Cardiology in the Young

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Coronary artery aneurysm and facial drooping in a infant with Kawasaki disease

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Abstract

Kawasaki disease is the leading cause of acquired heart disease in infants and young children. Kawasaki disease that manifests as facial nerve palsy is extremely rare, and the diagnosis is challenging. We report a 4-month-old girl with Kawasaki disease who presented with fever, redness and cracking in the lips and oral cavity, and a right facial nerve palsy. The infant received intravenous immunoglobulin, acetyl salicylic acid, and warfarin. The patient's fever subsided on the following day, and the right-sided facial nerve palsy was relieved a month later.

Brief Report

Cite this article: Yuan Y and Lu N (2020) Coronary artery aneurysm and facial drooping in a infant with Kawasaki disease. *Cardiology in the Young* **30**: 1957–1959. doi: 10.1017/S1047951120002980X

Received: 12 July 2020
Revised: 15 August 2020
Accepted: 26 August 2020
First published online: 25 September 2020

Keywords:
Complication; coronary artery aneurysm; facial nerve palsy; incomplete Kawasaki disease; pediatrics

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Background

Kawasaki disease is an acute, self-limiting febrile vasculitis, most common in infants aged 6 months–2 years. It is challenging to make a diagnosis of incomplete Kawasaki disease, particularly in infants younger than 6 months with fewer clinical manifestations.[1] Various neurologic complications have been described to occur in 1–30% of patients with Kawasaki disease,[2] but facial nerve palsy secondary to Kawasaki disease is an extremely rare (0.9%) and atypical presentation.[3] Patients with Kawasaki disease and facial palsy tend to be under 6 months old, and there is a higher association with coronary artery aneurysm in such patients compared to those without facial palsy who never received treatment.[4] Kawasaki disease associated with facial nerve palsy may indicate increased inflammatory burden, and patients may require additional anti-inflammatory agents and more vigilant echocardiography. Herein, we describe a 4-month-old female who presented with unilateral facial nerve palsy in the acute phase of incomplete Kawasaki disease, in which the diagnosis was delayed until day 10 of the illness.

Case Presentation

A 4-month-old female presented with fever, irritability, and reduced mobility of the right side of the face, in addition to an inability to close the right eye and epiphora for 3 days before admission. The onset of the disease, with fever and a slight cough, was 10 days before admission. Therefore, she was thought to have a respiratory tract infection and was admitted to a regional hospital where she was treated with antibiotics and antipyretics, but there was no improvement. On day 7 of illness, the patient developed a right facial droop, false right ptosis secondary to the inability to raise her right eyebrow, and decreased movement of the right aspect of her mouth. Her personal history revealed a 10-day episode of persistent fever and conjunctival injection that resolved spontaneously 5 days earlier. She also presented with generalised cracking and reddening of the lips and was transferred to our clinic with suspected Kawasaki disease.

Physical examination revealed fever (39°C), cracking and reddened lips, and extreme irritability. On examination, she was noted to have right painless cervical lymphadenopathy, left-sided deviation of the labial commissure, obliteration of the right nasolabial fold, and incomplete closure of the right eye, suggesting a right-sided infranuclear facial nerve palsy (Fig 1). A detailed neurologic assessment did not reveal any more defects. The remainder of the physical examination was unremarkable.

The laboratory tests on the day of admission revealed leukocytosis (36,590/μl) with neutrophilia (29,720/μl), anemia (Hb: 7.8 g/dl), thrombocytosis (1155,000/μl), hypo-albuminemia (2.59 g/dl), elevated N-terminal of the prohormone brain natriuretic peptide level (1130 pg/mL), elevated CRP (139 mg/l), and elevated erythrocyte sedimentation rate (65 mm/h). An echocardiogram was performed showing normal left ventricular function, mild pericardial effusion, and mitral regurgitation, in addition to aneurysms of the right main coronary artery (maximum diameter of 8.5 mm; Z-score 11.62), left main coronary artery (maximum diameter of 5.1 mm; Z-score 6.97), left anterior descending coronary artery (maximum diameter of 3.3 mm; Z-score 4.26), and left circumflex coronary artery (maximum diameter of 2.2 mm; Z-score 3.01), thus confirming the diagnosis of incomplete Kawasaki disease (Fig 2). The electrocardiogram demonstrated sinus tachycardia, and the chest X-ray and cerebral magnetic resonance imaging

