Giant right coronary artery aneurysm secondary to Kawasaki disease in an infant

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Kawasaki disease (KD) is a potentially decapacitating multisystemic vasculitis with unknown etiology that acquired worldwide attention due to associated coronary aneurysms leading to life-threatening complications in very young babies including thrombosis, ischemia, and rupture. High levels of suspicion for early diagnosis and prompt treatment are crucial in preventing serious complications. We report here one of the patients who developed a giant coronary aneurysm but fortunately not a life-threatening complication after 5 years of follow-up. We conclude that later intravenous immunoglobulins (IVIG) treatment could be an important factor—among others—that precipitate into such complications.

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1. Introduction

Kawasaki disease (KD) was first described in Japan in 1961 by Dr. Tomisaku Kawasaki and is now encountered worldwide [1]. One-fourth of patients developed coronary dilatation but only ~2% died suddenly after the acute stage due to myocardial infarction caused by acute thrombosis within coronary artery aneurysms or rarely from aneurysmal rupture [2]. The persistence of giant coronary artery aneurysms (with internal diameters >8 mm) decreases to 0.5% – 1% if the patients are treated adequately and in time [3].

A few small studies that addressed the incidence and prevalence of KD in Kingdom of Saudi Arabia showed no significant differences between national statistical data than what is observed worldwide [4,5].

2. Case report

A 5-month-old infant was admitted to our center with a history of persistent fever for 3 weeks. She also had a history of polymorphic skin rash, red eyes, cracked lips, and skin peeling that started at the periungual area. She was treated with antibiotics for a suspected throat infection in a local hospital. At admission she
was irritable with skin peeling on her toes and fingers.

The initial laboratory findings showed hemoglobin 7.5 g/dL, white cell count (WBC) 16,200 ($n = 36\%$, $L = 48\%$, mono = 13\%, others = 4\%) platelets 1,356,000/mm, total protein 77 g/L [normal range (NR) 64–83 g/L] albumin 35 g/L (NR 35–50 g/L), erythrocyte sedimentation rate 43 mm/h (NR up to 20 mm/h), C-reactive protein 44.5 mg/L (NR 1–3 mg/L), prothrombin time (PT) 11 seconds (NR 11–15 seconds), activated partial thromboplastin time (aPTT) 33 seconds (NR 28–41 seconds), Beta-natriuretic peptide (BNP) 149 pmol/L (NR up to 15.3 pmol/L), and creatine kinase (CK) 42 U/L (NR 26–192 U/L). Sepsis workup including blood and urine cultures were unremarkable. Echocardiography did not show signs of ischemia.

Echocardiography showed (Fig. 1A and B) gross dilatation of the left coronary artery with multiple large aneurysmal dilatations and a possible small clot, giant Right Coronary Artery aneurysm, preserved LV systolic function, and moderate pericardial effusion (PE) without signs of tamponade.

Five days after admission; CKMB/CK was 28/31 U/L (NR for CKMB up to 25 U/L) and Troponin T was 27 ng/L (NR = up to 14 ng/L). CT angiography confirmed the echocardiography findings.

Even though it was a late treatment the patient received one dose of IVIG 2 g/kg, and kept on aspirin 5 mg/kg/day, heparin 20 IU/Kg/hr with target aPTT 50–60 seconds, and warfarin 1 mg orally once daily with a target international Normalization Ratio (INR) of 2–3.

After 16 days of warfarin, the medication was changed to enoxaparin S/C 1 mg/kg/dose 12 hourly due to difficulty to keep the target INR range.

The patient remained clinically stable and serial echocardiography revealed persistent giant RCA aneurysm with formation of small shell of thrombosis appearing anteriorly, and gradual improvements of LCA dilatations and aneurysms, as well as normalization of other laboratory tests.

The patient was not found to be a candidate for additional therapy and was therefore discharged on aspirin and enoxaparin and followed up in the outpatient department (OPD).

Then at the age of 18 months the patient was admitted electively for cardiac catheterization as part of a giant coronary aneurysm follow up. Coronary angiography showed giant RCA aneurysm (19.9 × 26 mm) with fusiform medium sized LAD aneurysm (6.9 × 10.2 mm) (see Fig. 2) but no coronary obstruction.

