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Kawasaki disease manifesting as bilateral facial nerve palsy and meningitis: a case report and literature review

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Abstract

Background: Kawasaki disease (KD) is an acute multisystem vasculitic syndrome that predominantly affects infants and young children. Neurological complications are rare in patients with KD and the diagnosis is challenging. We report a case of KD that manifested as bilateral facial nerve palsy and meningitis.

Case report: A 6-month-old boy presented with a 10-day history of fever. Four days before admission, the patient developed a rash, conjunctival injection, perioral and perianal excoriation, and bilateral facial nerve palsy. Brain magnetic resonance imaging was normal. Echocardiography showed dilated coronary arteries and coronary artery aneurysms. A cerebrospinal fluid examination showed an elevated leukocyte count. A diagnosis of KD was made, and the patient was treated with gamma globulin and aspirin. The patient's fever subsided on the following day and the right-sided facial nerve palsy was relieved 1 month later. An 18-month follow-up showed that the left-sided facial nerve palsy persisted and the patient's condition remained stable.

Conclusion: KD manifesting as bilateral facial nerve palsy and meningitis is extremely rare. Clinicians should be aware of this condition, and early diagnosis and appropriate treatment should be emphasized.

Keywords

Kawasaki disease, facial nerve palsy, meningitis, leukocyte count, fever, coronary artery aneurysm

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Introduction

Kawasaki disease (KD) is an acute, multisystem, vasculitic syndrome that can involve any organ system in the body. KD predominantly affects infants and young children.[1] Neurological complications may occur in only 1% to 30% of all patients with KD. These neurological complications include irritability, aseptic meningitis, hemiplegia, cerebral infarction, ataxia, epileptic seizures, cranial nerve palsies, and focal encephalopathy.[2,3] Because cases of KD presenting with neurological abnormalities are unusual and the clinical symptoms are non-specific, the diagnosis is challenging. We report a case of KD that manifested as bilateral facial nerve palsy and meningitis, and we review the relevant literature.

Case report

A 6-month-old boy presented with a 10-day history of continuous fever. Antibiotic therapy (oral cefixime for 2 days and intravenous cefuroxime for 5 days) was administrated, but the fever persisted. Four days before admission, the patient developed left-sided facial nerve palsy. On the day before admission, the patient developed somnolence and bilateral facial nerve palsy. A physical examination showed impaired consciousness and enlargement of the bilateral submaxillary lymph nodes (the largest node was approximately 1.5×1.5 cm, with a hard nature and marked mobility), disappearance of the bilateral forehead wrinkles and nasolabial grooves, and an inability to close the eyelids completely. The bregma was slightly distended (2.0×2.0 cm). There was no neck rigidity or Brudzinski's sign, and bilateral Babinski's signs were positive. No rash or desquamation was noted. A laboratory examination showed leukocytosis (18.11×10^9 white blood cells [WBC]/L), a decreased hemoglobin level (93 g/L), reduced mean corpuscular volume (76.4 fL), reduced serum albumin level (29 g/L), increased C-reactive protein level (69.60 mg/L), increased erythrocyte sedimentation rate (120 mm/hour), increased urine erythrocytes (6.3 red blood cells/high-power field), and increased urine leukocytes (21 WBC/high-power field). A cerebrospinal fluid examination showed an elevated leukocyte count (36×10^6 WBC/L) with a monocyte percentage of 97%. Glucose, chlorine, and protein levels were normal, and bacterial culture was negative. Brain magnetic resonance imaging showed no abnormalities.

On admission, a diagnosis of bacterial infection was suspected, and the patient was treated with ceftriaxone, mannitol, and glucocorticoid to relieve the potential nervous edema. After 4 days of treatment, no symptomatic or laboratory improvement was observed. Echocardiography was performed and showed dilation of the bilateral coronary arteries and coronary artery aneurysms (Figure 1). Further examination of the medical history showed that the patient had experienced mild non-secretory conjunctival injection, and perioral and perianal excoriation for 1 day on the third day after onset. These symptoms spontaneously regressed.

