Kawasaki Disease with Profound Vascular Involvements: An Insightful Pediatric Case

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1 INTRODUCTION

Kawasaki Disease (KD) is a rare entity but one of the most common pediatric vasculitis. It predominantly affects the pediatric population, generally before the age of 5 years with coronary artery aneurysms being the most feared complication; however, due to the broader disease spectrum, patients can present with diverse clinical presentations [1]. Coronary aneurysms generally develop after two weeks of disease onset in about 25% of cases [2]. The standard treatment includes IV immunoglobulin and aspirin [3]. There are no specific laboratory markers and the diagnosis is mainly based on a set of clinical criteria, featuring prolonged fever, polymorphous rash, conjunctivitis, mucosal changes, lymphadenopathy, and extremity changes [4]. Systemic arterial aneurysm formation is a very rare entity, affecting 0.8% to 2% of the cases, approximately 3 months after disease onset [5].

2 CASE HISTORY/EXAMINATION

We present a rare entity of extensive Kawasaki Disease (KD) in a male infant aged 2 months and 21 days, who presented with tachycardia and tachypnea with a prolonged fever lasting 25 days, and a rash that appeared 17 days before admission. Key findings on physical examination included tachycardia, a 2/6 systolic murmur, and pronounced tachypnea without any added sounds and equal air entry in the chest bilaterally.

3 METHODS (DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS, AND TREATMENT)

Initial lab workup was remarkable for raised CRP, ESR, and TROP-I levels. The patient was shifted with high-flow oxygen to the intensive care unit after the initial care in the emergency department. Based on initial echocardiography findings in ER, which had demonstrated dilatation of coronary arteries, Focused Echocardiography was subsequently performed by an expert cardiologist, which revealed a severely dilated right coronary artery at its origin with aneurysm formation & distal ectasia, severely dilated left coronary artery, severe ectasia of the left anterior descending artery, and severely dilated left circumflex artery. Heart failure therapy including diuretics was optimized and intravenous immunoglobulin (2g/kg), and dexamethasone were included in the treatment plan after consulting the cardiology team. Intensive monitoring of vitals, cardiac electrolytes, and daily 12-lead ECG were performed. The direct breastfeeding trial was tolerable, which was continued.

As he remained stable on oral breastfeeding, he was stepped down to special care. High-flow oxygen therapy tapered gradually. While in special care, the patient developed watery diarrhea and spikes of fever. Stool cultures, detailed report and blood cultures were sent, which revealed multidrug-resistant pseudomonas.

Antibiotic therapy was optimized with meropenem and colistin, which improved the condition within days. Diagnostic assessment through CT angiography was followed, revealing aneurysmal dilation in both internal carotid arteries, tortuous aorta with aneurysmal dilatation at the origin of bilateral intercostal arteries, coeliac, superior mesenteric, and both renal arteries, as well as aneurysmal dilatation of all coronary arteries, as shown in figures 1-4.

