

Incomplete Kawasaki Disease Presenting with Pancarditis and Pericardial Effusion

Pankardit ve Perikardiyal Efüzyon ile Presente Olan İnkomplet Kawasaki Hastalığı

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ABSTRACT

Kawasaki disease is an acute self-limiting systemic vasculitis. Although its etiology is unknown, some infectious agents are strongly suspected. Untreated patients may develop severe complications, such as coronary artery aneurysm, congestive heart failure, myocardial infarction, and cardiac dysrhythmia. In this report, eight years-old girl, diagnosed as incomplete KD with pericardial effusion without conjunctivitis and lymphadenopathy, was presented.

Key words: Incomplete Kawasaki disease, Pancarditis, Pericardial effusion

ÖZET

Kawasaki hastalığı akut, kendini sınırlayan sistemik bir vaskülitir. Etyolojisi kesin olarak bilinmemekle birlikte enfeksiyon ajanlarından şüphelenilmektedir. Tedavi edilmemiş hastalarda koroner arter anevrizması, konjestif kalp yetmezliği, miyokardiyal enfarktüs ve aritmi gibi komplikasyonlar gelişebilmektedir. Bu yazıda perikardiyal efüzyonu olup konjunktivit ve lenfadenopatisi olmayan inkomplet Kawasaki hastalığı tanısı konulan sekiz yaşında kız olgu sunulmuştur.

Anahtar kelimeler: İnkomplet Kawasaki hastalığı, Pankardit, Perikardiyal efüzyon

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INTRODUCTION

Kawasaki disease (KD) is an acute self-limiting systemic vasculitis although its etiology is unknown, an infectious agent is strongly suspected (1). Genetic predisposition is also likely and seen more commonly among Asians. KD is primarily afflicts infants and young children (2,3). It was first described by Kawasaki in 1967 in

Japan. There is any pathognomonic clinical or laboratory finding for the diagnosis (4). Patients who do not fulfill the classical diagnostic requirements and lack some of the cardinal clinical features but presenting the laboratory findings which stated in guidelines are said to have incomplete or atypical KD (5,6). Untreated

patients may develop severe complications (20% to 25%), such as coronary artery aneurysm, congestive heart failure, myocardial infarction, and cardiac dysrhythmia (7). In this report, we describe an 8 years-old girl diagnosed as incomplete KD with pericardial effusion without conjunctivitis and lymphadenopathy.

CASE REPORT

An 8-old girl was admitted to our hospital with history of high fever and irritability for 10 days duration. Physical examination revealed high fever of 39° C, heart rate of 135/bpm, respiratory rate of 24 breaths per minute, and a blood pressure of 90/60 mmHg. She had oral and pharyngeal erythema, strawberry tongue, generalized erythematous rash, oral apths, edema and arthritis on the left hand, knees and foot, tachycardia with no apparent murmur (Fig. 1-3). There were no signs of meningeal irritation, conjunctivitis, lymphadenopathy and hepatosplenomegaly.



Fig. 1. Top left: Arthritis and edema of dorsum of left hand, Top right: arthritis of left knee, Bottom left: arthritis of left ankle, Bottom right: arthritis of right knee.



Fig. 2. Oral apths, strawberry tongue and angular cheilitis at the time of admission



Fig. 3. Dermal desquamation in first day of admission

Laboratory workup was initiated for suspected infections and rheumatologic causes. White blood cell count was $22.8 \times 10^3/\mu\text{L}$ with 80% neutrophils, 20% lymphocyte, hemoglobin level of 9.76 g/dl and a platelet count of $318.000/\text{mm}^3$. Erythrocyte sedimentation rate (ESR) was 36 mm/h, C-reactive protein (CRP) 239 mg/L ($N < 5 \text{ mg/L}$), total protein 5.4g/dl, albumin 2.5 g/dl, normal levels of serum alanine aminotransferase (17 iu/l), gamma glutamyl transferase and electrolytes levels. Her blood, throat and urine cultures were negative. She had

steril pyuria. Coxsackie Ig M was negative. The abdominal ultrasonography was normal. Electrocardiogram (ECG) and transthoracic echocardiography were in normal limits on admission.

