Kawasaki Disease: A Case Report Every Otolaryngologist Should Know

Saurav Sarkar · Sudip Dawn ·
Amitabha Roychoudhury · B. K. Roychaudhuri

Received: 29 August 2008 / Accepted: 28 March 2009 / Published online: 9 July 2011
© Association of Otolaryngologists of India 2011

Abstract Kawasaki disease is the acute febrile vasculitis of childhood, predominantly affecting medium sized arteries with predilection for coronary arteries. The diagnosis is mainly clinical, though investigations are helpful in early detection of complications. We report a case of Kawasaki disease in a 5 year old child who presented to our hospital with conjunctival congestion, lymphadenopathy and fever. When on treatment with antibiotics did not help resolve symptoms, particularly conjunctival congestion Kawasaki disease was suspected. She was treated with a single dose of intravenous immunoglobulin G, over a period of 12 h. She subsequently developed a systolic murmur with grade I mitral regurgitation not a known complication of Kawasaki disease. She responded to the treatment remarkably and promptly thus evading any fatal complication.

Keywords Fever · Cervical lymphadenopathy · Kawasaki disease · Immunoglobulin · Mitral regurgitation

Introduction

Kawasaki disease (KD), formerly known as the mucocutaneous lymph node syndrome is the acute febrile vasculitis of childhood first described by Dr. Tomisaku Kawasaki, Japan in 1967. This disease occurs worldwide with Asians at highest risk (Laupland et al. [1]).

Kawasaki disease is self-limited acute vasculitis of childhood of unknown etiology. Treadwell et al. [2] have suggested the possibility of the association between KD and the use of humidifier in the room of a child with an antecedent respiratory illness. Nomura et al. [3] has pointed out the possibility of relationship between streptococcal pyrogenic exotoxin A and KD. Shingadia et al. [4] suggested a ubiquitous virus of the gamma virus family to be the likely etiological agent for genetically susceptible individuals. Many hypotheses regarding etiology of KD exist but the debate is persistent as to whether the inflammatory response of KD is from a super antigen or a conventional antigen (Guzman et al. [5]).

Kawasaki disease is most commonly observed in middle and upper middle classes. It is more common in the Japanese population. This syndrome has now surpassed rheumatic fever as leading cause of acquired heart disease in USA among children younger than 5 years. The annual incidence in USA is 17.1/100,000 children and in Japan 111.7/100,000 children. Thus it may be considered as a rare disease but Taubert et al. [6] found out in his study that Kawasaki disease is not uncommon but since its atypicality it often goes unnoticed and unreported. Male to female ratio being 1.5:1, the peak prevalence is in children aged 18–24 months. The disease occurs predominantly in young children, 80% of the patients are below 5 years of age. Pannaraj et al. [7] recently noted that KD might occur in children younger than 6 months or older than 5 years.

Epidemiological and clinical features strongly support an infectious origin. Kawasaki disease causes severe vasculitis of all blood vessels but predominantly affects medium sized arteries with predilection for coronary arteries. On pathological examination there is intense inflammation with infiltration of vessel wall initially with neutrophils then macrophages and lymphocytes (mostly

S. Sarkar (✉)
Fulbright Scholar, Stanford University, California, USA
e-mail: kushalkushal79@rediffmail.com

S. Dawn · A. Roychoudhury · B. K. Roychaudhuri
Vivekananda Institute of Medical Sciences, Kolkata, India

CD8 T cells) and plasma cells. IgA plasma cells predominate the infiltrate. Vessel loses structural integrity and weakens causing dilatation or aneurysm. Thrombi may form in the lumen and obstruct blood flow. In healing phase, the lesion becomes fibrotic and may result in stenosis (Brown et al. [8]).

There is no specific diagnostic test for this disease (Burns et al. [9]). The diagnosis of this disease is based on the diagnostic guidelines for Kawasaki disease as per American Heart Association Council on coronary vascular disease in young, committee on rheumatic fever, endocarditis and Kawasaki disease.

According to Chantepie et al. [10] the primary complication is rupture of coronary artery aneurysm. Other complications being dehydration resulting from fever, anorexia and joint inflammation in acute phase limiting mobility.

