

# Facial nerve palsy and Kawasaki disease

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**We report on a case of facial nerve palsy associated with Kawasaki disease in a 2-year-old boy. Facial nerve palsy is one of the rare neurological manifestations of Kawasaki disease. Twenty-seven other cases that have been reported in the literature are reviewed. There is a high incidence of coronary artery aneurysm (52%) and a female predilection in patients with Kawasaki disease. The facial palsy associated with the disease is self-limiting. Recovery is spontaneous in surviving patients, although the use of intravenous immunoglobulin may be able to hasten the recovery.**

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## Introduction

Kawasaki disease (KD), or acute infantile febrile mucocutaneous lymph node syndrome, was originally described as a distinct clinical entity in Japanese children by Kawasaki in 1967.<sup>1</sup> The first report in the English literature appeared in 1974.<sup>2</sup> In the United States, 3000 cases of KD are diagnosed each year; 80% of cases occur in children younger than 5 years.<sup>3</sup> There is a slight increase in the prevalence of KD in winter and spring.<sup>3</sup> As of 1992, The Japan Kawasaki Disease Research Committee had already documented over 116 800 cases of KD in Japan, with a male to female ratio of 1.5:1.<sup>3</sup> Apart from irritability, lethargy, and aseptic meningitis, neurological complications of KD are uncommon. Facial nerve paralysis associated with KD was first reported by Murayama in 1974.<sup>4</sup> It has since been reported in the literature in only 26 other children (Table). In this report, we describe and discuss the clinical significance of a case of facial nerve paralysis occurring in a 2-year-old boy with KD.

## Case report

A 2-year-old Chinese boy was admitted to the Queen Elizabeth Hospital (QEH) in April 1999 with a 6-day history of fever, skin rash, left cervical lymphadenopathy, and bilateral non-exudative conjunctivitis. The

patient had a fever (temperature, 40°C) and was irritable. His conjunctivae were injected without exudate; his oropharynx was erythematous; and his lips were swollen, reddened, and cracked. A tender, enlarged cervical lymph node of 3 cm in diameter was palpable just below the angle of the left mandible. There were confluent, erythematous, papular rashes over the face, trunk, and limbs. Results from the cardiovascular examination were normal. The patient's peripheral white blood cell count was  $14.3 \times 10^9$  /L (normal range,  $3.2-9.8 \times 10^9$  /L), with 86% neutrophils and 8% lymphocytes. The haemoglobin level was 120 g/L (normal range, 136-172 g/L) and the platelet count was  $356 \times 10^9$  /L (normal range,  $150-450 \times 10^9$  /L); the erythrocyte sedimentation rate was 80 mm/h (normal range, 0-30 mm/h). The level of serum alanine aminotransferase was 40 U/L (normal range, 0-35 U/L). Urinalysis revealed one white blood cell per high-power field. The anti-streptozyme O titre was <200 IU/mL. Blood, throat, and urine cultures gave negative results.

A course of intravenous cefuroxime was started empirically after hospital admission, but intermittent fever persisted. Serological tests for the presence of Epstein-Barr virus and auto-antibodies gave negative results, as did the stool culture. Results from electrocardiography and echocardiography were normal. A clinical diagnosis of KD was made on day 2 of hospital admission. Cefuroxime treatment was discontinued, and intravenous immunoglobulin 2 g/kg and aspirin 30 mg·kg<sup>-1</sup>·d<sup>-1</sup> were given. Fever subsided 1 day after the initiation of immunoglobulin and aspirin therapy. Aspirin was then changed to dipyridamole as soon as it became known that the patient had glucose-6-phosphate dehydrogenase

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deficiency. He was discharged home on day 7 (13 days from the onset of fever) and was followed up at the cardiac out-patient clinic of the QEH. The patient was readmitted to hospital 4 days later for the acute onset of facial asymmetry. Neurological examination showed lower motor-neuron-type palsy of the right facial nerve. No other neurological deficit or cranial nerve palsy was detected. Computed tomography and magnetic resonance imaging of the brain gave normal results. Lumbar puncture and electroencephalography were not performed. The facial palsy recovered completely within 1 month, following conservative treatment. Repeated echocardiograms showed normal coronary arteries.

## Discussion

The exact aetiology of KD is unknown. According to the fourth edition of the Japanese diagnostic guidelines of KD,<sup>5</sup> a diagnosis is made on the recognition of five or six main symptoms (Box). If only four main symptoms are found, but echocardiography or angiography reveals coronary arterial changes, the Japanese guidelines also recognise the disease as KD. The Japanese criteria differ slightly from the North American diagnostic criteria, in which the presence of fever for at least 5 days and at least four other symptoms are necessary to establish the diagnosis.<sup>6,7</sup> Patients with fever and fewer than four other main symptoms can also be classified as having KD when coronary artery disease is detected.

