Clinical Case Report

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Facial nerve palsy presenting as rare neurological complication of Kawasaki disease

A case report

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Abstract

Rationale: Facial nerve palsy (FNP) is one of the rare neurologic symptoms of Kawasaki disease (KD), associated with a higher incidence of coronary arteries lesions and may be an indicator of more severe disease.

Patient concerns: A 3-month-old male infant with persistent fever, irritability, and facial asymmetry.

Diagnoses: KD with FNP.

Interventions: The infant received intravenous immunoglobulin (IVIG) (2 g/kg/16 hours) and aspirin (50 mg/kg/day) were started on the 8th day of illness.

Outcomes: Fever and FNP resolved within 48 hours after IVIG treatment. The inflammatory markers all improved to normal or near-normal levels before discharge; all infectious studies returned negative. His left facial weakness was unappreciable at day of discharge.

Lessons: FNP associated with KD is an uncommon finding but may indicate an increased risk of coronary artery involvement. KD should always be kept in mind in the differential diagnosis of a child who presents with prolonged unexplained fever, even with incomplete diagnostic features, as well as the need to be aware of unusual manifestations, such as FNP.

Abbreviations: FNP = facial nerve palsy, IVIG = intravenous immunoglobulin, KD = Kawasaki disease.

Keywords: facial nerve palsy, Kawasaki disease

1. Introduction

Kawasaki disease (KD) or mucocutaneous lymph node syndrome is a multisystem vasculitic disease. The diagnosis of KD is made on basis of fever at least 5 days duration with at least 4 of 5 principal criteria: changes in extremities, polymorphous exanthem, bilateral conjunctival injection, changes in lips and oral cavity, and cervical lymphadenopathy.[1] Incomplete KD refers to patients who do not fulfill all the clinical criteria. KD is an acute panvasculitis with a striking affinity for the coronary arteries (CA) and it can affect any organ system in the body. Facial nerve palsy (FNP), first reported in 1974 by Murayama, is a recognized but uncommon manifestation of KD.[2] FNP was added as one of the neurologic symptoms of KD in the Japanese diagnostic guidelines published in 1984.[3] FNP has been associated with a higher incidence of CA lesions and may be an indicator of more severe disease.[4] Clinically, FNP usually presents during the convalescent phase of KD, but may develop any time from the second day of fever until >1 month after the initial KD presentation.[5] FNP is usually transient, lasting from 2 days to 3 months, with spontaneous and complete recovery, except in 2 patients who died of cardiac complications.[6] Treatment with intravenous immunoglobulin (IVIG) seems to shorten the time to full recovery.[7]

2. Case report

We reported the case of a 3-month-old male infant admitted to our clinic with persistent fever, irritability, and facial asymmetry. The onset of the disease, including fever, rhinorrhea, and cough, was 7 days before the admission. Therefore, he was admitted to a community hospital where he benefited from antibiotics and antipyretics, but there was no improvement. On day 8 of illness, he presented with a reduced mobility of the left side of the face, in addition to an inability to close the left eye and epiphora. His personal history revealed an 8-day episode of persistent fever, conjunctival injection, and confluent, erythematous, papular rashes over the face, upper arms and legs, and trunk, which had spontaneously resolved 6 days earlier. As a result, he was thought to have hand-mouth-foot disease and was transferred to our clinic with suspected KD.

On examination, he was noted to be irritable and to have a temperature of 39.5°C. The physical examination revealed right painless cervical lymphadenopathy, right-sided deviation of the labial commissure, obliteration of the left nasolabial fold, and incomplete closure of the left eye, suggesting a left-sided

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Informed consent was obtained from the patient's mother before the publication of this case report.

The authors have no conflicts of interests to disclose.

