Study of erroneous diagnoses of Kawasaki disease

Meimei Zhang*

The Fifth Affiliated Hospital of Zhengzhou University, Zhengzhou, Henan 450001, China

Received January 29, 2009

Abstract

Kawasaki's disease (KD) is an acute febrile illness of infants and children in which redness of the mucous membrane; skin and tongue are associated with swelling and desquamation of the hands and feet. Forty-four samples of the erroneous diagnosis of Kawasaki disease were studied. The coronary artery pathological change was found very few in regular diagnosis. On the other hand, the most often erroneous diagnoses of Kawasaki disease being as hypersensitivity vasculitis, upper respiratory tract infection, cervical lymphadenopathy, acute nephritis and scarlatina are learned and demonstrated in this report. [Life Science Journal. 2009; 6(2): 90 – 92] (ISSN: 1097 – 8135).

Keywords: Kawasaki's disease; febrile illness; mucous membrane; diagnosis; hypersensitivity vasculitis; cervical lymphadenopathy; acute nephritis; scarlatina

1 Introduction

Some signs include swollen lymph nodes of the neck, redness and swelling of the eyes, sores in the mouth (stomatitis) and swollen lips (chelitis). After a week or two the skin of the hands and feet begin to peel starting around the nails. Upwards of 20% of patients develop coronary artery complications, however most patients have uneventful recoveries without any long term problems. The peak age of incidence is 1 year of age with a mean of 2.6 years and it is uncommon over 8 years of age. The cause of Kawasaki's disease (KD) is not known and is generally seen during the winter and spring of the year. There are 3 phases of the disease. Phase I: Abrupt onset of fever, lasting around 12 days or so, followed by most of the principal symptoms of the disease condition. This phase develops a red rash usually first seen on the palms and soles that then spreads to involve the torso within a couple days. The most common appearance is a hive-like rash; however it may also resemble measles (morbilliform rash), erythema multiforme or a scarletina like rash. It is more impressive on the hands and feet than the torso and the hands and feet generally develop some swelling as well. Phase II: This is a subacute phase can last around 30 days during which fever, arthritis that and arthralgia, thrombocytosis, desquamation, and carditis generally resolve. This is the phase with the highest risk of sudden death. The desquamation occurs in this phase with significant peeling of the hands and feet starting at the tips of the digits. In addition most of the dangerous cardiac abnormalities occur such as dysrhythmias, heart failure and left ventricular dysfunction. Phase III: This is the convalescent period and generally starts 8 – 10 weeks after the beginning of the illness; it basically starts when all symptoms resolve and lasts until all tests have returned to normal. Icelandic Moss (Cetraria islandica) has been used to treat inflammation and dryness of the pharyngeal mucosa in the naturopathy for many years. It contains substances with protective and antiinflammatory effects for the oral mucosa[1]. It was reported that the patient with advanced gastric cancer with Virchow's lymph node metastasis who successfully received curative resection following neoadjuvant chemotherapy with a single oral anticancer drug[2].

The diagnosis of KD depends on several criteria. First there must be an elevated temperature of greater than 102.50 F for at least 5 days; then 4 out of 5 of the following:

1) Bilateral conjunctival injection (redness of eyes); 2) At least one swollen lymph node in the anterior cervical lymph nodes (front of neck); 3) A widespread scarlet fever like erythroderma (redness of the skin), with areas

*Corresponding author. Email: Meimei20082008@gmail.com

of sharply marginated rash, deeply erythematous maculopapular rash and iris lesions; 4) At least one of the following: erythema of the palms and soles, edema of the hands and feet, generalized desquamation around the tips of the fingers and toes; 5) At least one of the following changes found in and around the mouth: injected/fissured lips, injected pharynx (back of the mouth) and "strawberry" tongue (a bright red appearance of the tongue).

The majority of people with KD recover without long term complications. However about 20% of patients develop some form of vascular involvement such as: coronary artery aneurysms, myocarditis, heart attack (myocardial infarction), peripheral vascular occlusion, small bowel obstruction or stroke. Historically about 1% of patients die from complications of KD.

