face. She had a history of bilateral conjunctival injections, strawberry tongue, reddened lips, and perianal excoriation. After 14 days, the patient was unable to shut her left eye and experienced drooping of the left side of her mouth. Seventeen days later, the patient was noted to be expressionless and had bilateral drooping of the sides of her mouth. Physical examinations revealed fever (38.5°C), lethargy, fingertips desquamation of the skin, left cervical lymphadenopathy, and

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XY and XL contributed equally to the work.

Informed consent: The written informed consent was obtained, and the patient's information was anonymized and de-identified.

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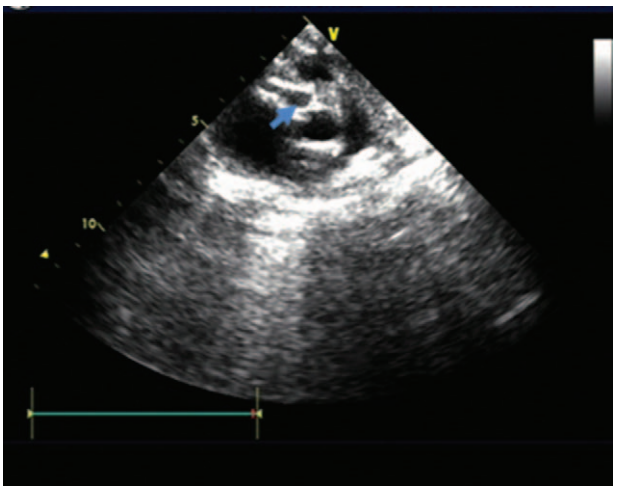

Figure 1. The echocardiographic image showing the aneurysm in the right coronary artery (the arrow).

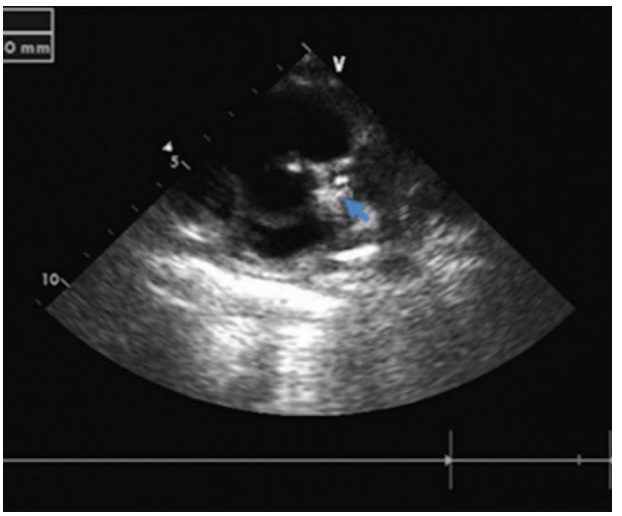

Figure 3. The echocardiographic image showing the aneurysm and mural thrombus in the aneurysm (the arrow).

bilateral peripheral FNPs without other cranial nerve defects. Laboratory findings revealed normocytic–normochromic anemia (9.9 g/dl), leukocytosis (18.11×10^9/L), thrombocytosis (828×10^9/L), high C-reactive protein level (69.6 mg/dl), and erythrocyte sedimentation rate (120 mm/h) with a normal urine examination and chest radiograph. Lumbar puncture revealed that white blood count was 36×10^6/L, and glucose concentration and protein concentration were normal. An echocardiogram showed a beaded sample dilatation of all coronary arteries, in addition to aneurysms of the middle of the right coronary artery (6.2 mm in diameter; 14.5 *Z* score, Fig. 1) and the left coronary artery (5.4 mm in diameter; 9.4 *Z* score, Fig. 2). Her medical history, physical and laboratory examinations converged to complete KD and she received intravenous immunoglobulin (IVIG) (2 g/kg) with high-dose aspirin according to the clinical guidelines 19 days after illness. With the above measures, her fever finally resolved after 2 days' IVIG. All inflammatory index turned out to be normal or near-normal levels prior to discharge. However, the echocardiogram remained unchanged. Low dose daily aspirin and warfarin were orally delivered. The patient was re-examined regularly. Three months after discharge, the echocardiogram was performed and showed that the maximum diameter of the left main coronary artery was up to 5.9 mm and that of right main coronary artery was up to 9.5 mm. Furthermore, 6 months after post-discharge, the echocardiogram showed the lumen diameter began to decrease. Unfortunately, when the child was followed up to 1 year and 7 months, it was found that there was a thrombus in the left coronary artery. About 2.5 years after post-discharge, the echocardiography showed aneurysms of the right main coronary artery (3.6 mm in diameter; 5.2 Z score), of the left main coronary artery (6.5 mm in diameter; 9.0 Z score, Fig. 3), and mural thrombus in the left aneurysm. Her right FNP recovered nearly normal, and unfortunately the left FNP has not been fully recovered until now.