Facial nerve palsy associated with KD is rare and was first added as one of the neurological symptoms and signs of KD in the Japanese diagnostic guidelines in 1984.<sup>8</sup> The occurrence of facial nerve palsy has been reported in 27 patients with KD (Table). The

age of onset of KD in these patients ranged from 3 to 25 months, but most of them were younger than 20 months. There is a 1.4:1 female predominance (in contrast to the 1.5:1 male predominance for KD in general). All facial nerve palsies were unilateral, with the left side being more commonly affected than the right. Among the 22 children who had echocardiography performed, 12 (54%) were found to have coronary artery aneurysms, compared with 25% of all cases of KD (without intravenous immunoglobulin infusion). The higher incidence of coronary artery aneurysm suggests that the occurrence of facial nerve palsy is an indicator of an increased severity of disease. The facial palsy is usually transient, with the duration ranging from 2 days to 3 months. Spontaneous and complete recovery without any treatment and without sequelae was observed in all but two reported cases. Two children died of complications related to cardiac involvement. In these two patients, the facial palsies persisted until death.

Amano and Hazama<sup>9</sup> have described the histological results from investigations of the nervous system of 30 patients with KD. Ganglionitis and neuritis of cranial and peripheral nerves were seen. Examinations of the brain showed endoarteritis, periarteritis, aseptic choriomeningitis, and leptomeningitis. These inflammatory lesions were similar to those found in the other affected organs. Facial nerve palsy is thus thought to be the result of vasculitis involving the facial nerve at a level beyond the facial nucleus. The pertinent feature of irritability may be associated with the involvement of the central nervous system.

At the QEH, this was the first documented case of KD complicated by lower motor-neuron facial palsy. To the best of our knowledge, this is also the first case reported in Hong Kong. Fifty-seven and 34 new cases of KD were seen at the QEH in 1997 and 1998, respectively (unpublished data, 1999). Apart from the patient described in this report, none of patients with KD experienced major neurological complications other than transient irritability, lethargy, and aseptic meningitis. Kawasaki disease is well recognised in local paediatric units and there is a low threshold for giving intravenous immunoglobulin for suspected or diagnosed cases.<sup>10</sup> Because the facial nerve palsy is likely to be caused by an inflammatory vasculitic process that affects the facial nerve, it has been postulated that the palsy may be ameliorated by giving intravenous immunoglobulin.<sup>11</sup> Bushara et al<sup>11</sup> reported a case of facial nerve palsy in a 12-week-old boy with KD who had complete resolution of facial nerve palsy within 36 hours of the administration of intravenous

### Diagnostic guidelines of Kawasaki disease<sup>4</sup>

#### Main symptoms

- (1) Fever persisting for 5 days or more
- (2) Changes in peripheral extremities:
  - initial stage: reddening of palms and soles, indurative oedema
  - convalescent stage: membranous desquamation from the fingertips
- (3) Polymorphous exanthema
- (4) Bilateral conjunctival congestion
- (5) Changes in lips and oral cavity: reddening of lips, strawberry tongue, diffuse injection of oral and pharyngeal mucosa
- (6) Acute non-purulent cervical lymphadenopathy

The presence of at least five of the six symptoms is required for a diagnosis of Kawasaki disease; patients with four main symptoms can be diagnosed as having Kawasaki disease when coronary aneurysm is shown by echocardiography or coronary angiography.

**Table. Characteristics of 28 patients with facial nerve palsy associated with Kawasaki disease\***

Patient	Age (months)	Sex	Onset of palsy (days)	Side affected	Duration (days)	Cardiovascular involvement	Immunoglobulin therapy
1	3	M	1.25	L	2	CAA <sup>†</sup>	After facial palsy
2	7	F	17	R	30	None	Before facial palsy
3	10	F	10	R	90	None	None
4	3	F	12	L	7	CAA	None
5	13	F	16	L	11	None	None
6	8	M	2	R	11	None	None
7	6	F	16	L	60	None	None
8	9	F	19	R	60	None	None
9	13	F	12	L	16	CAA	None
10	15	F	16	R	13	CAA	None
11	7	F	16	L	12	CAA	None
12	8	M	14	R	16	CAA	None
13	6	M	28	L	28	CAA	None
14	11	M	18	L	20	CAA	None
15	9	F	10	R	10	None	None
16	9	F	7	L	60	CAA	None
17	6	F	18	L	21	None	None
18	25	F	16	R	60	CAA	None
19	14	F	22	R	60	None	None
20	-	-	-	-	7	-	-
21	-	-	-	-	7	-	-
22	13	M	-	-	-	-	-
23	-	-	-	-	Until death	-	-
24	7	M	37	L	26	None	None
25	-	-	-	-	-	-	-
26	13	M	17	L	Until death	CAA	None
27	3	M	12	L	5	CAA	None
28 <sup>‡</sup>	24	M	17	R	32	None	Before facial palsy

\* References available from the corresponding author; some data for patients 20-33 and 25 were not provided in the original references

<sup>†</sup> CAA coronary artery aneurysm

<sup>‡</sup> This case

immunoglobulin. The precise mechanisms by which intravenous immunoglobulin may work remain unknown. The modulation of the synthesis and release of pro-inflammatory cytokines may play a role.

## Conclusion

Kawasaki disease is a paediatric febrile vasculitic disease and may affect any organ, although it seems to have a predilection for coronary vessels. Irritability, which signifies the involvement of the central nervous system, is a common and constant feature of KD. Facial nerve palsy associated with KD is uncommon and may indicate an increased risk of coronary artery involvement. It is important to bear in mind the diagnosis of KD in any child with unexplained high fever and facial nerve palsy. The palsy normally resolves spontaneously and completely. More data are required to demonstrate if intravenous immunoglobulin helps treat the neurological complications of KD.

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