Timely institution of treatment with IV IgG and high dose of aspirin as early as possible after diagnosis causes rapid defervescence of the disease (Dajani et al. [11]).

Case Report

A 5 years old female child presented to the emergency department Ramakrishna Mission Seva Pratishthan, Vivekananda Institute of Medical Sciences, Kolkata, with the history of remittent fever for the past 4 days along with congestion of eyes and cervical lymphadenopathy for the same duration (Fig. 1). Her parents gave history of appearance of rash over upper chest and abdomen on the second day of her illness, which resolved spontaneously. She had visited a general physician as per whose advice she had taken antibiotics without any remission of her symptoms. She was then referred to our hospital.

Clinical examination revealed multiple cervical lymphadenopathy on the left side involving levels I, II and V. There were four mobile and non-tender lymph nodes each measuring about 2 cm in diameter. Her conjunctiva, oropharynx and nasal mucosa, were found congested with high fever. Other systemic examinations were within normal limits. A screening echocardiography was done and was found to be normal. She was admitted and intravenous coamoxyclov was started presuming an infective etiology. Gradually the lymph node swelling started decreasing in size along with remission of fever.

Ophthalmologists were consulted for the persistent conjunctival injection. Antibiotic and antiallergic eye drops were applied but conjunctival congestion persisted even after 2 days. A provisional diagnosis of KD was considered in consultation with the paediatrician. She was given intravenous immunoglobulin G in a single dose of 2 gm/kg over a period of 12 h under close supervision. She

Fig. 1 The anxious child with conjunctival congestion, lymphadenopathy and congested fissured lips

responded to this remarkably with conjunctival congestion disappearing within 24 h. She had a periungual desquamation in fingers of both extremities from about 4th day of admission. She also developed a systolic murmur. The child was not irritable throughout the period of illness.

Echocardiography showed mild volume overload with left atrial and ventricular hypertrophy with grade I mitral regurgitation (MR) Fig. 2. Urine examination was found to be within normal limits and no growth was found on culture. On admission her blood profile showed—polymorphonuclear (PMN) leucocytosis, normal Hb%, normal platelet count, markedly increased ESR and positive C reactive protein. She was discharged with smiling face on the 14th day (Fig. 3); after which she was followed up in our clinic at weekly interval for 6 weeks without any signs of late complications.

Discussion

The diagnosis of KD is based on diagnostic criteria as per American Heart Association Council on coronary vascular disease in young, Committee on Rheumatic fever, endocarditis and Kawasaki disease diagnostic guidelines [12] for Kawasaki disease—fever lasting for at least 5 days along with presence of at least 4 of the following 5 signs—(1) bilateral conjunctival injection, (2) generally non purulent, changes in mucosa of oropharynx, including injected oropharynx, injected and dry fissured lips, strawberry tongue, (3) changes of the peripheral extremities such as edema

at 4 ml/kg/Hr can be infused with the 10% concentration starting at 0.5 ml/kg/Hr. If no adverse effects occur, the rate can be increased gradually up to a maximum of 8 ml/kg/Hr [18–21].

Our case had the typical presentation consistent with the diagnostic criteria for KD. On enquiry it was revealed that their family used humidifiers, the association of which with KD has been stated by Treadwell et al. [12]. She developed a systolic murmur during the course of her illness. It was confirmed by echocardiography showing a grade I mitral regurgitation which is a complication reported in other article by Raju Khubchandani and Susan D'Souza in 2004 [22]. She had been given intravenous immunoglobulin and high dose of aspirin to which she responded promptly with rapid defervescence, which was in accordance with the finding of Dajani et al. [11]. Chantepie et al. [10] noted that an important and fatal complication of the disease is rupture of coronary artery aneurysm, which luckily could be evaded in this case because of the timely institution of therapy.

Conclusion

From ENT perspective an awareness of Kawasaki disease is important as the condition can present with cervical lymph node swelling and fever with other signs and symptoms. We report this case because mitral regurgitation is not a known complication of Kawasaki disease.

A clinician must have a high index of suspicion and knowledge of the clinical features of the KD. Early institution of therapy with intravenous IgG minimizes the risk of serious complications. Clinical judgement is the mainstay in diagnosis and appropriate treatment results in complete recovery without any long-term side effects.