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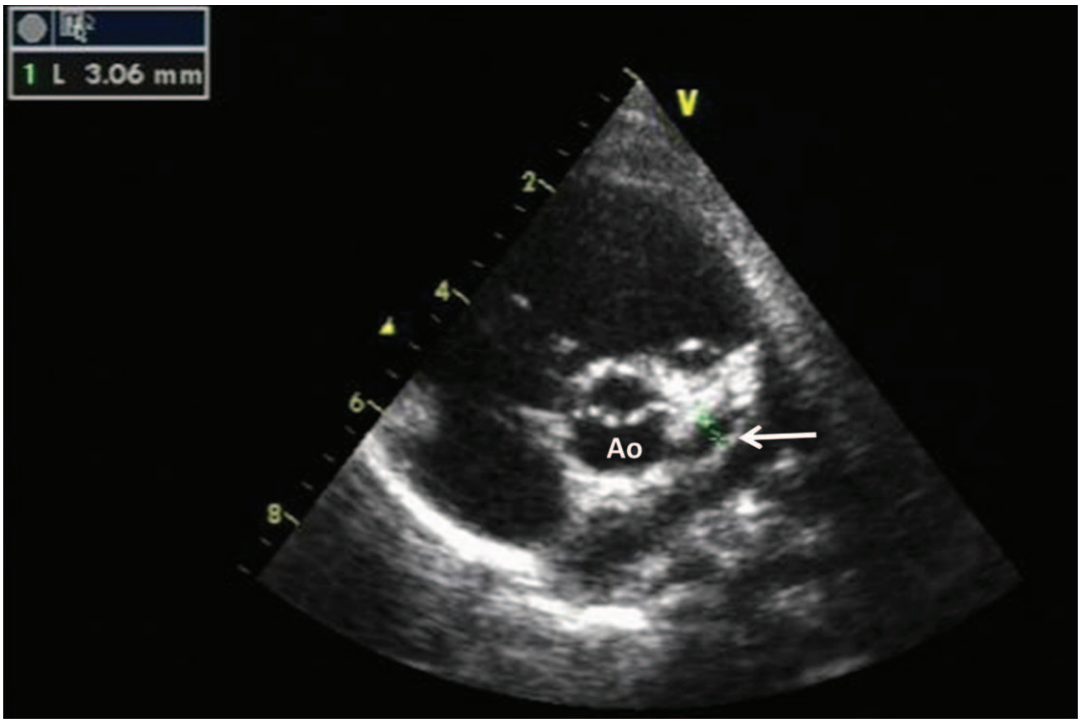

Figure 1. Parasternal short axis echocardiographic image showing dilatation (arrows) in left coronary artery. Ao = aortic root.

infranuclear FNP. A detailed neurologic assessment did not reveal any more defects. The remainder of the physical examination was unremarkable.

Laboratory findings revealed anemia (7.4 g/dl), leukocytosis (28,300/mm^3), thrombocytosis (980,000/mm^3), increased C-reactive protein level (109 mg/dl), and erythrocyte sedimentation rate (95 mm/h). An echocardiogram was performed showing normal left ventricular function, mild pericardial effusion, mitral regurgitation, and left coronary artery dilatation (3.06 mm in diameter) (Z-score 4.98) (Fig. 1). Electrocardiogram demonstrated sinus tachycardia and chest X-ray was normal. The results of cerebral magnetic resonance imaging were normal. Blood, urine, cerebrospinal fluid, and stool cultures were sterile. Throat and fecal cultures were negative for bacterial and viral pathogens. Serologic testing was negative for herpes simplex virus (HSV) types 1 and 2, enterovirus, adenovirus, Mycoplasma pneumoniae, Epstein–Barr virus, cytomegalovirus, and influenza A and B.

Based on the patient's medical history, the physical and laboratory findings were compatible with incomplete KD. He received IVIG (2 g/kg/16 hours) and aspirin (50 mg/kg/day) were started on the 8th day of illness. Fever and FNP resolved within 48 hours after IVIG treatment. An echocardiogram performed on day 4 of admission revealed similar coronary findings.

The patient was discharged on treatment with aspirin (5 mg/kg/day) and close cardiac follow-up. His inflammatory markers all improved to normal or near-normal levels before discharge; all infectious studies returned negative. His left facial weakness was unappreciable at day of discharge (14 days after onset). At follow-up 6 months after discharge, he remained asymptomatic and the repeated echocardiogram showed obvious improvement with normal left coronary arteries. Pericardial effusion and mitral regurgitation had disappeared.

