Photo Quiz

http://journals.sbmu.ac.ir/jpn
DOI: https://doi.org/10.22037/jpn.v7i4.28409

A 4-month-old Female Infant with 36 Hours of Persistent High-Grade Fever

A family physician referred a 4-month-old Caucasian female infant to the pediatric emergency department because of 36 hours of persistent high-grade fever (T 39.2°C).

Her mother mentions that before the fever began, she was well, feeding normally on breast milk. The fever started suddenly, peaked within a few hours and remained high despite receiving enough doses of oral acetaminophen. She had no other symptoms during these 36 hours, except intermittent lethargy and agitation. She is the result of a term birth with a birth-weight of 2970 g. She has two elder brothers (8 and 3 years, healthy). Her parents are healthy and non-consanguineous (Her mother is 29, and her father is 35 years old). Her general condition is poor probably because of high fever (T 39.2°C). However, she has normal growth and development. On physical examination, the heart rate is 150/min and the respiratory rate is 56/min. The conjunctiva is mildly erythematous in both eyes (Figure 1) and the pharynx is hyperemic. No peripheral lymph node is palpable. The heart, lung and abdomen examination are normal. The muscle tone is normal. There are no meningeal signs.

The laboratory tests on admission shows: Hemoglobin: 9 g/dL (reference 9.5-14.1 g/dL), red blood cells:3.35X10^6/mm^3 (reference 3.1-5.1X10^6/mm^3), MCV: 84 fl (reference 74-108 fl), WBC count: 26.1X10^3/mm^3 (reference 6-17X10^3/mm^3), neutrophils: 63% (reference <48%), platelet count:186X10^3/mm^3) (reference 150-250X10^3/mm^3), glucose: 85 mg/dL (reference 50-90 mg/dL), bilirubin: 0.67 mg/dL (reference <1.2 mg/dL), blood urea nitrogen: 13.4 mg/dL (reference 5–18 mg/dL), creatinine:0.36 mg/dL (reference 0.2-0.4 mg/dL), total protein:4.9 g/dL (reference 5.1–7.3 g/dL), albumin:2.7 g/dL (reference 2.2–4.8 g/dL), aspartate aminotransferase: 52 U/L (reference 9–80 U/L), alanine aminotransferase: 36 U/L (reference 13–45 U/L), C-reactive protein (CRP):14.23 mg/dL (reference <0.5 mg/dL). Chest radiography is unremarkable. Abdominal ultrasound reveals slight hepatosplenomegaly and mild peritoneal effusion.

What is your diagnosis?

Figure 1

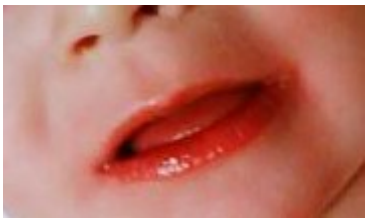

Figure 2

Figure 3

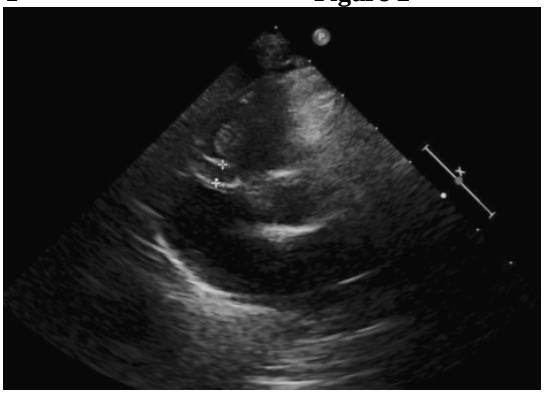

Figure 4

A 4-month-old Female Infant with 36 Hours of Persistent High-Grade Fever

Mehrnoush Hassa Yeganeh*

Pediatric Nephrology Research Center, Research Institute for Children Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Please cite this article as: Hassas Yeganeh M. A 4-month-old Female Infant with 36 Hours of Persistent High-Grade Fever. J Ped Nephrol. 2019;7(4).