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**Figure 1.** Short axis parasternal echocardiographic view showing (A) a giant Right Coronary Artery and multiple LCA aneurysms and (B) multiple Left Coronary Artery aneurysms.

**Figure 2.** Aortic root angiography showing a giant Right Coronary Artery with a small Left Anterior Descending artery aneurysm.
The patient was followed up in the OPD and on the last visit at 5 years and 5 months of age the patient was doing well. Echocardiography showed persistent giant right coronary artery aneurysm (diameter 18 mm) with a small old organized anterior shell of thrombosis and mildly dilated left main coronary artery but no aneurysm or thrombosis and normal function.

3. Discussion

The true incidence of KD in Middle Eastern countries is not known. The incidence was 7.4 per 100,000 children under 5 years of age in a retrospective study conducted in an eastern province of Saudi Arabia for the period from 1992 to 2012 [5,6].

KD is associated with increasing complications in infants. Many risk factors contribute to the increased risk of coronary aneurysm especially delayed IVIG administration, age <6 months, saccular shape of coronary aneurysm, and its giant size (>8 mm diameter) [1,7]. We reported an infant with KD who received IVIG after >2 weeks and already had a persistent giant saccular RCA aneurysm whereas LCA multiple fusiform aneurysms improved with time (see Fig. 3).

Burns et al. [11] reported that infliximab made 13/16 patients with IVIG-resistant KD defervescent and their laboratory measures improved within 48 hours [8] However, our patient was admitted after the acute stage had already ensued.

Warfarin is a good choice for complicated KD patients who need additional anticoagulation. However, this oral anticoagulant may be substituted with daily subcutaneous injections of enoxaparin in case of failure to maintain a safe therapeutic INR level [7–10].

The indications for revascularization surgery (coronary bypass graft procedures) in children have not been established in clinical trials, but such surgery should be considered when reversible ischemia is present on stress testing [7,8,10]. Our patient could not undergo stress testing as she was uncooperative.

4. Conclusion

Kawasaki disease is a debilitating disease which needs prompt and early action with regard to treatment with IVIG to prevent lifelong disability and complications [7,8,10]. A large scale epidemiologic data of KD however are not available for KSA. According to our knowledge this is the first giant coronary aneurysm clearly related to KD reported in KSA that persisted for 5 years fortunately with an uneventful course.

References

[1] Sei YS, Jin HO, Jong-Hyun K, Ji-Whan H, Kyung-Yil L, Dae KK. Giant coronary and axillary aneurysms in an infant with Kawasaki Disease associated with thrombocytopenia. J Korean Pediatr 2005;48:901–6.
[2] Zhang S, Liu G, Yu T, Zhou G, Zheng R. Giant right coronary artery aneurysm secondary to Kawasaki disease in child. Int J Clin Exp Pathol 2015;8:9468–70.
[3] Barron KS. Kawasaki disease: Etiology, pathogenesis, and treatment. Cleve Clin J Med 2002;69:SII69–78.
[4] Alharbi KM. Kawasaki disease in western Saudi Arabia. Saudi Med. J 2010;31:1217–20.
[5] Lardhi AA. Kawasaki disease: A university hospital experience. Saudi J Med Sci 2013;1:35–9.
[6] Rosenfeld EA, Corydon KE, Sculman ST. Kawasaki disease in infant less than one year of age. J Pediatr 1995;126:524–9.
[7] Shah R, Hirzallah H, Fares M, Mallad A, Karlsson G, El-Hamdani M, et al. J Med 2018;4.
[8] Eleftheriou D, Levin M, Shingadia D, Tulloh R, Klein NJ, Brogan PA. Management of Kawasaki disease. Arch Dis Child. 2014;99:74–83.
[9] El-Segaier M, Galal M. Intracoronary thrombus in an infant with Kawasaki disease and giant coronary aneurysm. Acta Paediatr 2013;102:e227–8.
[10] Newburger JW, Takahashi M, Gerber MA, Gewitz M, Tani LY, Burns JC, et al. Diagnosis, treatment, and long-term management of Kawasaki Disease. Circulation 2004;110:2747–71.
[11] Burns JC, Mason WH, Hauger SB, Janai H, Bastian JF, Wohrley JD, et al. Infliximab treatment for refractory Kawasaki syndrome. J Pediatr 2005;146:562–7.