Early diagnosis is critical so as to try and prevent cardiovascular complications. It is generally accepted that failed infrainguinal bypass with prosthetic material significantly compromises arterial run off, which may limit future revascularization. It is well known that the negative consequences of early vein graft thrombosis are limited, but the effect of failed peripheral angioplasty on the distal vasculature is poorly studied[3]. It was reported that A coronary stent may be lost in the peripheral or visceral arterial system with an incidence ranging from 0.9 to 8.4%, however, a limb or organ ischemia after stent migration is very uncommon[4]. Patients should be hospitalized during the phase I period to monitor for complications. The primary treatment is aspirin 100 mg/kg/day until the fever has passed, after which the dose is reduced to 5 – 10 mg/kg/day until all lab tests return to normal. High dose intravenous gamma globulin (IVIG) has been used to reduce the risk of coronary aneurysms and myocardial infarction. KD is also named as mucocutaneous lymph node syndrome found by Dr. Kawasaki in 1967 at Japan. The special pathognomics of KD include coronary vasculitisand coronary vasculitis. The male to female ratio of morbidity of KD is about 1.5% and 80% of the patients being under five years old. The mortality is 0.3% to 0.5%. Rate of recrudesce is about 3%.

2 Data Acquisition and Analysis

Forty-four samples were collected where 30 are males and 14 females. The ages of the patients were between 3 months to seven years old. The male to female ratio is 2.1. Thirty two samples are for the ages between 1 year to 3 years old, 72.7% of the total forty four samples. In Table 1, it depicts the clinical manifestation of the 44 samples.

Table 1. List of KD-clinical manifestation

Description	Percentage
Fever,7 – 15 days averagely 9.4 days	44 (100%)
Edema, hands and feet	39 (88.6%)
Edema, hands and feet in 7 days	41 (93.2%)
Bulbus oculi tunica in 7 days	44 (100%)
Chapped shin in 7 days	44 (100%)
Scaly rash	34 (77.3%)
Anus decrustation	17 (38.6%)
Neck lymph glad swelling	40 (90.9%)
Proteinuria	13 (29.5%)
Leukocytosis	33 (75%)
Thrombocythemia	34 (77.3%)

3 Erroneous Diagnoses of KD

Erroneous diagnoses of KD may happen due to the clinical manifestations look the same with other diseases. The possibilities of the erroneousness include upper respiratory tract infection, cervical lymphadenopathy, acute nephritis, scarlatina and medical rash etc. Table 2 lists several erroneous diagnoses of KD in our study.

All the patients listed were cured by administration with oral aspirin, cortex and IVID.

Table 2. Erroneous diagnoses of KD

Erroneous diagnoses	Specimen (percentage)
Upper respiratory tract infection	1 (47.7%)
Cervical lymphadenopathy	3 (6.8%)
Acute nephritis	1 (2.3%)
Medical rash	2 (4.5%)
Scarlatina	1 (2.3%)
Pulmonitis	1 (2.3%)

4 Discussion

Heart failure (HF) decompensation continues to account for approximately 1 million hospitalizations per year in the United States. Pulmonary congestion is the hallmark sign of worsening HF[5].

The origin of disease of KD is still not clear. After the massive epidemiology research, a preferred thought is that KD is an immunity disease caused by one kind or many kinds of pathogenic microorganism that enters into human immune system. Although KD has the self-

recovery tendency, its coronary artery expansion and the coronal aneurism formation rate reaches as high as 25% to 40%[7]. KD is an acute self-limited vasculitis of childhood that is characterized by fever, bilateral nonexudative conjunctivitis, erythema of the lips and oral mucosa, changes in the extremities, rash, and cervical lymphadenopathy. Coronary artery aneurysms or ectasia develop in approximately 15% to 25% of untreated children and may lead to ischemic heart disease or sudden death[6]. Because of being deficient in the specificity diagnosis standard, early diagnosis receives certain limitation. The rate of erroneous diagnosis appears very high. Recommendations for the initial evaluation, treatment in the acute phase, and long-term management of patients with KD are intended to assist physicians in understanding the range of acceptable approaches for caring for patients with KD. The ultimate decisions for case management must be made by physicians in light of the particular conditions presented by individual patients[6].

