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A Case Report of Facial Nerve Palsy in Kawasaki Disease

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ABSTRACT

Introduction: One of the most prevalent childhood vasculitis is kawasaki disease (KD), which mainly affects medium-sized arteries throughout the body. This disease usually does not cause many neurological symptoms and only rarely results in facial nerve palsy (FNP).

Case presentation: A 4.6-month-old iranian girl presented due to a prolonged fever. Lab tests showed increased CRP, ESR, ferritin, WBC, and platelet counts, and echocardiography revealed dilated right and left main coronary arteries, without any vegetations, pericardial effusion, and atrioventricular valve regurgitation. The patient was diagnosed with incomplete KD. At the same time, she developed left-sided peripheral FNP. Patient treated with intravenous immunoglobulin and aspirin. The fever resolved after about 36 hours, and the patient was discharged on the 16th day of hospital admission. Follow-up echocardiography demonstrated gradually resolution of her coronary arteries dilation. FNP was recovered completely after about 2 months.

Conclusion: FNP is an uncommon neurological symptom of KD. However, it typically resolves on its own and without any lasting effects in those who survive the illness. Nevertheless, it may indicate a higher likelihood of coronary artery involvement, and as a result, additional anti-inflammatory treatments and more closely monitored echocardiography may be necessary.

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Introduction

Kawasaki disease (KD), also known as acute infantile febrile mucocutaneous lymph node syndrome, is a disease that primarily affects young children and is characterized by inflammation of the medium-sized arteries . It was first identified in Japanese children by Kawasaki in 1967, but it is now found worldwide and is the leading cause of acquired heart disease in developed nations (1). KD can affect children of all ethnic backgrounds, but it is more common in Asian children, and slightly more common in males than females. Approximately 85-90% cases of KD occur in children under 5 years of age (2).

The cause of KD is not yet known, but it is thought to be related to infection,

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autoimmune reactions, and genetic predisposition (3). It can affect many body systems, but neurological involvement is rare. One of the rarest complications of KD is facial nerve palsy (FNP) (4). We present the case of a 4.6-month-old female who was diagnosed with KD, which involved her coronary arteries and resulted in FNP.

Case presentation

A 4.6-month-old iranian girl presented only with a prolonged fever. The physical examination documented fever without any other signs. Laboratory findings revealed WBC count (7.500/mm3), anemia (8.4g/dl), mild thrombocytosis (518.000/mm3), increased C-reactive protein (CRP) level (63 mg/dl) and erythrocyte sedimentation rate (ESR) (46 mm/h). Serum electrolytes, BUN, creatinine, and chest radiograph were normal. She was hospitalized and broad spectrum intravenous antibiotics (ceftriaxone and vancomycin) were started. Abdominal sonography was normal except for mild edema and thickening of the gallbladder wall. Liver function tests were normal (SGOT=20 U/L, SGPT=11 U/L, Alk-P=373 U/L, Bilirubin total 1.5 and direct 0.4 mg/dl). Total serum protein, albumin, and LDH were normal (5.8, 3.8 gr/dl, and 419 U/L respectively), but ferritin increased (185 mg/dl). Wright, Widal agglutination tests, PPD test, peripheral blood smear for malaria, and immunoassays viral hepatitis panel were negative. Blood and urine cultures also were sterile .

On the 4th day of admission, transthoracic echocardiographic examination was performed. It showed normal findings. The fever continued, and physical examination was normal, so lab tests were repeated that showed increased CRP (96 mg/dl), WBC (17.200 /mm3 with 61.5% neutrophil), and platelet counts (669.000/ mm3). Therefore, meropenem was added to the antibiotic regimen. Because the fever had not stopped, repeated echocardiography was done on the 10th day of admission. The second echocardiography showed dilated right and left main coronary arteries (z score >2.5) without any vegetations, pericardial effusion, and atrioventricular valve regurgitation (Fig. 1). Left and right ventricular functions were normal. The patient's medical history, and the physical, laboratory, and echocardiography findings were compatible with incomplete KD. Electrocardiogram was normal. Serum troponin I and CPK were normal. Unfortunately on the same day, she developed left-sided peripheral FNP. Other neurologic examinations were normal. Intravenous immunoglobulin (IVIG) (2/g/kg/ infused over 12 hours) and aspirin (80 mg/kg, per day, divided/6h) was started and antibiotics were discontinued. The fever was resolved after about 36 hours and the general condition became better.

