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ATYPICAL KAWASAKI DISEASE IN AN INFANT: A CASE STUDY OF DIAGNOSTIC CHALLENGES AND DISCHARGE AGAINST MEDICAL ADVICE

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ABSTRACT

Objective: This case study evaluates the diagnostic difficulties and socio-medical challenges encountered during the sequential hospitalization of an 11-month-old infant with atypical Kawasaki disease (AKD).

Methods: A retrospective review of medical records, laboratory values, imaging studies, and documented clinical interactions was conducted at a regional hospital.

Results: The patient initially presented with isolated acute febrile cervical lymphadenopathy. A recent routine Varicella immunization was noted in the medical history. The clinical trajectory was confounded by early corticosteroid exposure for airway stridor and a congenital bicuspid aortic valve with an associated dilated ascending aorta, leading to the initial exclusion of Kawasaki disease. Upon readmission with delayed periungual desquamation and borderline left anterior descending coronary artery Z-scores, a diagnosis of incomplete vasculitis was established, and intravenous immunoglobulin (IVIG) therapy (total 1.65 g/kg) was completed. However, during both admissions, the patient's parents demanded immediate discharge against medical advice (DAMA) upon observing temporary symptomatic relief, equating clinical improvement with full recovery.

Conclusion: Incomplete clinical criteria and co-existing structural cardiac anomalies can cause significant diagnostic delays in infant vasculitis. Furthermore, this case suggests that parental interpretation of early symptomatic improvement may contribute to DAMA decisions, highlighting the need for highly specific risk communication regarding invisible cardiovascular complications.

KEYWORDS

Atypical Kawasaki Disease, Pediatric Vasculitis, Discharge Against Medical Advice (DAMA), Health Communication, Patient Compliance

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Introduction

The clinical management of rare and atypical pediatric conditions poses a profound challenge not only to medical science but also to the structures of healthcare communication and parental decision-making frameworks. Kawasaki disease (KD) is an acute, self-limiting systemic vasculitis that predominantly affects infants and young children, carrying a significant risk of coronary artery aneurysms if left untreated. While classic KD manifests with a well-defined constellation of signs—including prolonged fever, bilateral conjunctival injection, mucosal changes, polymorphous rash, extremity changes, and cervical lymphadenopathy—incomplete or atypical variants frequently lack these hallmark features. Atypical Kawasaki disease (AKD) often presents with isolated symptoms, mimicking common bacterial or viral infections, which leads to frequent misdiagnoses in outpatient settings and primary care networks. As emphasized in recent epidemiological syntheses, the clinical presentation of AKD can be misleading, particularly in young infants where incomplete criteria significantly delay the initiation of intravenous immunoglobulin (IVIG) therapy, thereby increasing the baseline risk of cardiac sequelae (Conte et al., 2023).

The complexity of AKD is further compounded by external clinical variables, such as recent immunizations or coincidental structural cardiac anomalies, which can cloud the diagnostic picture. From a socio-medical perspective, the diagnostic odyssey of an infant with AKD highlights critical friction points in healthcare communication. Misunderstandings regarding the severity of systemic inflammation, coupled with parental misalignment with clinical protocols, can result in families seeking discharge against medical advice (DAMA). DAMA remains a problematic phenomenon in pediatric care, heavily influenced by communication failures, sociocultural factors, and gaps in parental health literacy regarding subclinical risks (Al-Mohammadi, 2019). This case study evaluates the diagnostic challenges and communication breakdowns encountered during the dual hospitalization of an 11-month-old infant presenting with atypical Kawasaki disease.

Methodology

This paper utilizes a descriptive case study design to analyze a complex clinical scenario at the intersection of pediatric medicine and healthcare communication systems. A retrospective review of medical records, laboratory findings, imaging studies, and documented clinical interactions was conducted. The primary data source consisted of the patient's longitudinal medical records and diagnostic archives across two distinct, consecutive admissions at a regional hospital. To preserve patient anonymity and comply with international reporting standards, timeline events are described relative to the day of initial presentation. Clinical parameters were benchmarked against established international diagnostic protocols, specifically the American Heart Association (AHA) scientific statement on Kawasaki disease guidelines.