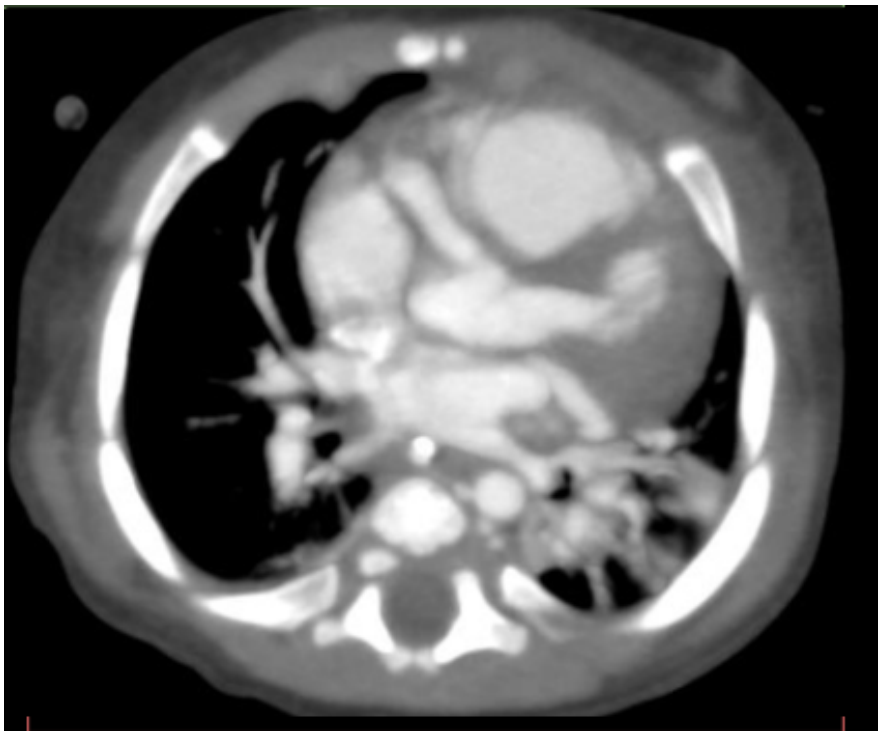
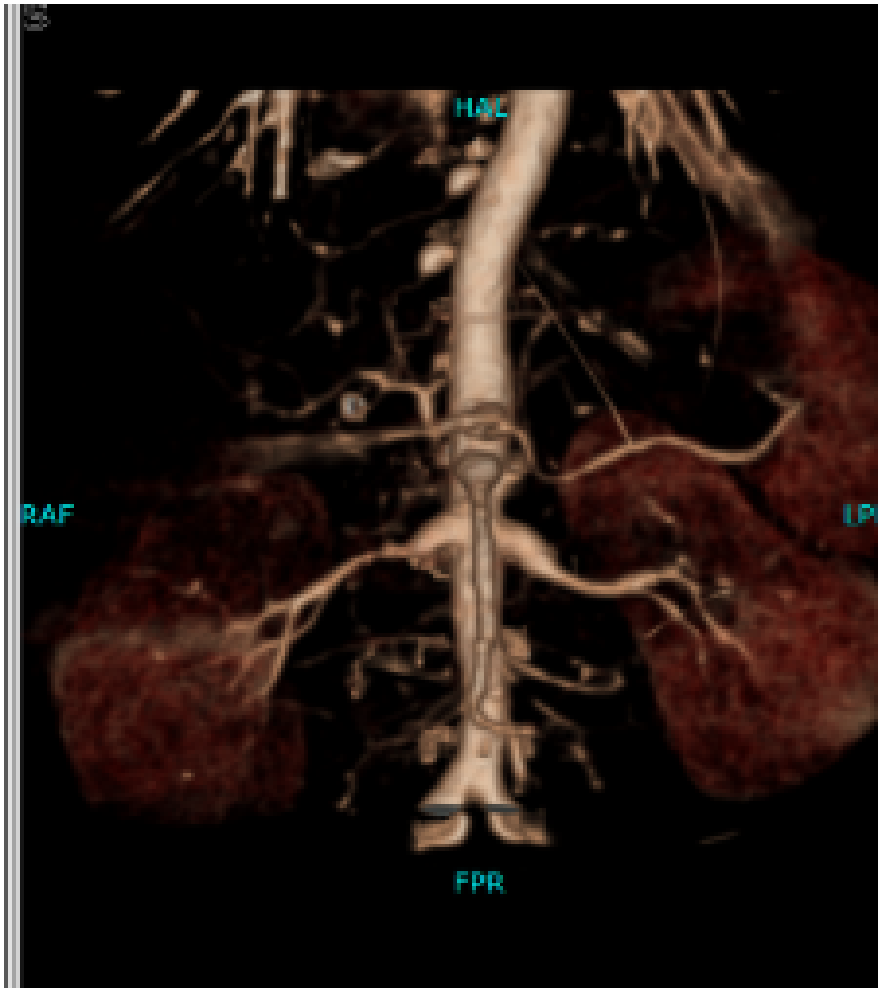
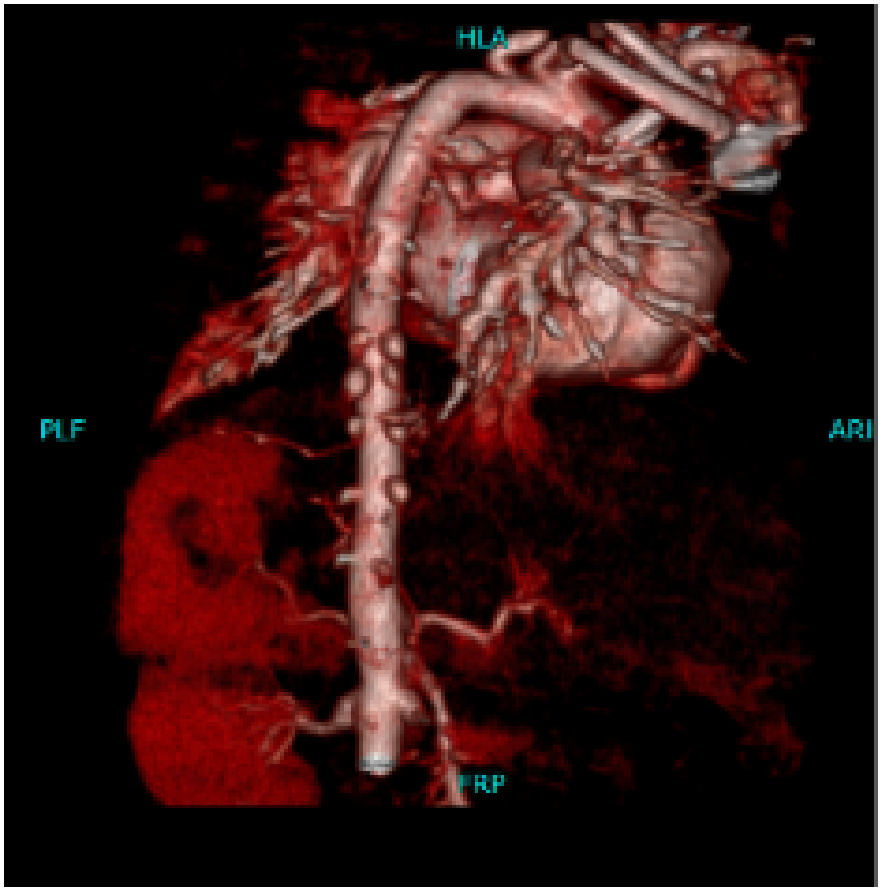