A diagnosis of KD was made. The patient was treated with intravenous gamma globulin (2 g/kg, intravenous drip for 5 days) and oral aspirin (four doses of 30–50 mg/kg/day). The fever subsided on the following day. Three days later, the dose of aspirin was reduced to 3 to 5 mg/kg/day. Five days later, the laboratory parameters were all within the normal range. The right-sided facial nerve palsy was relieved 1 month after treatment. An 18-month follow-up showed that the left-sided facial nerve palsy persisted and the patient's condition remained stable. Echocardiography showed that the coronary arteries were still dilated, with a left coronary diameter of 4 mm and a right

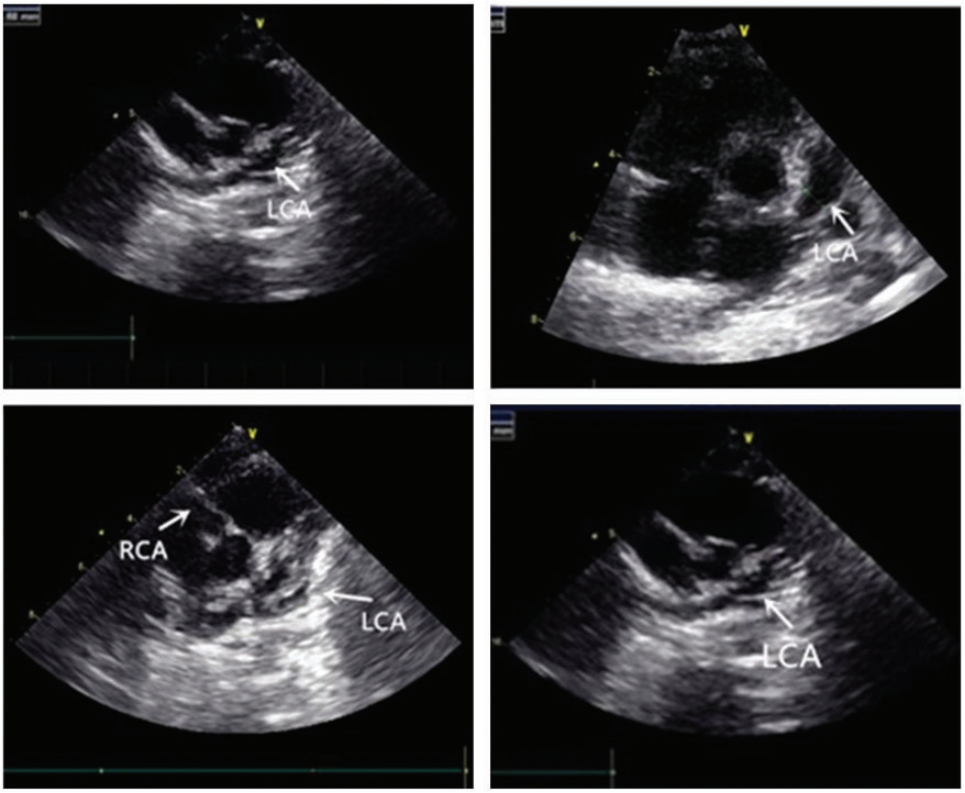

Figure 1. Echocardiography of the patient. Echocardiography shows aneurysmal dilatation of the bilateral coronary arteries. The maximal diameter of the left coronary artery (LCA) reached 5.5 mm and that of the right coronary artery (RCA) reached 6.2 mm

coronary diameter of 5.2 mm. Oral aspirin administration was continued.

Written informed consent was obtained from the patient's parents. This study was approved by the ethics committee of the First Hospital of Jilin University.

