Familial Occurrence of Kawasaki Disease in an Indian Family

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Conflict of Interest: None declared

This article has been peer reviewed.

Article Submitted on: 1st September 2011

Article Accepted on: 18th September 2011

Funding Sources: None declared

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Cite this article: Bannajji MH, Puttaiah K, Patil S. Familial occurrence of Kawasaki disease in an Indian family. J Pak Med Stud. 2012; 2(4):151-153

ABSTRACT

The etiology of Kawasaki disease (KD) continues to be elusive to the researchers. Although epidemiological studies support infectious origin, familial occurrences across the world suggest possible genetic predisposition. We are reporting two cases of KD diagnosed among cousins. The diagnosis in both these cases was based on clinical criteria. Laboratory data and response to therapeutic measures was also noted. This study reports the occurrence of familial KD in Indian population, which has not been reported in Indian literature before.

Keywords: Kawasaki disease; Familial; Diagnosis

INTRODUCTION

Kawasaki disease (KD), formerly known as the mucocutaneous lymph node syndrome, is an acute febrile illness of childhood seen worldwide. It is a vasculitis with a predilection for the coronary arteries, with approximately 20%-25% of the untreated patients experiencing coronary artery abnormalities, including aneurysms. Attempts to find an etiological agent for Kawasaki disease, one of the leading causes of acquired heart disease in children, have not found a definitive answer, although epidemiological studies support infectious origin. Continued research to solve the mystery of KD has found an interesting aspect to the occurrence of the disease in the form of its familial occurrence. Surveys from Japan and North America have reported KD in family members, siblings and offspring, suggesting a possible genetic predisposition for the occurrence of the disease and for the involvement of coronary vessels [1] [2]. There have been reports of recurrence of KD with coronary artery involvement in siblings [3]. There is not much information on the familial occurrence of KD in Indian population. Herewith presenting an Indian child with KD whose cousin had suffered from the same disease earlier on, suggesting familial occurrence of KD in Indian population.

CASE REPORTS

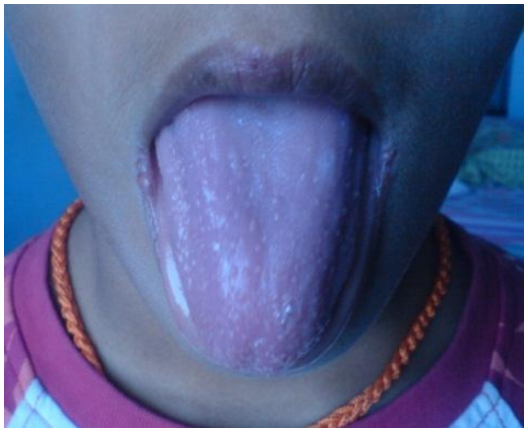
Case 1: The Patient

An 8-year-old male child presented with history of high grade intermittent fever of 5 days, with redness of tongue and peeling of the skin of hands and feet for 3 days. He was the only child born to non-consanguineous parents. He has no significant medical or past history. On examination (Figure 2), he was febrile, with bilateral bulbar conjunctival congestion, strawberry tongue, angular cheilitis, periungual desquamation (Figure 3), bilateral cervical lymphadenopathy and mild hepatomegaly. A diagnosis of KD was made based on his clinical presentation. Following admission, his initial blood investigations showed leukocytosis with normal platelet count, and raised erythrocyte sedimentation rate (ESR) and raised C-reactive protein (CRP). Echocardiogram done at admission revealed dilated left coronary artery. Child was treated with intravenous immunoglobulin (IVIg) and aspirin. Fever subsided subsequently by fourth day of treatment. His condition improved significantly and was later discharged from the hospital. On follow-up after 3 weeks, repeat echocardiogram revealed normal left coronary artery, showing improvement from the previous findings.

Case 2: The Cousin

The maternal cousin of the above mentioned child was admitted with similar complaints 5 months previously at our hospital. The second case was also a male child, aged 4 years, who presented with a history of high grade fever for 7 days. He had no significant medical or past history. On examination, the child had bulbar conjunctival congestion, desquamation of the oral mucosa and peeling of the skin on hands and feet. He also had significant cervical lymphadenopathy. Based on these signs and symptoms, a diagnosis of KD was made, and he was admitted to the hospital for further management. At admission, the blood investigations revealed leukocytosis with normal platelet count, and raised ESR and CRP. Echocardiogram done at admission was normal. The child was also treated with IVIg. He responded well to the treatment, showing improvement over a period of five days.