Figure 1: *Axial CECT Coronary angiogram demonstrating multiple bilateral coronary artery aneurysmal dilatations.*





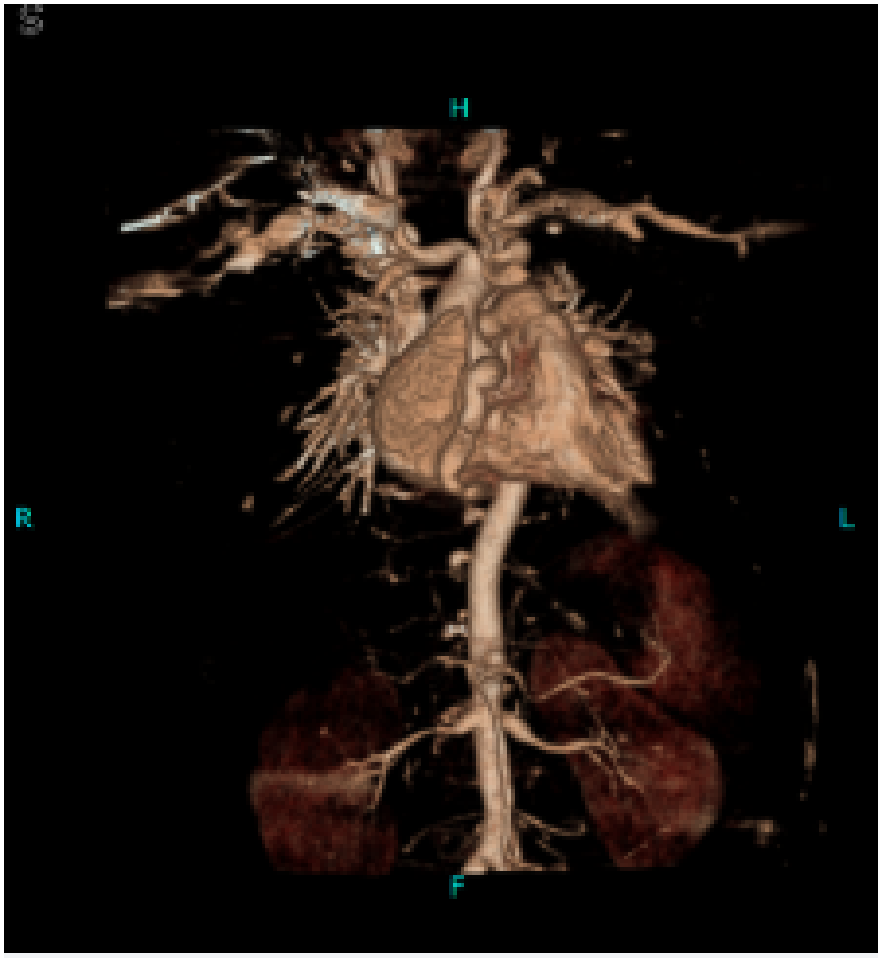


Figure 2, 3 & 4 *Maximum intensity projection (MIP) with coronal reconstruction and virtual cinematic rendering images, demonstrating multiple systemic arterial aneurysms in this patient, including coronary, coeliac, superior mesenteric, intercostal, and bilateral renal arteries.*

