Kawasaki Disease with Bilateral Ptosis-A Case Report

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Abstract
Kawasaki disease (KD) is an acute self-limiting vasculitis and probably the most common vasculitis in pediatric age group. It has become the commonest cause of acquired heart disease in west as well as in the east replacing the rheumatic fever. The diagnosis of Kawasaki disease is entirely clinical with none of the clinical features are pathognomic. It may present with many typical as well as atypical features. Here we present a case of Kawasaki disease where a 3 years old boy presented with atypical KD with bilateral ptosis.

Key words: Kawasaki disease, ptosis, levator palpebral superioris

Introduction
Kawasaki Disease (KD) is a vasculitis of the medium- and small-sized vessels. Though few clinical features are typical, none are pathognomonic. Recognition of a clinical pattern in association with laboratory evidence is needed to establish the diagnosis. It can affect almost any system. Involvement of the Central Nervous System is not uncommon, predominantly manifesting as extreme irritability, or even aseptic meningitis. Seizures, cerebrovascular accidents, and cranial nerve paralysis have also been reported. But involvement of bilateral Levator Palpebrae Superioris muscle to cause bilateral ptosis is a rare phenomenon. Here we report a case of atypical KD who presented with bilateral ptosis and cardiac involvement.
Case Report
A 3-year-old boy was admitted with fever for 4 days, with one episode of generalized seizure on day 3 of fever. There was a history of maculopapular rash on day 2 of fever, which subsided before admission.

On admission, the boy was febrile, with erythematous hue on the lips, upper eyelids and palms. There was no conjunctival congestion, rash, or lymphadenopathy.

Initial investigations revealed a total leucocyte count of 22,380/cmm, N70, L28, M2, E0, B0, Hb – 9.1 gm%, platelet – 356,000/cmm, ESR – 64 in 1st hour, CRP – 186 mg/L (Normal 6<mg/l), normal liver and renal function, and normal urine microscopy.

CSF study revealed 46 cells, which were all lymphocytes with normal protein and sugar. MRI of the brain was normal.

From day 3 of admission (day 7 of fever) he started having bilateral drooping of the upper eyelids with continuation of high spikes of fever (figure 1). There was no other neurological involvement. Pupils were normal in both the eyes and pupillary reaction was normal. As the pupils were unaffected, ptosis was due to the paralysis of Levator Palpebrae Superioris muscles.

Repeat investigations revealed neutrophilic leucocytosis, thrombocytosis, high CRP and ESR, and raised transaminases with hypoalbuminemia. Serum sodium dropped down to 128 mEq/l. Features of cardiac involvement in the form of gallop rhythm with a short systolic murmur became prominent. Echocardiography showed increased perivascular brightness of coronaries with ectasia, mild pericardial effusion, and moderate MR.

Since KD was a strong possibility, IV immunoglobulin was started at a dose of 2 gram/kg with 30 mg/kg/day of aspirin. Following initiation of IV Ig, there was defervescence and gradual improvement of ptosis over the next 24 hours (figure 2). Repeat echocardiography after 6 weeks was normal. The child has been on regular follow-up in our Pediatric Rheumatology clinic for the last 8 months and remains asymptomatic.

Discussion
Kawasaki disease was first described by Tomisaku Kawasaki in Japan in 1967 [1]. It is a form of acute multisystem necrotizing vasculitis involving small- and medium-sized arteries with a predilection for widespread involvement. This is now said to be the commonest cause of acquired heart disease in children in developed countries [2,3], surpassing the incidence of rheumatic heart disease. In the developing countries it is now being recognized more frequently. The disease primarily affects children below 4 years of age [4,5]. The most common organ affected is the heart, followed by the gastrointestinal tract and central nervous system.

Neurological complications of KD are well recognized in the form of hemiplegia, seizures, myositis [6], cranial nerve involvement, peripheral neuropathy, cerebral infarctions, and aseptic meningitis [7]. In one large series [8], neurological complications arose in 1.1% of cases. There have been 18 previously reported cases of facial nerve palsy in Kawasaki disease.
The most common CNS manifestation is aseptic meningitis, which probably accounts for the extreme irritability. A review of these cases [9] noted that six of the 10 children, i.e., 60%, in whom cerebrospinal fluid was sampled had pleocytosis. Ghosh et al. reported a case of KD with palatal palsy [10]. Thapa et al. also reported a case of KD with oculomotor palsy [11].

Ophthalmologic manifestation in KD usually is bilateral non-purulent bulbar conjunctivitis. Bilateral ptosis is an extremely rare manifestation. Until today, only Zhao SH et al. had reported a single case of KD with ptosis due to paralysis of palpebrae superioris muscle [12].

Timely (particularly within the first 10 days) administration of immunoglobulin can reduce the occurrence of coronary aneurysms, but whether this can prevent neurological complications is not known, though it has been seen that the irritability decreases after immunoglobulin. In our index case, the ptosis started improving within 12 hours of completion of immunoglobulin infusion, and completely disappeared after 24 hours.

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References

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