

subgroup with associated paroxysmal choreoathetosis has also been described [6]. As per ILAE classification [7], both BFIS and BIS are considered similar, except for the family history.

Generally, no further seizures are observed in cases treated pharmacologically. In untreated cases, there can be isolated or brief clusters within one year of age. Treatment with antiepileptic medication is not mandatory [8]. We started antiepileptic drugs in first two patients, but with increasing confidence about this diagnosis, we discharged the next two infants without drugs, and they did not have any seizure recurrence over the next 5 to 9 months.

The most characteristic feature of the syndrome is the occurrence of a cluster of few brief seizures, lasting for 1-3 days, with the child being well inter-ictally [9]. Recognition of this syndrome helps in avoiding long term anti-epileptic therapy.

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during patient management and manuscript preparation; BT: reported EEG findings. All authors approved the final manuscript.

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## Recurrent Kawasaki Disease

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**Background:** Recurrent Kawasaki disease is rare. **Case characteristics:** An eight-month old infant had classic Kawasaki disease with transient coronary artery dilatation. **Observations:** Recurrence of incomplete Kawasaki disease after two years of initial diagnosis. **Outcome:** The index episode of Kawasaki disease was resistant to single infusion of immunoglobulin, while repeat episode responded within 24 hours of institution of therapy. **Message:** Early recognition of recurrent Kawasaki disease requires a high index of suspicion.

**Keywords:** *Incomplete Kawasaki disease, Outcome, Recurrence, Resistant.*

**K**awasaki disease (KD) is an acute, self-limiting, medium-size vessel vasculitis of unknown etiology that predominantly involves the skin, mucous membranes, lymph nodes and coronary arteries. Standard therapy of KD is with a single intravenous infusion of immunoglobulins (IVIG) and high-dose aspirin until the acute phase reactants normalize. IVIG-resistant KD, which occurs in approximately 15% of children, can be defined as the persistence of fever beyond 36 hours of the initial IVIG

infusion, and mandates a 2nd or even 3rd dose of IVIG [1-3]. Recurrent KD is mostly reported in Japan and the US, occurring in 3-4% and 0.8% of cases, respectively [4], but is only rarely reported from India [5].

#### CASE REPORT

An 8-month-old boy was referred to us with cough and cold for 15 days, fever for 5 days, and loose motions for 2 days. On examination, he was irritable and febrile. He had tachypnea and tachycardia with normal blood pressure.

His eyes and oral mucosa were injected, and he had strawberry tongue. His hands and feet were edematous. Systemic examination was unremarkable. The BCG scar mark was inflamed.

Hematological investigations revealed hemoglobin (Hb) 7.5 g/dL, total leucocyte count (TLC)  $36.8 \times 10^9/L$ , P:76%, L:22%, platelet count  $400 \times 10^9/L$ , C-reactive protein (CRP) 12 mg/dL, erythrocyte sedimentation rate (ESR) 38 mm/h, aspartate aminotransferase (AST) 15.6 U/L, alanine aminotransferase (ALT) 10.0 U/L, serum sodium 137 meq/L, potassium 3.6 meq/L, serum albumin 2.3 g/dL, blood urea 25.2 mg/dL, and serum creatinine 0.72 mg/dL. Routine microscopy of urine and cerebrospinal fluid was normal. Antistreptolysin O (ASO) titer and Mantoux test were negative. Malarial parasites were not seen in smear. Chest X-ray showed cardiomegaly. Abdominal ultrasound was normal. He received injections of ceftriaxone, amikacin, metronidazole and gatifloxacin for four days before being referred to us. In our hospital, injections of meropenem, vancomycin and artesunate were administered empirically. In spite of 10 days therapy, fever was unresponsive, and the child was provisionally diagnosed as KD. Two-dimensional echocardiography (2D-Echo) revealed left coronary artery dilatation (6.9 mm), no fistula, and mild pericardial effusion with ejection fraction of 60%. Subsequently, IVIG (2 g/Kg stat dose) and aspirin (85 mg/kg/day in divided doses) were administered. Fever was persistent even after 36 hours of IVIG, in view of resistant KD; a second dose of IVIG was given [2]. Consequently fever subsided within 24 hours of the second dose of IVIG. Aspirin was prescribed at high doses till acute phase reactants normalized (7 weeks), following which the dose was tapered to 5 mg/kg/day. Echocardiography at 4 months revealed reduction in left coronary artery dilation (3.4 mm) that completely resolved at 9 months; aspirin was stopped subsequently.