After cultures were taken, the patient was put on intravenous ceftriaxone empirically for unknown fever with bacterial etiology. On the 3th day of the treatment, fever remained almost unchanged with any improvement of sign and symptoms. On the 3th day of admission she developed 1/6 systolic murmur and friction rub. Platelet count was 318.000/mm³. Control echo showed mitral insufficiency and pericardial effusion (2-3mm).

Because of the prolonged history of unexplained fever, per oral cheilitis, oral aphts, strawberry tongue, pharyngeal erythema, dermal rash and desquamation in the abdomen and inguinal region, edema and arthritis on the left hand and foot, extremely high CRP level and mild elevation in ESR, sterile pyuria, hypoalbuminemia, thrombocytosis, endo-myocarditis (tachycardia, murmur and friction rub) incomplete Kawasaki disease was considered in this case although lacking some of cardinal features of disease. The antibiotic was stopped and per oral aspirin (80 mg/kg/day) was administered while waiting for obtaining intravenous immunoglobulin (IVIG). The fever resolved next day immediately after started aspirin. Erythematous skin rash, fatigue, pericardial effusion, tachycardia and arthritis completely regressed in 2-3 days. On the 3th day of aspirin treatment, CRP and sedimentation levels decreased, albumin level was elevated, arthritis, angular cheilitis and aphthous stomatitis were subsided dramatically. The dermal desquamation improved (Fig. 4,5).

Echocardiographic findings were within normal limits, with any positive findings regarding mitral insufficiency, pericardial effusion and coronary artery aneurism. Thrombocytosis was developed after 4th day of aspirin treatment (600.000/mm³). Patient was put on aspirin 3mg/kg/day and discharged. Later she visited for three times with one month apart. During follow-up she was doing well. Physical exam, ECG and echocardiographic findings were in normal limits.



Fig. 4. Oral appearance one week after treatment



Fig. 5. Dermal desquamation one week after treatment

DISCUSSION

Kawasaki disease (KD) is an acute, self-limited vasculitis. It occurs predominantly in infant and young children (5). Diagnostic criteria are summarized and prepared by Japanese Kawasaki Disease Research Committee (3), and The American Heart Association (AHA) (1). Fever for more than 5 days along with four or more of following findings without any alternative explanation for the findings may establish the diagnosis:

1. Bilateral, nonpurulent conjunctivitis;
2. Erythematous mouth and pharynx, strawberry tongue, and red, cracked lips;
3. A polymorphous, generalized, erythematous rash that can be morbilliform, maculopapular, or scarlatiniform or may resemble erythema multiforme;
4. Abnormalities of the extremities consisting of induration of the hands and feet with erythematous palms and soles, often with later periungual desquamation; and
5. Acute, nonsuppurative, usually unilateral, cervical lymphadenopathy with at least 1 node measuring 1.5 cm in diameter (4,5,8,9).

For the diagnosis of incomplete KD, guideline developed by AHA can be used (5).

In our case, despite lacking lymphadenopathy, organomegaly and conjunctivitis, based on prolonged fever resistant to treatment along with oral and pharyngeal erythema, strawberry tongue, erythematous maculopapular rash, indurations of hands and feet, supporting by laboratory findings, incomplete Kawasaki disease was considered. She developed pancarditis on the 3th day of admission (13th day of diseases). Fever, pericardial effusion and murmur surprisingly disappeared immediately after receiving aspirin (80mg/kg/day) without

IVIG treatment. Sonçağı et al. reported that pericardial effusion improved

with IVIG administration (10). Although we obtained IVIG in 24 hours after starting aspirin therapy, we did not infuse it because fever, murmur and pericardial effusion disappeared (shown by echocardiography) dramatically and arthritis subsided and other clinical findings, including oral aphthae and cheilitis, regressed significantly.