https://doi.org/10.22037/jpn.v7i4.28409

*Corresponding Author
Mehrnoush Hassas Yeganeh, MD.
Email: mehrnoushyeganeh@gmail.com

The patient is initially treated with intravenous antibiotic therapy (ceftriaxone) and acetaminophen. Despite IV antibiotic therapy, the fever persists (39.4^{o}C) and there is no change in her condition. After 48 hours of antibiotic therapy, a generalized erythematous macular rash appears on the trunk and extends to extremities. Generalized edema appears in the trunk and extends rapidly to extremities including hands and feet. Conjunctivitis develops in both eyes. Lips become red, and the tongue is inflamed and red (Figures 2 and 3). Blood and urine cultures are negative after 48 hours. On echocardiography, the right coronary artery (RCA) is within the maximum limits of the normal range. A small pericardial effusion was also detected.

We administer IVIG (2 g/kg administered as a single infusion over 8 to 12 hours) and aspirin (80 mg/kg per day in divided doses) with the diagnosis of Kawasaki Disease.

Nevertheless, the child continues to be febrile, and the rash continues to extend. New echocardiography performed after 48hours from the first dose of IVIG shows dilatation of RCA (2.8 mm). We administer the second dose of IVIG. However, following this second dose of IVIG, the patient still has a high fever and develops fissuring of the lips, ectasia of left coronary artery (LCA) and a worsening of the dilatation of RCA (RCA: 3.9 mm; LCA: 3.1 mm) (Figure 4).

Lab tests show platelet count to increase to 303X10^{3}/mm^{3} and a further increase of CRP value (15.96 mg/dL).

Q1: What is the reason for not responding to the treatment?

A1: The treatment was not sufficient, because the age <12 months is a single risk factor for high-risk Kawasaki disease; the patient should have been additionally treated with systemic glucocorticoids (1,2). We start IV methylprednisolone (IVMP, 30 mg/kg/day). After 24 hours of beginning IVMP, the body temperature begins to decrease. However, in new lab tests after four days, aspartate aminotransferase increases to 267 IU/L, alanine aminotransferase increases to 312 IU/L. The echocardiography after five days shows the maximum diameters were 6.3 mm for RCA, 4.1mm for LCA (2,3).

Q2: What is the best plan for her now?

A2: Because of the marked increase of transaminase levels, we gradually reduce the dose of aspirin to 40 mg/kg/day (2,4).

As long as fever or coronary vasculitis is not adequately controlled, IVMP therapy is continued for at least five days to ensure that absorption of glucocorticoids is optimized. Patients are switched from intravenous to oral glucocorticoids to cover a total of 15 days of corticosteroid, five days on prednisone 2 mg/kg, and five days on prednisone 1 mg/kg. CRP is measured two to three times per week until the concentration is ≤5 mg/L. When the inflammatory markers become normal and clinical picture improves, aspirin can be changed to clopidogrel (1mg/Kg/day) its anti-aggregating effects because of the persistence of the ectasia and the dilatation of the coronary arteries (2,4,5).

References:

1. Sundel R, Kawasaki disease: Clinical features and diagnosis - UpToDate. 2019:1-12.
2. Sundel R, Kawasaki disease: Initial treatment and prognosis - UpToDate. 2019:1-12.
3. Hassas Yeganeh M. A 4.5-year-old boy with Diffuse Purpuric Lesions all Over his Extremities. J Ped Nephrol. 2019;7(3).
4. Kobayashi T, Inoue Y, Takeuchi K, et al. Prediction of intravenous immunoglobulin unresponsiveness in patients with Kawasaki disease. Circulation. 2006;113(22):2606–2612.
5. Kobayashi T, Saji T, Otani T, et al. RAISE Study Group Investigators. Efficacy of immunoglobulin plus prednisolone for prevention of coronary artery abnormalities in severe Kawasaki disease (RAISE study): a randomized, open-label, blinded-endpoints trial. Lancet. 2012;379(9826):1613–1620.