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Case Report

Kawasaki Disease in Disguise: Case Reports

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Abstract

Kawasaki disease is a vasculitis syndrome that presents with prolonged fever and mucocutaneous manifestations. Some cases have unusual presentations leading to delayed diagnosis and intervention. We report on two cases with atypical presentations.

Keywords: Atypical Kawasaki Disease; Incomplete Kawasaki Disease; Coronary Artery Aneurysm; Unresolving Pneumonia; Vasculitis

Abbreviations

KD: Kawasaki Disease; CAA: Coronary Artery Aneurysm; CRP: C-Reactive Protein; ESR: Erythrocyte Sedimentation Rate; IVIG: Intravenous Immunoglobulins

Introduction

Kawasaki Disease (KD) is among the common pediatric vasculitides [1]. It is a self-limited disease characterized by fever and acute inflammatory manifestations affecting the skin, mucous membranes and occasionally other organs [2]. Coronary artery aneurysms (CAA) are the most significant complication of KD leading to significant morbidity and mortality. Early diagnosis and intervention have a major role in reducing such risks. The typical diagnostic criteria of KD established by Tomisaku Kawasaki in 1967 [3] are not always fulfilled in a significant percentage of patients. Diagnosis depends on a combination of clinical and laboratory criteria none of which is solely reliable for making the diagnosis. Other more common diagnoses usually take precedence in the differential diagnosis adding to the challenge of diagnosing atypical or incomplete KD cases.

Case I:

A four-year old girl of South Eastern Asian descent with a history of hyperreactive airway disease and asthma presented to the ER department with high fever, and shortness of breath. Examination was notable for grunting, intercostal and subcostal retractions, generalized chest wheezing and diminished air entry bilaterally. Her oxygen saturation was 88%. She improved on oxygen and salbutamol nebulization. Blood count was notable for leucopenia but otherwise unremarkable. C-reactive protein (CRP) level was normal at 1.47. Chest radiography was significant for bilateral peribranchial cuffing with lower lung zone patches of consolidation. Presenting during the Flu season, the patient was admitted with an initial diagnosis of viral (probably influenza) pneumonia with asthma exacerbation. She was started on Oseltamivir, and Azithromycin together with salbutamol nebulization and supportive oxygen therapy as needed. Systemic steroids were administered due to poor response to bronchodilator therapy. On the next day of admission, fever started to subside, but the patient still needed respiratory support. Two days later, fever started to rise again reaching up to 39.5˚C with persistent respiratory distress. Repeat chest X-ray showed segmental infiltration of right upper lung zone, haziness of right lower lung base, increased left retrocardiac density and blunting of right costophrenic angle. Augmentin was added to the treatment due to progression of the pneumonia. However, fever and respiratory distressed persisted. Blood work was repeated, and it showed moderate leukocytosis at 13000, slightly lower hemoglobin at 10.9, platelet count of 600,000. C-reactive protein was elevated at 41.57. The patient started to develop glazed erythematous tongue. This, together

with the resolving pneumonia and elevated platelet count raised the possibility of KD. We reviewed the literature for similar cases and started to do further investigations to establish the diagnosis. Erythrocyte Sedimentation rate (ESR) was found elevated at 120, serum albumin slightly low at 31.8 and +1 urine leucocytes. Echocardiography showed mild dilatation of left coronary artery further confirming the diagnosis at day 8 of the fever. Patient received one dose of intravenous immunoglobulins (IVIG) at 2g/kg and Acetyl Salicylic acid at 75 mg/kg. Fever subsided for one day then recurred less than 48 hours after completion of the IVIG dose. Another dose of IVIG was administered after which fever completely subsided with marked improvement in the general condition and resolution of the respiratory distress. Echocardiographic follow-up showed complete recovery of the coronary aneurysm 6-months after the presentation. This patient is now at 12-years of age and is still with good health apart from occasional asthma exacerbations and mild allergic rhinitis.