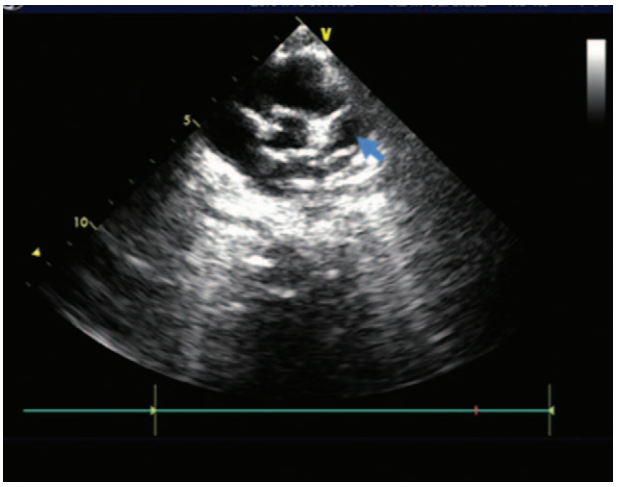

Figure 2. The echocardiographic image showing the aneurysm in the left coronary artery (the arrow).

3. Discussion

The diagnosis of KD[2] is based on the presence of clinical features of persistent fever (≥5 days) together with polymorphous exanthema, cervical lymphadenopathy, non-purulent conjunctival injection, changes of the lips, oral cavity, and extremities. Complete KD is defined as fever and ≥4 out of above 5 symptoms. Neurological complications of KD include irritability, lethargy, aseptic meningitis, ataxia, seizures, focal encephalopathy, cranial nerve palsies, cerebral infarction, transient hemiplegia, and acute demyelinating lesions of the upper thoracic spine.[3,4] FNP is one of the neurological complications of KD. The consensus has not been drawn regarding the exact incidence of facial nerve palsy in patients with KD. Only 41 cases have been reported in the literature,[4] concluding that patients tend to be 18-month-old or less (86.1%), in which 63.9% were under 12 months and the median onset of facial palsy is 16 days in the course. The facial nerve palsy pathogenic mechanisms may be the dysfunctions of both ischemic vasculitis of the arteries and immunologic mechanisms associated with the facial nerve.[4,5] Although spontaneous remission of facial nerve palsy occurs in 1 week to 3 months, IVIG therapy seems to improve recovery,[4] being the most effective treatment for the first 10-d of KD[2].

FNP could be a sign of significant inflammatory burden that leads to high occurrence of CAAs.[4] CAAs occurred in more than

half of the patients with FNP reported by Poon.[6] The use of *Z* scores allows for evaluating the severity of coronary artery dilation by correcting for body surface area and allows for comparisons across time and populations. A classification scheme based solely on *Z* scores has been proposed in recent KD guidelines[7]: CAAs are considered small if *Z* scores are 2.5 to <5; medium if *Z* scores are >5 to <10; and large or giant if *Z* scores are either >10 or >8 mm in diameter.

The current treatment for KD is a high dose of 2 g/kg IVIG, given within the first 10 days after disease onset.[5] Apart from IVIG, high-dose aspirin is advised by the American Heart Association. The majority of patients respond rapidly to IVIG, yet approximately 10%–20% of all patients do not respond well or have recurrent fever within 36–48 h after IVIG.[7] A second dose of IVIG is commonly advised, particularly in patients who have partially responded. Furthermore, corticosteroids are also advised and Warfarin is recommended if giant aneurysms are present.[2,7] Other secondary treatment possibilities are infliximab (TNF-α inhibitor), cyclosporine (calcineurin inhibitor), and statins, yet efficacy remains to be further investigated.[5,7]

Many CAAs regress to a normal-sized lumen mainly within the first 5 years.[8] While the lumen diameter may return to normal, it has become apparent that the vascular wall is often still damaged.[7] Giant CAA may have serious long-term consequences. Apart from thrombosis within the CAA and perfusion abnormalities after CAA, there is an increased risk of stenosis just proximal or distal to the CAA.[7] The risk for patients with small- to medium-sized CAA is unclear.[8] All aneurysms that reduced in size to a normal luminal dimension were originally small or medium in size.[7,8]

In this patient, according to her medical history and physical examinations, KD was "complete" and her FNP was bilateral. The patient's left FNP has not yet fully recovered up to now. This feature of long term non-recovery of FNP in KD has not been reported in published literature. Before being admitted to our hospital, her bilateral facial nerves and coronary arteries had been damaged. Although her clinical manifestations gradually relieved after IVIG, the lesions of the coronary artery persisted due to the worsening of disease at follow-up, which supports the observation of excessive inflammatory burden. Thus, delayed diagnosis and treatment might result in a higher rate of coronary artery aneurysms.

In summary, FNP in KD is uncommon. Yet, it may be a marker of disease progression. One should be aware of the diagnosis of KD when children suffer from high fever and FNP, even with incomplete diagnostic features.

Author contributions

Data curation: Yongchun Wang.
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Software: Mengjiao Wang.
Supervision: Li Sun.
Validation: Li Sun.
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Writing – review & editing: Xia Yu.

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