CASE REPORT

Ptosis as a complication of Kawasaki disease

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SUMMARY

Kawasaki disease is an acute febrile exanthematous disease that affects children younger than 5 years of age. It is regarded as the most common cause of childhood acquired heart disease, but ocular and neurological problems are among the other important clinical findings. We present a 3-year-old boy who developed bilateral ptosis on day 21, 5 days after intravenous immunoglobulin. The ptosis was due to bilateral paralysis of the levator palpebrae superioris muscles and resolved spontaneously on day 25. There were no cardiac sequelae.

BACKGROUND

Kawasaki disease (KD) is a small-to-medium vessel vasculitis that affects children in the age group below 5 years. It results in a variety of clinical signs and symptoms including the skin, mucous membranes, eyes and neurological system.^{1,2}

The exact underlying cause of KD remains unknown and there is no specific pathognomonic test for confirmation of the diagnosis. However, the American Heart Association established criteria for diagnosing typical and atypical KD.³

Typical KD must have fever for at least 5 days together with four clinical features from the following list: skin rash, conjunctivitis, acral skin changes, cervical lymphadenopathy and oral mucosal changes. When only two or three of these features are present, then the diagnosis is atypical KD.

Coronary artery thrombosis and aneurysms are the most frequent and important complications occurring in 25% of untreated cases. Early diagnosis and prompt therapy can reduce the probability of such serious complications.⁴

Typical ocular manifestations include bilateral bulbar conjunctival non-purulent injection, uveitis, iridocyclitis, papilloedema and keratitis.⁵ We report a childhood case of KD complicated by bilateral ptosis.

CASE PRESENTATION

A 3-year-old boy presented with 2 days' history of headache and abdominal pain followed by fever at the next day along with prominent unilateral cervical lymph node enlargement (figure 1). At the fourth day of illness, a maculopapular rash started on the lower limbs. The fever persisted.

At the end of the first week, bilateral upper eyelid oedema started to appear. Laboratory investigations showed a leucocytosis $18.0 \times 10^9/L$ (39.4% lymphocytes; 42.6% neutrophils), haemoglobin 11.1 g/dL,

platelet $350.0 (\times 10^9/L)$, erythrocyte sedimentation rate 80 mm/1st.hr and an unremarkable general urine test. A non-specific viral exanthem was diagnosed at this stage and paracetamol suppositories were recommended for the fever.

By day 8, the fever and eyelid oedema persisted, the rash had spread to the trunk and was visible faintly on the extremities (figure 2).

During the second week of the disease, a resolution of the eyelid oedema as well as the rash on the lower limbs were noted, while the skin rash became more prominent on the trunk and face. Along the second week of illness, no decrease of body temperature recorded in spite of the oral antibiotic intake (cefixime 8 mg/kg/day for 5 days).

Repeat investigations showed a leucocytosis $20.0L$ (30.5% lymphocytes; 30.6% neutrophils, 28.1% eosinophils), haemoglobin 10.5 g/dL, platelet count $345.0 (\times 10^9/L)$ and normal echocardiography. Atypical KD was diagnosed based on the prolonged fever, exanthem, unilateral cervical lymphadenopathy, leucocytosis and high ESR.³

Figure 1 Large prominent unilateral cervical lymphadenopathy developed early in the course of Kawasaki disease.

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Figure 2 Generalised non-itchy maculopapular rash is a major criterion of Kawasaki disease.

At the beginning of the third week, the lips became fissured and crusted and a bilateral non-purulent bulbar conjunctivitis developed which now fulfilled the criteria for typical KD.

On day 16, aspirin (20 mg/kg orally four times per day) was started and next day an infusion of 30 mg/kg of intravenous immunoglobulin was given over 12 hours.

According to recommendations of the American Heart Association, intravenous immunoglobulin can be prescribed within 10 days of disease presentation or later if the patient has persistent fever, aneurysms or inflammation.³

This resulted in complete resolution of the cutaneous rash and fever over 12 hours. After 48 hours, the dose of aspirin reduced to 81 mg single dose daily and continue for 8 weeks.

The child was then well, completely free from any signs and symptoms. However, a bilateral ptosis of the eyelids occurred on day 21. Ophthalmological and neurological evaluation revealed bilateral paralysis of the levator palpebrae superioris muscle (figure 3).

The pupils showed normal reactions to light and there was no diplopia. MRI was normal, excluding a central cause for the ptosis. Spontaneous resolution of the ptosis occurred on day 25.

OUTCOME AND FOLLOW-UP

At the 6-month follow-up, the child was completely healthy without any residual findings of KD. Ophthalmological examination and echocardiography revealed no abnormalities.

DISCUSSION

Ptosis developing in KD is very rare. To the best of our knowledge, we present only the fourth case report.⁶⁻⁸

In the previous reports, the ptosis developed early in the course of the disease occurring on day 7 and associated with fever and cardiac involvement. Our case differed in that the ptosis appeared late occurring first at day 21 when the child was afebrile and there has been no abnormality found on cardiac follow-up.

Our case was also unique in that although the rash and fever resolved within 24 hours of the intravenous immunoglobulin treatment, the ptosis appeared 5 days after the intravenous immunoglobulin therapy. This contrasts with the 2013 case where the ptosis and fever resolved with intravenous immunoglobulin.⁷

Our case is similar to Falcini *et al* where both have late occurrence of ptosis, no associated cardiac complications and spontaneous resolution of the ptosis.⁸

Neurological complications are not uncommon in KD. There have been more than 40 cases of facial nerve palsy reported in the medical literature.⁹ Transient unilateral oculomotor palsy has been reported with accompanying double vision and unilateral ptosis.¹⁰ The ptosis in our case was not the result of oculomotor nerve injury as the eyelid drop was bilateral and symmetrical, the pupil examination was normal and central cranial pathology was excluded by MRI. It is likely the ptosis was initiated by the inflammatory process affecting the levator palpebrae superioris muscles,

Figure 3 Bilateral eyelid ptosis was a late sign complicating Kawasaki disease.

Learning points

- ▶ Kawasaki disease should be considered in the evaluation of ptosis associated with a febrile exanthem in young children.
- ▶ Ptosis can occur even after apparent resolution of the disease process or after intravenous immunoglobulin treatment.
- ▶ Ptosis of Kawasaki disease can resolve spontaneously.

and the bilateral eyelid oedema may have been due to an early inflammatory myopathy of the eyelid muscles.

Although in the previous case reports KD-associated ptosis responded well to immunoglobulin treatment, we observed the ptosis can spontaneously resolve.

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