Case II:

A 6-month old male patient of Chinese descent with past history of cow-milk protein allergy and NICU admission for prematurity and respiratory distress presented to the pediatric ER with fever, severe diarrhea and irritability for 3 days. He was moderately dehydrated and inconsolable. Initial blood count and inflammatory markers were unremarkable. While in ER, he developed vomiting and was admitted for further intravenous rehydration and further evaluation. The initial diagnosis was viral gastroenteritis, and intussusception was suspected in view of vomiting and irritability. Radiological examination ruled out intussusception, but symptoms persisted. There was marked abdominal distension. Nasogastric tube was inserted to relieve the distension. Bilious fluid started to appear from the nasogastric aspirate. Abdominal plain X ray showed suspected obstruction, so laparotomy was done. Exploration showed no bowel obstruction. The appendix was removed, and a sample of inflamed lymph node was taken. Histopathological examination showed neutrophilic infiltration confined to appendicular mucosa and non-specific reactive inflammation of the mesenteric lymph nodes. Fever and irritability persisted despite proper hydration and supportive treatment. Ultrasound done to rule out intussusception showed moderate hepatomegaly. Liver function testing showed elevated hepatic enzymes and low albumin of 2.2. Repeat blood count showed marked leukocytosis of 35,000 with low hemoglobin of 8.2 g/dl and platelet count of 216,000. The low albumin level considering the fever, irritability, and the infant's ethnic background raised the possibility of KD. Further investigations showed an ESR of 140 and CRP of 138. Platelet count started to rise to around 1,500,000. Echocardiography showed moderately dilated right coronary artery. Diagnosis of KD was established about 10 days of the start of fever. The patient responded well to one dose of IVIG and high dose Acetyl Salicylic acid. At seven years of age, the patient has no residual coronary artery dilatation and is faring very well.

Discussion

We report on two cases of atypical KD. The first case presented with a picture consistent with viral pneumonia with poor response to treatment. Radiographic pulmonary abnormalities mainly peribronchial cuffing and pleural effusion were reported in about 15% of KD cases. [4] However, Singh and colleagues reported that less than 2% of confirmed KD cases had a predominant pulmonary presentation. [5] Peribronchial cuffing is usually seen in viral pneumonia and hyperreactive airway disease. Our patient was a known asthmatic patient and the initial picture matched the more common and reasonable diagnosis of asthma exacerbation triggered by viral infection. KD is predominantly a vasulitis disease that leads to increased vascular permeability and inflammatory exudate [6]. The pulmonary changes seen in our patient most probably reflect such changes rather than infective pneumonia. The case report published by Uziel and colleagues in 2003 of unresolving pneumonia as the main manifestation of atypical KD helped us in diagnosing our case [7].

The second case complained of fever, diarrhea, irritability and vomiting. Gastroenteritis possibly complicated with intussusception was the most probable diagnosis. Ultrasonography and ruled out intussusception but plain X-ray of the abdomen showed possible obstruction. Laparotomy was done due explore the possible cause of the obstruction. Diarrhea, irritability and vomiting occur in 40-60% of KD patients [2,8]. however, these are non-specific symptoms caused by much more common diseases. Gall bladder hydrops and hepatitis have been reported in cased with KD [9]. A recent publication review reported 48 case reports of KD patients showing pseudo intestinal obstruction. Half of these reported cases underwent surgery [10].

Conclusion

Diagnosis of KD can be very challenging especially in atypical and incomplete cases. Atypical presentations usually mimic many common diseases that take priority in the differential diagnosis.

The typical mucocutaneous changes of KD can be absent at the time of presentation. Laboratory changes suggestive of KD appear in later stages and sometimes after more than one week of the onset of symptoms. Early diagnosis and intervention within 10 days of disease onset markedly reduce morbidity and mortality related to CAA [11,12]. A high level of clinical suspicion and awareness of the different atypical presentations of KD is needed to achieve this goal. We managed to diagnose both cases within this favorable time frame. This may have contributed to their complete uneventful recovery.

Conflict of Interest

There is no financial benefit or conflict of interest to be reported for this case report.

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