Results**Initial Presentation and Outpatient Course**

An 11-month-old male infant (birth weight: 3650 g, born via normal spontaneous vaginal delivery at 39 weeks of gestation, Apgar score: 10) was admitted to the pediatric ward of a regional hospital on Day 1 of the clinical timeline. The chief complaints upon presentation were a persistent fever lasting for seven days (onset on Day -6, maximum temperature 38.2°C, recurring initially every 6 hours) and a sudden right-sided cervical swelling. The child's routine immunization schedule was up-to-date, including the administration of the first dose of the Varicella vaccine (Varilrix) on Day -7, exactly two days prior to the onset of the febrile episodes.

In the days preceding admission, the patient had been evaluated twice in outpatient settings. He was initially diagnosed with viral pharyngitis and prescribed symptomatic treatment (ibuprofen and paracetamol). Despite regular antipyretic therapy, the fever persisted. A rapid antigen test for Group A Streptococcus performed on Day -1 was negative. The parents reported progressive hyporexia, a sharp decline in fluid intake, and decreased diuresis characterized by concentrated, orange-colored urine. On the morning of admission, a firm, non-fluctuant swelling appeared on the right side of the infant's neck.

First Hospitalization (Day 1 – Day 6)

Upon physical examination at admission, the infant was in a moderate general status, exhibiting visible signs of dehydration, a hoarse voice, a notable inspiratory stridor, and sinus tachycardia (140 bpm). Local examination revealed an erythematous pharynx with hypertrophied palatine tonsils without exudates. A firm, fixed nodular mass measuring 5 x 5 cm was palpable at the right mandibular angle; the overlying skin was normal. There were no mucosal fissures, conjunctival injections, skin rashes, or peripheral edema.

Initial laboratory investigations revealed an intense systemic inflammatory response:

- C-reactive protein (CRP) was markedly elevated at 172 mg/L; Ferritin was 410 ng/mL. Procalcitonin levels remained low.

- Leukocytosis (31,000/ μ L) with prominent neutrophilia and absolute lymphopenia was noted.

- Severe thrombocytosis (942,000/ μ L) and decreased red blood cell indices (microcytic anemia) were recorded. Erythrocyte sedimentation rate (ESR) was elevated.

- Random hyperglycemia was detected. Serum troponin levels were normal, while NT-proBNP was mildly elevated. Organ functions (renal and hepatic) and baseline coagulation profiles were within normal physiological limits.

- Serological assays for Epstein-Barr Virus (EBV) and *Mycoplasma pneumoniae* were negative. Stool culture returned normal flora; a *Campylobacter* assay was negative. Repeated blood and urine cultures remained sterile, effectively ruling out a conventional bacterial urinary tract infection or sepsis.

Intravenous empiric antibiotic therapy with a third-generation cephalosporin (Cefotaxime) was initiated. Due to the severity of the inspiratory stridor and secondary upper airway compromise, a single dose of intravenous Dexamethasone was administered alongside nebulized bronchodilators, achieving airway stabilization. By the second day of hospitalization (Day 2), the cervical mass resolved completely; a follow-up ultrasound of the neck, an abdominal ultrasound, and a chest X-ray revealed no residual pathological deviations.

A specialized pediatric cardiology consultation was obtained on Day 2. Electrocardiography (ECG) demonstrated respiratory sinus arrhythmia but was otherwise unremarkable. Echocardiography (ECHO) revealed an incidental structural anomaly: a bicuspid aortic valve (BAV) without secondary stenosis or regurgitation, accompanied by a mild dilation of the ascending aorta. The origins of the coronary arteries were anatomically normal, though their absolute dimensions were noted to be at the upper borderline threshold (Z-scores). A trace pericardial effusion was present, with preserved biventricular systolic function and no signs of pulmonary hypertension.

Due to the infant's rapid clinical defervescence following antibiotic initiation, normal coronary calibers, and the absence of cutaneous-mucosal criteria, Kawasaki disease was initially excluded. The reactive thrombocytosis persisted, peaking at 1,081,000/ μ L. A hematology consultation recommended implementing antiplatelet therapy (Acetylsalicylic Acid) only if platelet counts exceeded 1,500,000/ μ L.