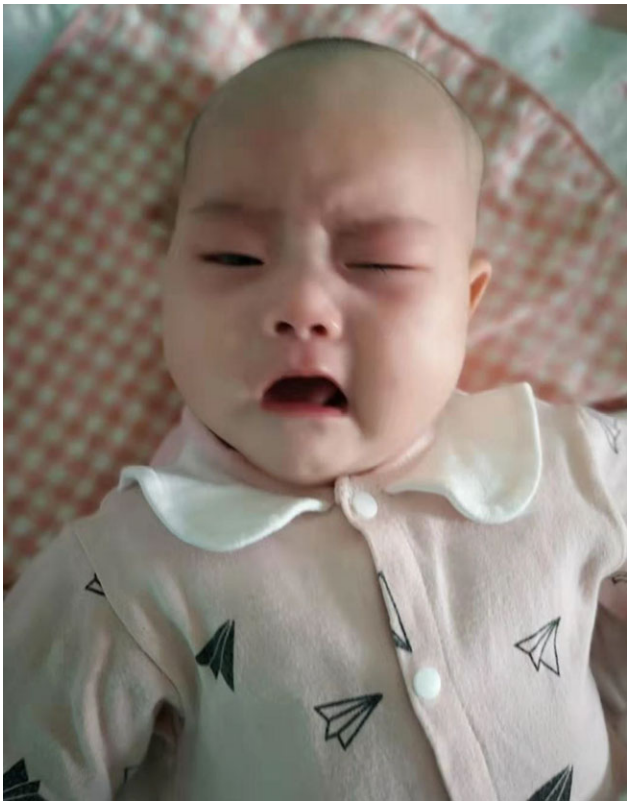

Figure 1. Photograph of the patient showing right-sided facial nerve palsy at presentation.

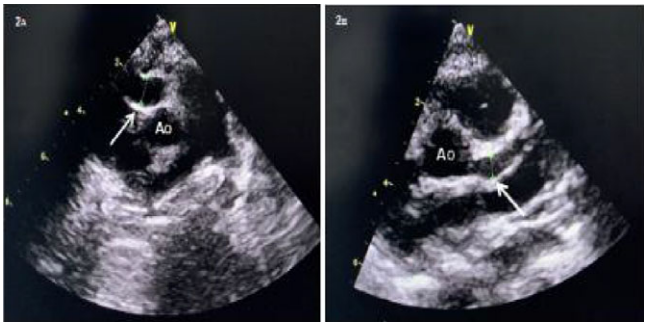

Figure 2. Two-dimensional Doppler echocardiogram. Parasternal short axis plane showing a large-size aneurysm of the left main coronary artery (2A: RMCA; maximum diameter of 8.5 mm; Z-score 11.62), and a small-size aneurysm of the right main coronary artery (2B: LMCA; maximum diameter of 5.1 mm; Z-score 6.97). Ao = aortic root.

were normal. Blood, urine, cerebrospinal fluid, and stool cultures were sterile. Throat and fecal cultures were negative for bacterial and viral pathogens.

Based on the patient's medical history, the physical and laboratory findings were compatible with incomplete Kawasaki disease. She received intravenous immunoglobulin (2 g/kg/16 hours), and acetyl salicylic acid (50 mg/kg/d) was started on the 10th day of illness. The patient's fever and facial nerve palsy resolved within 48 hours after intravenous immunoglobulin treatment. An echocardiogram performed on day 5 of admission revealed similar coronary findings. Enoxaparin was stopped, and anticoagulation was continued with a combination of warfarin and low-dose acetyl salicylic acid after the 2nd week of the treatment.

The patient's inflammatory markers all improved to normal or near-normal levels prior to discharge; all infectious studies returned to negative. Her right facial weakness was unappreciable on the day of discharge (20 days after onset). At the 3-month follow-up after discharge, the right-sided facial nerve palsy had resolved completely, and echocardiography showed improvement in the right coronary artery aneurysms, left coronary artery and left anterior descending coronary artery, and normal left circumflex coronary arteries. The pericardial effusion and mitral regurgitation disappeared completely.