3. Discussion

The age of reported patients with KD and FNP ranges from 3 to 25 months, but the majority were younger than 20 months. The average time of onset of palsy is the 16th day of illness. Median time to resolution is 18 days. There is a 1.4:1 female predominance (in contrast to the 1.5:1 male predominance for KD in general).[8] All FNPs described to date have been unilateral, and were left sided in 2 thirds of the cases.[6] Our patient, a male infant, also had left-side FNP.

The neurological complications of KD have been well described but they are quite rare with only 41 cases of FNP reported in the previous literature. FNP is considered to be a part of the spectrum of immune activation consequences in KD.[9] It is likely that both ischemic vasculitis of the arteries supplying the facial nerve and immunologic mechanisms contribute to the facial nerve dysfunction. Most of FNP cases were defined before the introduction of the use of IVIG therapy in KD. Although FNP resolves spontaneously within 1 week to 3 months, IVIG therapy seems to shorten the time to full recovery. However, it is reported that FNP in KD patients may be associated with more severe clinical progresses and higher incidence of coronary aneurysms, which may reflect late KD diagnosis, more severe inflammation and vasculitis.[1] Notably, there is only coronary artery dilatation without coronary artery aneurysm in our case even though FNP was present, and it may be because of the timely identification and treatment.

The early diagnosis of KD is hindered by the important similitudes between its clinical appearance and that of other infectious diseases. Therefore, in these cases, echocardiography can be helpful. A normal echocardiography within the first 7 days of fever cannot rule out KD, but positive findings can confirm the diagnosis.[10] Incomplete KD usually appears in young infants,

and it must never be mistaken as a mild form of KD.[11] Our case presented with an incomplete form of KD, because, in addition to fever, he had only 3 clinical features: polymorphous rash, bilateral nonexudative conjunctival injection, and right painless cervical lymphadenopathy. Also, similar to the data mentioned above, he was a young infant. Despite this incomplete presentation, the diagnosis in our case was not delayed more than 10 days, and the treatment was initiated 8 days after the onset with both IVIG and aspirin due to the positive echocardiographic findings.

4. Conclusion

FNP associated with KD is an uncommon finding but may indicate an increased risk of coronary artery involvement. KD should always be kept in mind in the differential diagnosis of a child who presents with prolonged unexplained fever, even with incomplete diagnostic features, as well as the need to be aware of unusual manifestations such as FNP.

Author contributions

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References

[1] Poon LK, Lun KS, Ng YM. Facial nerve palsy and Kawasaki disease. Hong Kong Med J 2000;6:224–6.
[2] 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. Circulation 2004;110:2747–71.
[3] Hattori T, Tokugawa K, Fukushige J, et al. Facial palsy in Kawasaki disease. Report of two cases and a review. Eur J Pediatr 1987;146: 601–2.
[4] Wright H, Waddington C, Geddes J, et al. Facial nerve palsy complicating Kawasaki disease. Pediatrics 2008;122:e783–5.
[5] Stowe RC. Facial nerve palsy, Kawasaki disease, and coronary artery aneurysm. Eur J Paediatr Neurol 2015;19:607–9.
[6] Gallagher PG. Facial nerve paralysis and Kawasaki disease. Rev Infect Dis 1990;12:403–5.
[7] Bushara K, Wilson A, Rust RS. Facial palsy in Kawasaki syndrome. Pediatr Neurol 1997;17:362–4.
[8] Rowley AH, Gonzalez-Crussi F, Shulman ST. Kawasaki syndrome. Rev Infect Dis 1988;10:1–5.
[9] Yeung RS. Pathogenesis and treatment of Kawasaki's disease. Curr Opin Rheumatol 2005;17:617–23.
[10] Tizard EJ. Complications of Kawasaki disease. Curr Paediatr 2005;15:62–8.
[11] Jiao F, Jindal AK, Pandiarajan V, et al. The emergence of Kawasaki disease in India and China. Glob Cardiol Sci Pract 2017;2017:e201721.