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Case Report

Incomplete Kawasaki Disease in a 27 days old Infant – A Case Report

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Abstract

We report a case of incomplete Kawasaki disease in a 27 days old neonate. The neonate presented with fever, cough, cold, and poor feeding, accompanied by a polymorphous rash. Neurological manifestations, including encephalopathy and convulsions, were also observed. Laboratory findings showed thrombocytosis, anemia, leukocytosis, elevated alanine transaminase and C-reactive protein. Echocardiography confirmed dilated coronary arteries. The administration of intravenous gamma globulin led to a prompt improvement in the infant's condition. Although Kawasaki disease is uncommon in neonates, it can manifest as a rapidly progressing and severe illness.

Introduction

Kawasaki disease (KD) is an acute inflammatory disease that typically affects medium- and small-sized muscular arteries, particularly the coronary vessels. It primarily affects young children under the age of five years and is extremely rare in neonates.[1] Following the global spread of the COVID-19 pandemic, a new disease called multisystem inflammatory syndrome in children (MIS-C) has emerged. Interestingly, patients with KD and MIS-C can exhibit remarkably similar presentations, making the initial diagnosis and treatment decisions quite challenging. It is crucial for clinicians to have a comprehensive understanding of the variations in epidemiology, diagnostic criteria, organ involvement, and laboratory markers to effectively differentiate between these two. We present a case of incomplete KD in a 27 days old neonate

Case Report

A 27-day-old female neonate with no significant birth history had presented with fever, cough, cold and poor feeding for six days. A generalized polymorphous rash had appeared three days prior. At admission to NICU, the infant had depressed sensorium, poor spontaneous breathing effort and only responding to painful stimuli. She had fever, tachycardia and hypotension with deranged capillary refill time and 66% SpO_2. Subsequently, the baby was intubated and initiated on ionotropic support. The baby had seizures and exhibited signs of encephalopathy. The baby was given phenobarbitone for seizures. Chest X-ray revealed findings consistent with viral pneumonia. IV antibiotics Meropenem and Netilmicin were initiated in view of sepsis. Routine hematological investigations were within normal limits. CSF study revealed 5 cells, all lymphocytes, sugar 76 mg / dl and protein 56 mg / dl. Respiratory virus panel test (multiplex PCR) collected from nasopharyngeal swab - negative, SARS-CoV-2 RT-PCR – negative, SARS CoV-2 nucleocapsid antibody (AU/ml) IgM & IgG - positive. Bacterial cultures from blood, urine, and cerebrospinal fluid were sterile. Partial orogastric feeding was started four days later. After six days, the neonate was successfully extubated and IV antibiotics stopped after 10 days. After 14 days of being admitted, the infants' condition again deteriorated, marked by repeated episodes of apnea and active convulsions. Consequently, the infant

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had to be reintubated and was administered levetiracetam to control the convulsions. This time, there was raised leucocytes (25000 / mm^3), platelets (0.75 million / mm^3) and CRP (23.8 mg / L). There was also raised serum ferritin – 1146 mcg / L, procalcitonin 24 ng / ml and serum IL 6 - 416 pg / ml. Serum albumin was normal, while ALT was elevated (173 U / L). The coagulation profile was deranged (PT-29 seconds and INR: 2.3). Serum D-dimer was 4.59 ng / ml FEU. Cardiomegaly was observed on the chest X-ray (Figure 1). Echocardiography showed a dilated left coronary artery (Z-scores for left main coronary artery 4.62, left anterior descending artery 3.5, Left circumflex artery-2.8, right coronary artery < 2.0) (Figure 2). Electrocardiogram was normal.