Discussion

Coronary artery aneurysms are the most important and life-threatening complication of Kawasaki disease. Coronary artery complications were significantly more common in infants than in children older than 1 year. It is challenging to make a diagnosis of incomplete Kawasaki disease, particularly in infants younger than 6 months with fewer clinical manifestations. Moreover, the rate of incomplete Kawasaki disease was also higher in infants aged 6 months. Chang et al[5] showed a higher rate of incomplete Kawasaki disease and coronary complications in Kawasaki disease patients <6 months than in older patients, and Rosenfeld et al[6] noted that Kawasaki disease patients younger than 6 months had a higher risk of developing coronary artery aneurysm. Many of these patients were outside the expected age range for Kawasaki disease, and diagnosis and treatment were often delayed.

The neurological complications of Kawasaki disease have been well described, but apart from extreme irritability, they are quite rare.[7] However, it has been reported that facial nerve palsy in these patients may be associated with more severe clinical progression and a higher incidence of coronary aneurysms. It is of great concern that >50% of patients with Kawasaki disease and facial nerve palsy develop coronary artery aneurysm.[8] We speculate that facial nerve palsy may be an indicator for evaluating the severity of Kawasaki disease. The mechanism of facial nerve palsy in Kawasaki disease remains unclear, but it may be considered to be a part of the spectrum of immune activation consequences in Kawasaki disease. It is likely that both ischemic vasculitis of the arteries supplying the facial nerve and immunologic mechanisms contribute to facial nerve dysfunction.[9,10]

Facial nerve palsy usually presents in females (female:male ratio 1.4:1) and infants <18 months of age (86%),[8,9] which is consistent with our report. Most reported cases are unilateral left-sided facial nerve palsys (60%), occurring during the convalescent phase (median onset is day 16 of illness); however, the case we reported was a unilateral right-sided facial nerve palsy and occurred on day 7 of the course of disease. Although facial nerve palsys resolved spontaneously within 1 week–3 months, intravenous immunoglobulin therapy seems to have shortened the time to full recovery.[10]

Conclusion

This case highlights that a Kawasaki disease diagnosis should be suspected in any child with prolonged unexplained fever, even with incomplete diagnostic features; moreover, physicians need to be aware of unusual manifestations such as facial nerve palsy. Atypical cases such as this may indicate an increased risk of coronary artery aneurysm because of the delayed diagnosis and greater inflammatory burden; thus, a more aggressive treatment (intravenous immunoglobulin plus steroids) may be necessary.

Acknowledgements. None.

https://doi.org/10.1017/S104795112000298X Published online by Cambridge University Press

Financial Support. This study was funded by the grants from Natural Science Foundation of Jilin Province (No.20200201617JC to Lu Na).

Conflicts of Interest. None.

References

1. Uysal F, Bostan OM, Celebi S, Uysal B, Hamitoglu S, Cil E. Outcomes of Kawasaki disease: a single-center experience. Clin Pediatr (Phila) 2015; 54: 579–584.
2. Gallagher PG. Facial nerve paralysis and Kawasaki disease. Rev Infect Dis 1990; 12: 403–405.
3. Alves NR, Magalhaes CM, Almeida Rde F, Santos RC, Gandolfi L, Pratesi R. Prospective study of Kawasaki disease complications: review of 115 cases. Rev Assoc Med Bras (1992) 2011; 57: 295–300.
4. Yeo Y, Kim T, Ha K, et al. Incomplete Kawasaki disease in patients younger than 1 year of age: a possible inherent risk factor. Eur J Pediatr 2009; 168: 157–162.
5. Chang FY, Hwang B, Chen SJ, Lee PC, Meng CC, Lu JH. Characteristics of Kawasaki disease in infants younger than six months of age. Pediatr Infect Dis J 2006; 25: 241–244.
6. Rosenfeld EA, Corydon KE, Shulman ST. Kawasaki disease in infants less than one year of age. J Pediatr 1995; 126: 524–529.
7. Newburger JW, Takahashi M, Gerber MA, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Pediatrics 2004; 114: 1708–1733.
8. Stowe RC. Facial nerve palsy, Kawasaki disease, and coronary artery aneurysm. Eur J Paediatr Neurol 2015; 19: 607–609.
9. Wright H, Waddington C, Geddes J, Newburger JW, Burgner D. Facial nerve palsy complicating Kawasaki disease. Pediatrics 2008; 122: e783–e785.
10. Poon LK, Lun KS, Ng YM. Facial nerve palsy and Kawasaki disease. Hong Kong Med J 2000; 6: 224–226.

https://doi.org/10.1017/S104795112000298X Published online by Cambridge University Press