On Day 6, noting that the child was afebrile, active, and feeding well, the parents concluded that the minor clinical improvements meant the child was fully cured. Consequently, the parents demanded immediate discharge against medical advice (DAMA) before full diagnostic monitoring could be completed. The patient was discharged with a prescription for oral Cefixime (Cetix).

Second Hospitalization (Day 9 – Day 11)

On Day 9, three days post-discharge, the infant was brought back to the hospital. The parents reported that the previous day (Day 8) they had observed an extensive, fine periungual desquamation (peeling of the skin) affecting the fingertips of both hands. The infant had remained afebrile at home but exhibited a mild residual hoarseness and a sporadic cough.

Upon readmission, the patient was afebrile (37.3°C), responsive, and in a stable general condition. Cutaneous and mucosal examinations were otherwise negative for new eruptions, and the cervical lymphadenopathy had completely regressed. Cardiorespiratory auscultation confirmed a stable, soft systolic murmur (grade 2/6) at the cardiac base, and respiratory sinus arrhythmia (100 bpm).

Repeat laboratory assessments showed a significant drop in acute phase reactants (CRP 2.9 mg/L, low PCT), but a persistently elevated ESR (50 mm/h). Leukocytosis had normalized (12,000/ μ L), but severe thrombocytosis remained sustained at 1,048,000/ μ L. Transient mild electrolyte anomalies (hyperkalemia and hyponatremia) and persistently high ferritin were documented.

An urgent follow-up echocardiogram was performed on Day 9. While the structural BAV and dilated ascending aorta remained unchanged, the pediatric cardiologist identified borderline Z-score expansions

localized specifically to the left anterior descending (LAD) coronary artery. When synthesized with the history of a 7-day unexplained fever, delayed fingertip desquamation, and persistent thrombocytosis, the clinical picture met the criteria for Incomplete/Atypical Kawasaki Disease (AKD).

The standard AKD therapeutic protocol was initiated immediately. The infant received a continuous 10-hour intravenous immunoglobulin (IVIG) infusion at an initial fractional dose of 1.1 g/kg, alongside antiplatelet therapy (Aspirin/Acard), gastric protection (Omeprazole/Helicid), and continuous pulse oximetry monitoring. A second fractional IVIG dose (0.55 g/kg) was completed on the evening of Day 9, bringing the total accumulated dose to 1.65 g/kg, without immediate adverse events.

On the morning of Day 10, approximately seven hours post-infusion, the infant experienced a single episode of emesis, mild lethargy, and a low-grade temperature spike (37.5°C), which resolved following a single dose of ibuprofen. No signs of myocardial failure or localized edema developed. By Day 11, the patient had recovered fully, displaying normal physical activity and full oral intake, with follow-up hematology demonstrating a clear downward trend in the platelet count.

Despite detailed counseling regarding the critical risk of coronary artery aneurysm development and the absolute necessity of post-infusion cardiac monitoring, the parents intervened a second time, demanding a premature discharge against medical advice (DAMA) on Day 11, exactly 36 hours after IVIG completion, as they believed that the child's stable clinical baseline was sufficient to warrant home management. The family was discharged with mandatory instructions for urgent outpatient pediatric cardiology follow-up and tracking of the coronary Z-scores.

Discussion

This case study illustrates the complex diagnostic and communicative labyrinth that often surrounds incomplete or atypical Kawasaki disease in infants. The initial presentation of isolated febrile cervical lymphadenopathy—frequently termed "Kawasaki disease mimicking cervical lymphadenitis"—occurs in approximately 10% to 15% of KD cases, often resulting in primary misdiagnoses as bacterial infections or adenitis. The diagnostic challenge was further complicated by the timeline of the Varicella vaccination administered 48 hours prior to the fever, which initially led clinicians to suspect a normal post-vaccinal reaction or a common viral exanthem. As documented by Matsubara et al. (2019), identifying AKD becomes an intricate diagnostic puzzle when typical mucocutaneous symptoms are completely missing or appear only as late, isolated phenomena during the convalescent phase. Infants under one year of age represent the highest risk group for developing coronary abnormalities precisely because their clinical manifestations are poorly aligned with classic diagnostic criteria.