The child again presented at three years of age (after 2 years of first episode) with fever for four days, pain in legs and swelling of hands and feet for two days. He had no history of cold, cough, allergy, insect bite or drug intake. On examination, the child was febrile and irritable with tachycardia and normal blood pressure. His oral mucosa, tongue and pharynx were diffusely injected; ulcers, enanthem and exudates were not noted. Right sided cervical lymphadenopathy (>1.5 cm) along with dry, crusted and cracked lips were present. Edema was noted over hands and feet. Skin examination did not revealed eruptions or rashes. Rest of the systemic examination was normal.

Laboratory tests revealed Hb 8.7 g/dL, TLC  $16.8 \times 10^9/L$  with 73% neutrophils, platelet count  $521 \times 10^9/L$ ,

serum albumin 3.2 g/dL, AST 154.0 U/L, ALT 159.7 U/L, ESR 48.0 mm/h, CRP 4.8 mg/dL, Serum sodium 128 meq/L and potassium 3.9 meq/L; ASO titer was negative. Peripheral smear for malarial parasite and Mantoux test were negative. Blood culture was sterile. Renal function tests and routine microscopic examination of urine was normal. Chest X-ray and abdominal ultrasound was normal. The child was administered intravenous ceftriaxone, oral antihistaminics and antipyretics. Despite 72 hours of therapy, fever persisted and there was worsening of oral mucosal congestion and swelling of extremities. Widal and malaria antigen tests were negative. Subsequently, child developed periungual desquamation of fingers, and the child was diagnosed as recurrent KD with incomplete presentation (>5d fever along with 3 clinical criteria: changes of mucosae of oropharynx, cervical lymphadenopathy and changes of peripheral extremities). Concomitant echocardiogram revealed normal coronary arteries. On the 7th day of fever, the child was administered with IVIG (2 g/Kg/day) and aspirin (80 mg/Kg/day) in three divided doses. Defervescence was achieved within 24 hours and remaining features improved over next 2 days. Follow-up after 15 days revealed desquamation of skin of both soles (**Fig. 1**). Skin peeling was in sheets and vastly intense to involve the entire sole, which further substantiated our diagnosis. Coronary arterial abnormalities (CAA) were not noted in 2D-echo at 6 weeks, 6 months and 12 months



**FIG. 1** Desquamation of skin over the soles during second episode of Kawasaki disease. (Color image at website)

follow-up. The child is being followed up regularly for the last four years without any symptoms.

## DISCUSSION

KD presents with classic or incomplete manifestations and is diagnosed after excluding similar disease conditions with the help of clinical criteria proposed by the American Heart Association [3]. The differential diagnosis of KD includes viral infections (measles, adenovirus, rubella, erythema infectiosum, infectious mononucleosis and herpangina) which shares acute oropharyngitis, fever and cervical adenopathy but with less evidence of systemic inflammation and lack the extremity changes as seen in KD. Systemic onset juvenile idiopathic arthritis mimics KD but lack of arthritis in the child over long term follow up excluded it. Absence of skin rashes negated scarlet fever, rickettsial infections and polyarteritis nodosa. The blood pressure was normal, ruling out streptococcal toxic-shock syndrome. Thus, we diagnosed the case as recurrent KD with incomplete presentation, which was supported by laboratory reports. Sheetlike skin peeling is pathognomonic of KD and additionally supported our diagnosis [2]. Both the episodes responded well to IVIG. However, the initial episode was resistant to the single infusion of IVIG and responded only after repeat dose [1]. If the symptoms of KD persist or recur after IVIG therapy, macrophage activation syndrome as a complication of KD should be considered. However it was excluded, as hepatosplenomegaly and pancytopenia were not noted in the child during entire course of illness.

The risk factors predictive of recurrent KD in a child are: younger age ( $\leq 2$  years) at the onset, male sex, treatment with IVIG [6,7], longer durations of fever, lower hemoglobin levels [8] and presence of CAA at the first episode [4,9]. All these risk factors for recurrence at the first episode were present in our case as well. High levels of transaminitis at first episode also poses risk for recurrent KD but were normal in our patient.

Hirata, *et al.* [8] reported recurrence of KD within a year while, the present case had recurrence after two years of first episode. Recurrence of KD and Incomplete KD are generally associated with CAA [2,3]; however, this was not seen in our case. Kato, *et al.* [2] reported CAA occurrence in 20% children of untreated KD in Japan. Daniels, *et al.* [10] reported that undiagnosed or

partially treated KD in childhood contributes CAA in 5% of adults in the USA.

Kawasaki disease should be included in the differential diagnosis of fever unresponsive to appropriate therapy, as all the criteria may not appear simultaneously but emerge serially, more so in a child with previous history of Kawasaki disease.

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