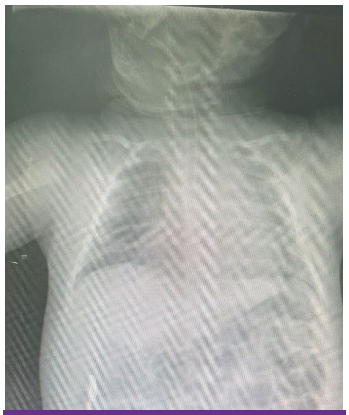

Fig 1 : Chest-X-ray showing cardiomegaly

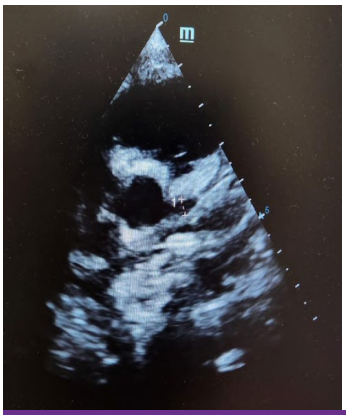

Fig 2: Echocardiography - parasternal short axis view showing left main coronary artery (LMCA) dilatation (Arrow mark red)

The infant received intravenous gamma globulin (IVIG) at a dosage of 2 g / kg. Within 48 hours of administration, there was a noticeable improvement in the infant's condition. The baby was extubated after four days. High-dose aspirin (30 mg / kg) was initiated and followed by a maintenance dose. The infant was discharged after 25 days. Upon a follow-up echocardiography at six weeks, the aneurysm was found to persist. The neurological examination conducted at the time of discharge was normal. The visual acuity was measured at 2.20 c / d at a distance of 38 cm with TAC (70% reliability). The Bruckner Test showed good and equal glow, and the screening BERA (Brainstem Evoked Response Audiometry) indicated both ears passed the test.

Discussion

KD primarily affects young children, with approximately 77% of affected patients being under the age of five years. Approximately 10% of cases of KD are observed in infants younger than six months of age.[2] In Japan, the occurrence of KD in infants below the age of three months is reported to be as low as 1.67%.[3] The youngest patients are more prone to displaying atypical symptoms and are at a higher risk of developing aneurysms.[4] Timely diagnosis and prompt administration of IVIG are crucial to minimize the occurrence of cardiac complications.

In our case, the neonate had fever, polymorphous rash, thrombocytosis, elevated ALT, and CRP, along with coronary artery dilatation which served as important indicators for making a diagnosis. Notably, the baby did not exhibit cervical lymphadenopathy, changes in extremities, or conjunctival injection. The infants also had encephalopathy, raised inflammatory markers and positive SARS CoV-2 nucleocapsid antibody.

According to a study conducted by Yoon et al, which examined 26 infants below the age of six months with KD, it was observed that infants in this age group rarely presented with cervical lymphadenopathy or non-exudative conjunctival injection compared to KD patients aged 6 months or older.[5] While uncommon, a small portion of patients with KD may experience neurological complications. Although encephalopathy is less commonly reported, there have been documented cases of its occurrence.[6] Neurological symptoms associated with KD include sensorineural hearing loss, facial nerve palsy, severe irritability, and aseptic meningitis.[6,7] Following the COVID-19 pandemic, a new disease called multisystem inflammatory syndrome in children (MIS-C) has emerged. Both KD and MIS-C are characterized by auto-immune and hyper-inflammatory responses that affect multiple organ systems.[7,8]

The most commonly observed CVS complication in MIS-C is ventricular dysfunction, although coronary artery dilation or aneurysms and pericardial effusions can also occur.[9] During the acute phase of MIS-C, elevated levels of CRP and ESR are typically observed, along with leukopenia instead of leukocytosis. Similar to KD, platelet activation occurs, but bone marrow suppression leads to thrombocytopenia. Studies have shown that a significant proportion of patients with MIS-C may meet the diagnostic criteria for either KD or atypical KD.[10] As a result, differentiating between these two diseases can be challenging. Therefore, it is crucial to have a comprehensive understanding of the subtle variations in clinical presentation, organ involvement, and laboratory markers in order to effectively distinguish between these conditions.

Conclusion

Diagnosing neonatal KD poses a clinical challenge due to the potential for atypical presentations. It is essential to maintain a high index of suspicion for KD in febrile neonates who do not respond to antibiotics. Timely echocardiography examination and early intervention are essential to prevent cardiovascular complications.

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