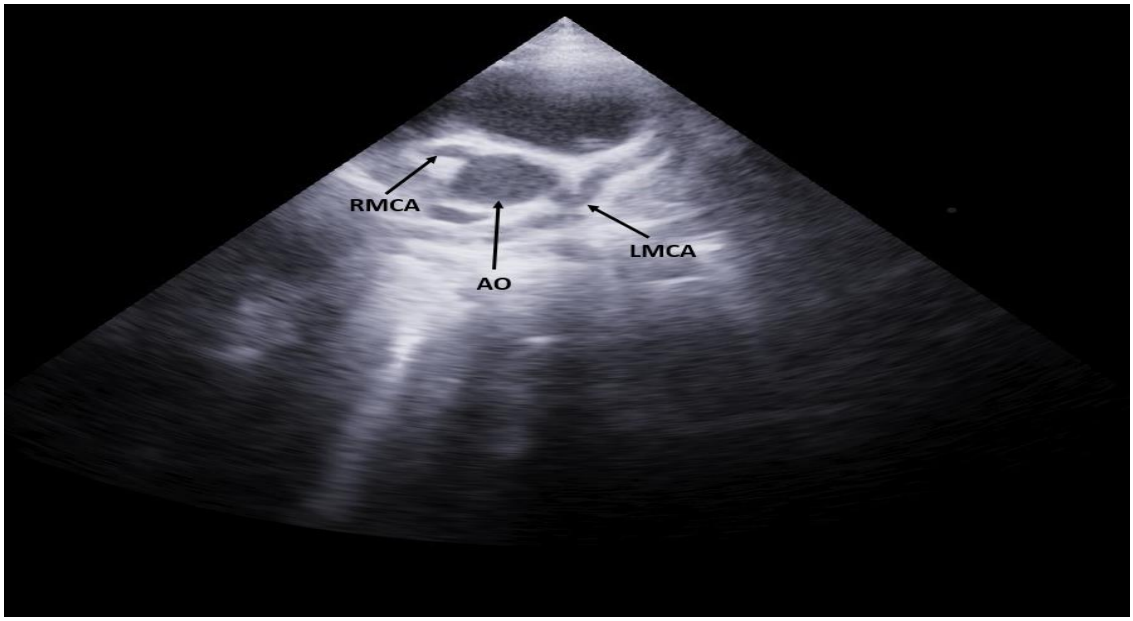

Figure 1. Two-dimensional Doppler echocardiogram. Parasternal short axis plane shows dilate of the left main coronary artery (LMCA), and right main coronary artery (RMCA). Ao; Aortic root.

On the 14th day of admission, WBC count and CRP decreased (5.7/ mm3 with 57% Lymphocyt, 84 mg/dl respectively), but platelet count increased to 714/mm3, and no changes in echocardiography findings. Patient was discharged on the 16th day of hospital admission with a low dose of ASA (5 mg/kg/day). A repeat echocardiogram 1 week after discharge showed mildly decreased both RCA and LCA dilations but after about 3 weeks post discharge showed normal size RCA and mild LCA dilation. Echocardiography 6 weeks after discharge showed normal sizes of RCA and LCA. FNP gradually resolves during 2 weeks and completely recovers after about 2 months post discharge. At follow-ups 4, 6, and 12 months after admission, she remained asymptomatic, normal physical findings including neurologic examination with normal echocardiogram.

Discussion

KD is a type of vasculitis that can affect various parts of the body, including the skin, nervous system, gastrointestinal tract, kidneys, lungs, eyes, and hematological system (5). There are two forms of KD: complete and incomplete (6). In the current study, a diagnosis of incomplete KD was made due to the absence of typical findings of KD and the presence of coronary artery dilation. Although neurological manifestations are not common in KD patients (prevalence of 1.1 to 3.7%), they can have a severe clinical course and result in a poor prognosis (7). The primary neurological symptoms that are most noteworthy encompass headache, aseptic meningoencephalitis, subdural effusion, cranial nerve palsy (CNP), cerebral vasculitis and stroke, encephalopathy and seizures, Neurodevelopmental disorders (NDDs), and Sensorineural hearing loss (8). The facial nerve (FN) represents the most commonly encountered CNP among pediatric patients diagnosed with KD with an incidence rate ranging from 0.9% to 1.3% (9). It is interesting to note that the existing literature only documents a total of 45 reported cases of KD accompanied by FNP. Consistent with these documented cases, the age at which KD manifested in these individuals varied from 3 to 25 months, with a predominant occurrence below the age of 20 months. Furthermore, there is a higher female prevalence at a ratio of 1.4:1, in contrast to the overall male predominance ratio of 1.5:1 for KD. Typically, the FNP tends to affect the left side (10). In the present investigation, the patient under scrutiny was a 4.6-month-old female infant displaying left-sided FNP .

FNP may serve as a potential indicator of a more serious pathological condition and heightened susceptibility to coronary artery involvement (4). In the reported case, although the first echocardiography was normal, repeated one showed coronary arteries dilatation which also suggests more watchful echocardiography.

IVIG and aspirin represent the principal therapeutic modalities for FNP stemming from an inflammatory vasculitic mechanism (1). The administration of IVIG and aspirin in the present investigation led to the resolution of fever within a timeframe of 36 hours. Subsequent to discharge, echocardiographic evaluations were conducted at intervals of approximately 1, 3, and 6 weeks, revealing a gradual amelioration of coronary dilatation. Notably, in the context of our clinical scenario, FNP emerged as the solitary neurological manifestation of KD, exhibiting a notable improvement after a span of 2 weeks, with full recovery achieved in approximately 8 weeks. The majority of individuals manifested spontaneous resolution of FN symptoms within a 3-month follow-up period.

Conclusion

FNP is an uncommon neurological symptom of KD. However, it typically resolves on its own and without any lasting effects in those who survive the illness. Nevertheless, it may indicate a higher likelihood of coronary artery involvement, and as a result, additional anti-inflammatory treatments and more closely monitored echocardiography may be necessary.

Abbreviations

KD: Kawasaki disease ; **FNP:** Facial nerve palsy, **IVIG:** Intravenous immunoglobulin; **CNP:** Cranial nerve palsy; **NDDs:** Neurodevelopmental disorders; **FN:** Facial nerve.

Ethics approval and consent to participate

All procedures performed in this study follow the ethical standards of the institutional and national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. written informed consent was obtained from parent for participant under 16 years old.

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