4 CONCLUSION AND RESULTS (OUTCOME AND FOLLOW UP)

An autoimmune profile was sent and the rheumatology team was taken on board. While staying in special care, a second dose of IVIG, naproxen, and immunosuppressive therapy including azathioprine and hydroxychloroquine were started. The patient remained stable and was shifted to the ward. Repeated cultures did not reveal MDR pseudomonas. The baby was later discharged after a total hospital stay of 11 days and his family was counseled for early follow-up with infectious disease, cardiology, and hematology clinic outpatient.

5 DISCUSSION

Although KD is a self-limiting disease, the main feared complication of developing coronary aneurysms can occur in up to 30% of untreated cases [1]. This complication is one of the major causes of mortality in untreated cases, necessitating timely management. The incidence of this complication can be reduced with effective & prompt IVIG administration [3]. Initial presentation can be variable, posing significant challenges for diagnosis, as there are no laboratory diagnostic markers and diagnosis relies mainly on classical physical signs [4]. However, in our described case, the variability in the presentation of KD and the potential for extensive vascular spectrum, beyond the coronary arteries is key to consider. The aneurysmal dilatation in

multiple arterial sites, including the bilateral internal carotid arteries, descending aorta, intercostal arteries, coeliac artery, superior mesenteric artery, bilateral renal arteries, and all coronary arteries, is a rare presentation, occurring in less than 2% of cases [5]. This needs a systematic clinical as well as imaging approach while evaluating such patients to look for multi-organ involvement, as the disease has the potential to show variable complexities.

While reviewing the computed tomographic study as a radiologist, one should be aware of this rare entity to look for in their checklist. This also raises a special concern for the team of infectious diseases, as our case had an added complexity in management due to MDR Pseudomonas sepsis. The successful management of this case involved a multidisciplinary approach, highlighting the importance of collaboration among various specialties in managing complex KD cases. IVIG, aspirin, and IV dexamethasone are the key treatments recommended for managing acute KD [3]. However, the extensive vascular involvement in this case required vigilant monitoring and a tailored approach to address the unique challenges posed by the multiple aneurysms.

This case emphasizes acute as well as long-term prospective plans for management. It emphasizes the need for long-term follow-up and monitoring of patients with KD, especially those with extensive vascular involvement, to detect and manage potential complications such as aneurysm progression or thrombosis and multi-organ involvement [5]. Previously, there have been case reports of young patients presenting with calcified coronary aneurysms, highlighting the importance of screening patients for the progression of the disease [6]. This also highlights the importance of family education regarding disease progression, prognosis, and regular follow-ups. The limitation of our case is the lack of a long-term follow-up date, which would provide valuable insights regarding the progression of other possible vascular involvement and multi-organ complications. Further research is needed in these patients with extensive vascular involvement, which would explain and redefine the standards of approach towards such patients in the long run.

AUTHOR CONTRIBUTIONS

Naila Nadeem: Conceptualization; data curation; investigation; project administration, writing – original draft; writing – review & editing. **Muhammad Nadeem Ahmad:** Conceptualization; Writing – original draft; writing – review & editing. **Muhammad Haseeb Malik:** Conceptualization; data curation; supervision; validation.**Mallick Muhammad Zohaibuddin:** Conceptualization; data curation; supervision. **Muhammad Ahmed:** Conceptualization; data curation; validation. **Faheemullah Khan:** Conceptualization; supervision; validation. **Hatem Eltaly:** Conceptualization; data curation; investigation; validation; writing – original draft. **Uffan Zafar** : Conceptualization; data curation; investigation; project administration, writing – original draft; writing – review & editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

ETHICS STATEMENT

Not applicable.

DATA AVAILABILITY STATEMENT

The data supporting the findings of this study are available from the corresponding author upon reasonable request.

CONSENT

Written informed consent was obtained from the patient's parents for the publication of this case report and any accompanying images.

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