Discussion

In the current study, a diagnosis of KD was made according to the widely used criteria proposed by Tomisaku Kawasaki in 1967 as follows: presence of prolonged fever (>5 days), rash, conjunctival injection, perioral and perianal excoriation, and enlargement of submaxillary lymph nodes. Moreover, echocardiography and laboratory findings supported this diagnosis.[4,5] Our patient was only 6 months old, and fever developed for longer than 5 days with meningitis and bilateral facial palsy. Because the typical symptoms of KD were absent, and there was coronary artery dilation, a diagnosis of atypical and incomplete KD was made.[6]

KD mainly affects infants and young pediatric patients, in which any organ can be involved, such as the heart, skin, digestive tract, skeleton, respiratory tract and the nervous system. Nervous system involvement is observed in approximately 1% to 30% of cases of KD. Clinical manifestations of KD include irritability, somnolence, aseptic meningitis, ataxia, seizures,

focal encephalopathy, cranial nerve palsies, cerebral infarction, and transient hemiplegia.[7,8] To the best of our knowledge, this is the first case of KD that presented with bilateral facial nerve palsy and meningitis simultaneously.

Dengler and colleagues investigated the cerebrospinal fluid features of patients with acute KD.[9] They found that KD was characterized by an elevated leukocyte count and an elevated monocyte percentage, without remarkable changes in glucose or protein levels. In our case, the cerebrospinal fluid findings were consistent with these features. The patient responded poorly to antibiotics, and there were no bacteria in the cerebrospinal fluid. After treatment with gamma globulin, the cerebrospinal fluid results returned to normal, and thus infection or neoplasms were excluded. We determined that there was concurrent aseptic meningitis.

Facial nerve palsy secondary to KD is extremely rare. We reviewed the relevant literature and found only 45 reported cases of KD with facial nerve paralysis. A summary of the clinical characteristics of these cases is as follows. 1) The median age of these patients was 9 months and 62.5% of the patients were aged younger than 1 year. 2) The median clinical course of facial nerve palsy from onset to diagnosis was 16 days and the median clinical course from diagnosis to complete remission was 18 days (the longest duration was approximately 3 months). 3) There was a slight female predominance, with a female-to-male ratio of 1.2:1. 4) In the majority of the previous cases, facial nerve palsy was unilateral, and only one case showed bilateral facial nerve palsy.[10] 5) Cerebrospinal fluid findings were all normal in the previous studies. 6) Facial nerve palsy had no sequela. 7) The incidence of coronary artery complications in this cohort (approximately 62.5%) was significantly higher than that in general patients with KD.

We speculate that facial nerve palsy may be an indicator for evaluating the severity of KD. The mechanism of facial nerve palsy in KD remains unclear, but it may be associated with arterial vasculitis and immune dysfunction. Ischemic vasculitis of arteries supplying the facial nerve and inflammatory mechanisms may contribute to facial nerve palsy in KD. Additionally, we noted that facial nerve palsy was greatly improved in our patient after administration of intravenous gamma globulin. Therefore, immune function may be involved in the pathogenesis of facial nerve palsy in KD.[11,12] In the current case, left-sided facial nerve palsy persisted during the 18-month follow-up period. We hypothesize that this poor prognosis was due to inflammatory overburden. A much longer anti-inflammatory treatment and close follow-up are still required. Our findings of KD, which presented with bilateral facial palsy and meningitis, have not been previously reported. Because the onset age is early, and clinical symptoms were atypical, the diagnosis of KD was challenging in our case. Clinicians should consider KD as a differential diagnosis when a young child presents with long-term fever. The diagnostic value of coronary artery ultrasound should be highlighted, which facilitates early diagnosis and treatment.

Conclusion

KD manifesting as bilateral facial nerve palsy and meningitis is extremely rare. Clinicians should be aware of this condition because it is usually underdiagnosed owing to the non-specific clinical manifestations. Early diagnosis and appropriate treatment of KD are important.

Declaration of conflicting interest

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