Group case of illness gets reliable diagnose in three days being only 16.25%. Misdiagnoses diseases reach to six different kinds of erroneous diagnosis. The upper respiratory tract infection is a common and frequently-occurring disease to be in erroneous diagnosis of KD. The early symptoms including high fever, cornea hyperemia red, cervical lymphadenopathy must be distinct from KD[8,9]. Specifically, diagnoses of KD may show similarity of the upper respiratory tract infection skin rash, cornea hyperemia only happens in single side and echocardiogram inspection lacks of the confirmation of coronary artery harm etc. Scarlet fever may be another erroneous diagnosis for similar clinical early symptoms. The major symptoms including the skin rash, rubra must be distinct from KD. Basically, scarlatina should have the contact history. The skin rash of scarlatina may have its characteristic value. Etiology inspection can exhibit a group of second grade hemolytic streptococcus as the dissimilarity.

The childhood reheumatoid arthritis can be seen in 2 – 4 years old babies and infants. The symptom begins with high fever, accompanies by polymorphic skin rash, lymph node tumescent and heart damage. None hand and foot rigid dropsy with typical coronary artery damage can be found in our cases study. However, mainly because the acquisition of information being in short of analysis of the early echocardiogram, the diagnosis may not be able to find the temporary coronary artery expands. Most of the cases, patients recovered in 30 days.

Our study revealed especially that KD needs a long-term observation. KD has the peak contract to damage the coronary artery in course of 15 days. Patients leave the hospital may still keep most of the trouble of having coronary artery expansion and the aneurism danger. Early diagnosis, treatment, close observation of the patients enhance the KD curing rate as well as reducing the possibilities to damage the coronary artery.

References

1. Kempe C, Gruning H, Stasche N, Hormann K. [Icelandic moss lozenges in the prevention or treatment of oral mucosa irritation and dried out throat mucosa]. Laryngorhinootologie 1997; 76: 186 – 8.
2. Iwazawa T, Kinuta M, Yano H, Matsui S, Tamagaki S, Yasue A, Okada K, Kanoh T, Tono T, Nakano Y, Okamoto S, Monden T. An oral anticancer drug, TS-1, enabled a patient with advanced gastric cancer with Virchow's metastasis to receive curative resection. Gastric Cancer 2002; 5: 96 – 101.
3. Joels CS, York JW, Kalbaugh CA, Cull DL, Langan EM, Taylor SM. Surgical implications of early failed endovascular intervention of the superficial femoral artery. J Vasc Surg 2008; 47: 562 – 5.
4. Siani A, Siani LM, Mounayergi F, Baldassarre E. Lower limb ischemia after migration of a coronary artery stent into the femoral artery. Interact Cardiovasc Thorac Surg 2008; 7: 447 – 8.
5. Daleiden-Burns A, Stiles P. Proactive monitoring: implications of implantable devices for future heart failure management. Crit Care Nurs Q 2007; 30: 321 – 8.
6. Newburger JW, Takahashi M, Gerber MA, Gewitz MH, Tani LY, Burns JC, Shulman ST, Bolger AF, Ferrieri P, Baltimore RS, Wilson WR, Baddour LM, Levison ME, Pallasch TJ, Falace DA, Taubert K A. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Circulation 2004; 110: 2747 – 71.
7. Wang HW. Questions of the cornary damages by Kawasaki disease. Journal of Chinese Pediatrics 2006; 21(10): 730 – 1.
8. http://www.nlm.nih.gov/medlineplus/ency/article/000989.htm
9. http://www.americanheart.org/presenter.jhtml?identifier=4634.