In a prospective study conducted by Chantepie et al, noted pericardial involvement in 12 of 94 KD cases (%24) (11). Another study reported pericardial effusion in 6.3 % of 96 patients with KD and myocarditis is only one case (12). In a case despite IVIG and high dose methylprednisolone treatment, the patient developed cardiac tamponade and died hours later (13).

Laboratory findings are almost the same in both typical and incomplete cases. In our patient laboratory findings were consistent with KD. She had high leukocytosis with neutrophils predominance, hypoalbuminemia, sterile pyuria and elevated sedimentation rate and extremely high CRP levels. Thrombocytosis may occur in KD three to four weeks after clinical onset and in our case it occurred 14-15th day of disease was developed. It is known that coronary aneurysm may develop in patients with KD in children under five years of age more frequently. Such a complication was not observed in our patient during 3 months follow-up.

In conclusion, pancarditis may develop in untreated incomplete KD. Aspirin therapy should not be delayed in case of lacking IVIG.

REFERENCES

1. Freeman AF, Shulman ST, Kawasaki disease: summary of the American Heart Association guidelines. *Am Fam Physician* 2006 1;74:1141-8.
2. Kawasaki T. Acute febrile mucocutaneous syndrome with lymphoid involvement with specific desquamation of the fingers and toes in children. *Arerugi* 1967;16:178–222.
3. Kawasaki T, Kosaki F, Okawa S, Shigematsu I, Yanagawa HA. New infantile acute febrile mucocutaneous lymph node syndrome (MLNS) prevailing in Japan. *Pediatrics* 1974;54:271-6.
4. Barone SR, Pontrelli LR, Krilov LR. The differentiation of classic Kawasaki disease, atypical Kawasaki disease, and acute adenoviral infection: use of clinical features and a rapid direct fluorescent antigen test. *Arch Pediatr Adolesc Med* 2000;154:453-6.
5. Newburger JW, Takahashi M, Gerber MA, Gewitz MH, Tani LY, Burns JC, et al. Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. 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. *Pediatrics* 2004;114:1708-33.
6. Manlhiot C, Christie E, McCrindle BW, Rosenberg H, Chahal N, Yeung RS. Complete and incomplete Kawasaki disease: two sides of the same coin. *Eur J Pediatr* 2012;171:657-62.
7. Dahlem PG, von Rosenstiel IA, Lam J, Kuijpers TW. Pulse methylprednisolone therapy for impending cardiac tamponade in immunoglobulin-resistant Kawasaki disease. *Intensive Care Med* 1999;25:1137-9.
8. Fujita Y, Nakamura Y, Sakata K, Hara N, Kobayashi M, Nagai M, et al. Kawasaki disease in families. *Pediatrics* 1989;84:666-9.
9. Sonobe T, Kiyosawa N, Tsuchiya K, Aso S, Imada Y, Imai Y, et al. Prevalence of coronary artery abnormality in incomplete Kawasaki disease. *Pediatr Int* 2007;49:421-6
10. Sonçağı A, Devrim I, Karagöz T, Dilber E, Celiker A, Ozen S, et al. Septated pericarditis associated with Kawasaki disease: a brief case report. *Turk J Pediatr* 2007;49:312-4.
11. Chantepie A, Mauran P, Lussion JR, Vaillant MC, Bozio A. Cardiovascular complications of Kawasaki syndrome: results of a French multicenter study *Arch Pediatr* 2001;8:713-9.
12. Salice P, Pietrogrande MC, Barbier P, Ghiglia S, Laicini E, Fesslova V. [Cardiovascular abnormalities in Kawasaki disease. An Italian prospective study]. *Cardiologia*. 1998;43:1367-74.
13. Ozdogu H, Boga C. Fatal cardiac tamponade in a patient with Kawasaki disease. *Heart Lung* 2005;34:257-9.