The administration of Dexamethasone for upper airway stridor represents a critical pivot point in this case. Recent evidence indicates that inappropriate or early corticosteroid exposure prior to definitive IVIG therapy can mask systemic inflammatory markers, transiently lower CRP, and suppress fever, thereby delaying the correct diagnosis of AKD (Chen et al., 2024). This mechanism may have contributed to the diagnostic delay observed in the present case during the first hospitalization, contributing to the premature exclusion of KD based on the rapid clearance of the cervical mass and normal early coronary dimensions.

The detection of a bicuspid aortic valve (BAV) and an associated dilated ascending aorta introduced an additional structural confounding factor. Dilation of the aortic root and ascending aorta is a well-known structural complication in pediatric patients with BAV (Warren et al., 2006). However, transient aortic root dilation can also occur acutely during the intense vasculitic phase of Kawasaki disease. Distinguishing whether the ascending aorta's dimensions were purely a reflection of the congenital BAV or an active manifestation of KD-induced vasculitis underscores the diagnostic conundrum noted by rheumatologists (Manlhiot et al., 2012).

An additional management-related consideration concerns the administration of a cumulative IVIG dose of 1.65 g/kg, which is lower than the standard 2.0 g/kg regimen recommended by the American Heart Association (McCrindle et al., 2017). Although the patient demonstrated short-term clinical improvement, deviation from guideline-based dosing may complicate the interpretation of treatment efficacy and long-term coronary outcomes. This suboptimal dosing was entirely due to the early discontinuation of care by the family, which prevented the administration of additional fractional dosing.

One possible contributing factor to the repeated DAMA events may have been limitations in risk communication and parental understanding of delayed cardiovascular complications. The phenomenon of Discharge Against Medical Advice (DAMA) in pediatrics represents a complex sociological problem. As analyzed by Al-Mohammadi (2019), parental initiation of DAMA is rarely a random event; it is fundamentally driven by critical factors in communication, suboptimal health literacy, and a perceived resolution of danger

once immediate symptoms diminish. In this instance, the parents mistook the infant's temporary defervescence and symptomatic relief during both admissions as a sign of complete recovery. They lacked the medical literacy required to understand that while the external symptoms (fever, neck swelling) had abated, the internal systemic vasculitis was still actively evolving, as evidenced by the delayed periungual desquamation and emerging borderline coronary artery Z-scores. Al-Mohammadi (2019) highlights that to mitigate DAMA, physicians must move away from brief, abstract clinical updates and instead use structured, empathetic communication frameworks that explicitly detail the severe invisible risks—such as silent coronary aneurysms—associated with premature termination of hospital monitoring. This case strongly reinforces that counseling must explicitly address the difference between absolute biological resolution and superficial symptomatic improvement to prevent high-risk premature discharges.

Beyond the clinical diagnosis itself, this case illustrates how healthcare outcomes may be affected by communication processes, parental risk perception, and decision-making behavior. These socio-medical factors can substantially influence adherence to evidence-based treatment recommendations, particularly when disease progression is not directly observable by caregivers.

This report has several limitations. It describes a single patient, causal relationships between corticosteroid exposure and diagnostic delay cannot be definitively established, and the precise socio-cognitive reasons underlying parental DAMA decisions could not be formally assessed through structured interviews.

Conclusions

- Atypical Kawasaki disease can mimic bacterial cervical lymphadenitis and delay diagnosis.
- Confounding factors such as early corticosteroid exposure and congenital cardiac anomalies may complicate clinical assessment.
- This case suggests that effective clinician-parent communication may improve adherence to recommended monitoring and treatment plans in atypical pediatric vasculitis.

Ethical Considerations: Ethical approval was waived because this study describes a single anonymized clinical case report. Written informed consent for the publication of this case study was obtained from the patient's legal guardians.

Conflict of Interest: The authors declare no conflicts of interest.

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