References

1. Laupland KB, Dele Davis H (1999) Epidemiology, etiology, and management of Kawasaki disease: state of art. *Pediatr Cardiol* 20(3):177–183
2. Treadwell TA, Maddox RA, Homan RC et al (2002) Investigation of Kawasaki syndrome risk factors in Colorado. *Pediatr Infect Dis J* 21(10):976–978
3. Nomura Y, Masuda K, Yoshinaga M et al (2004) Possible relationship between streptococcal pyrogenic exotoxin A and Kawasaki syndrome in patients older than six months of age. *Pediatr Infect Dis J* 23(8):789–791
4. Shingadia D, Bose A, Booy R (2002) Could a herpes virus be the cause of Kawasaki disease? (Review). *Lancet Infect Dis* 2(5):310–313
5. Guzman-Cottrill JA, Shulman ST (2004) Recent developments and controversies in Kawasaki disease. *Minerva Pediatr* 56(1): 51–61
6. Taubert KA, Rowley AH, Shulman ST (1995) In: Kato Hed KD (ed) A 10 year (1984–1993) US hospital survey of Kawasaki disease, 1st edn. Elsevier Science, Amsterdam, pp 34–38
7. Pannaraj PS, Turner CL, Bastian JF et al (2004) Failure to diagnose Kawasaki disease at the extremes of the paediatric age range. *Pediatr Infect Dis J* 23(8):789–791
8. Brown TJ, Crawford SE, Cornwall M et al (2001) CD8 T cells and macrophage infiltrate coronary artery aneurysms in acute Kawasaki disease. *J Infect Dis* 184:940–943
9. Burns JC, Glode MP (2004) Kawasaki syndrome (review). *Lancet* 364(9433):533–544
10. Chantepie A, Mauran P, Lusson JR et al (2001) Cardiovascular complications of Kawasaki syndrome: results of a French multicentre study. *Arch Pediatr* 8(7):713–719
11. Dajani AS, Taubert KA, Takghashi M et al (1994) Guidelines for long-term management of patients with Kawasaki disease. *Circulation* 89:916–922
12. American Heart Association (2001) Council on CVD in young, committee on rheumatic fever, endocarditis and Kawasaki disease: diagnostic guidelines for Kawasaki disease. *Circulation* 103:335–336
13. Burns JC, Kushner HI, Bastian JF, Shike H, Shimizu C, Matsubara T, Turner CL (2000) Kawasaki disease—a brief history. *Paediatrics* 106(2):e27
14. Chang RK (2002) Epidemiologic characteristics of children hospitalized for Kawasaki disease in California. *Paediatr Infect Dis J* 21(12):1150–1155
15. Gardener-Medwin JM, Dolezalova P, Cummins C, Southwood T (2002) Incidence of Henoch-Schönlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. *Lancet* 360(9341):1197–1202
16. Brogan PA, Bose A, Burger D, Shingadia D, Tulloh R, Michie C et al (2002) Kawasaki disease: an evidence based approach to diagnosis, treatment and proposal for future research. *Arch Dis Child* 86(4):286–290
17. Stiehm ER (1979) Standard and special human immune serum globulins as therapeutic agents. *Pediatrics* 63:301–319
18. Furusho K, Sato K, Soeda T et al (1983) High dose intravenous gammaglobulin for Kawasaki disease [letter]. *Lancet* 2:1359
19. Nagashima M, Matsushima M, Matsucka H, Ogawa A, Okumura N (1987) High dose gammaglobulin therapy for Kawasaki disease. *J Pediatr* 110:710–712
20. Engle MA, Fatica NS, Bussell JB, O'Laughlin JE, Snyder MS, Lesser ML (1989) Clinical trial of single-dose intravenous gammaglobulin in acute Kawasaki disease. *AJDC* 143: 1300–1304
21. Isawa M, Sugiyama K, Kawase A et al (1986) Prevention of coronary artery involvement in Kawasaki disease by early intravenous high dose gammaglobulin. In: Doyle EF, Engle MA, Gersony WM, Rashkind EJ, Talner NS (eds) *Pediatric cardiology*. Springer-Verlag, New York, pp 1083–1085
22. Khubchandani R, D'Souza S (2004) Kawasaki disease in India. *Pediatr Rheumatol* 2(2):162–171