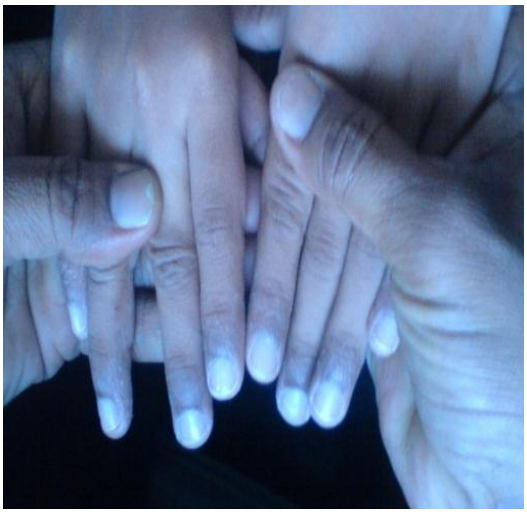
Figure 1: Strawberry tongue and angular cheilitis in the patient



DISCUSSION

Familial occurrence of KD has been well documented by the Japanese researchers. Studies from America have documented that the disease is more common among Asians and Asian-American populations [4]. In Japan, the siblings of an index case were found to have 10-fold increased relative risk of KD. The incidence of KD is 2-fold higher in Japanese parents of children with the disease. The incidence of recurrent KD in siblings is 5 to 6 times higher in multi-generational KD families as compared to those with only one affected child.

Figure 2: Periungual desquamation in the patient

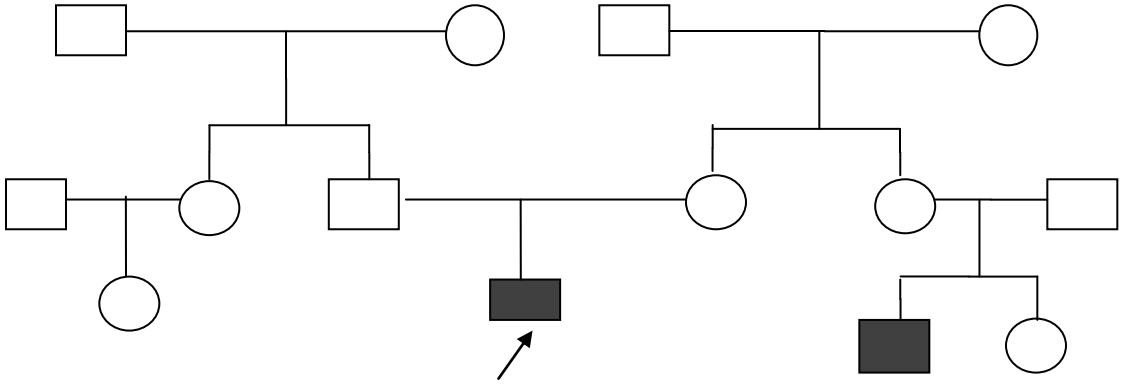


Familial occurrence points towards possible genetic predisposition and also helps in understanding or predicting the severity and course of the disease, including the risk of coronary artery involvement. It has been reported in previous studies that the disease severity and risk of coronary involvement is higher in those with familial occurrences [5]. This case report.

Table I: Clinical and lab parameters cousins with Kawasaki disease

Characterist-ics	Patient	His cousin
Age (years) Sex	8yrs Male	4yrs Male
Fever (days) Skin rashes Extremity changes Lips and oral cavity changes Eye changes Cervical Lymphadenop-athy	5 No Yes Yes Yes Yes	7 No Yes Yes Yes Yes
Hemoglobin	12.4 g/dl	11.2 g/dl
Total leukocyte count [neutrophils]	21600 cells/cumm[75]	14900 cells/cumm [82]
ESR CRP Platelets SGOT SGPT Albumin	40 mm/hr 15 mg/dl 3.56 x 10^5 cells/cumm 20 15 3.4 g/dl	55 mm/hr 18 mg/dl 5.5 x 10^5 cells/cumm 56 53 3.1 g/dl
Echocardiogram	Dilated left coronary artery	Normal

Figure 3: Family tree of the Indian Family. Arrow on black box denotes patient. Second black box denotes cousin



highlights the familial occurrence of KD in Indian population

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