Case Report

Incomplete Kawasaki Disease Presenting as Anterior Mediastinitis and Neck Cellulitis

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Abstract

Background: While classical Kawasaki disease (KD) is easily diagnosed by a well-established set of diagnostic criteria, the diagnosis of incomplete KD remains elusive. **Clinical Description:** A 1-year-old girl presented with high fever, painful swelling, and redness over the anterior part of the neck, associated with swallowing difficulty. There was no rash, redness of eyes, or lymphadenopathy. **Management and Outcome:** Considering cellulitis of the neck, antibiotics were initiated. She developed stridor and received dexamethasone. Fever subsided, dysphagia and neck swelling improved by 3 days, but fever re-appeared on day 5, with progressively rising total leukocyte counts, platelets, and C-reactive protein. Magnetic resonance imaging neck showed features of anterior mediastinitis. Bacillus Calmette–Guérin reactivation was noted on day 8. Suspecting atypical KD, echocardiography was done thereafter, which showed left main coronary artery aneurysm. Intravenous immunoglobulin and aspirin were started. Because of medium sized aneurysm at repeat echo, increasing in size, treatment was intensified with infliximab and prednisolone. Follow-up echo after 2 weeks, showed diminution in the size of aneurysm with complete regression by 2 months. **Conclusion:** The case highlights yet another atypical manifestation of KD. Only a high index of suspicion in an unusual course of febrile illness in a child can diagnose such incomplete forms of KD.

Keywords: Atypical Kawasaki disease, bacillus Calmette–Guérin reactivation, coronary aneurysm

Kawasaki disease (KD) is the most common cause of acquired heart disease among children. Coronary artery aneurysms can lead to myocardial ischemia, infarction, and sudden death. The definition of atypical KD should be reserved for patients who have clinical manifestations not fulfilling the criteria for classic KD.[1,2] The potentially severe outcome of either classic or incomplete KD without therapy emphasizes the importance of early identification and treatment of all patients with the disease. We report a case of incomplete KD with a rare presentation.

Clinical Description

A 1-year-old girl presented with fever and cough for 5 days, painful swelling, and redness at the front of the neck, associated with difficulty in swallowing for 1 day. There was no rash, lymphadenopathy, altered sensorium, redness of eyes, icterus, vomiting, diarrhea, or urinary symptoms. She was being treated by a local practitioner with oral amoxicillin/clavulanic acid along with paracetamol since the first day of illness. Past history and family history were insignificant, and she had received all her vaccines till date.

On admission, the child was conscious but irritable with a temperature of 101°F, heart rate of 130/minute, respiratory rate of 32/min, SpO_2 of 97% in room air, and normal volume pulses with a blood pressure of 86/68 mmHg. Her anthropometry was appropriate for age. Mild pallor was present, but icterus, cyanosis, clubbing, edema, and lymphadenopathy were absent. The swelling on the anterior part of the neck was diffuse, 10 cm × 5 cm, with erythema of overlying skin and tender on palpation. Examination of the throat and oral cavity was unremarkable, and systemic examination was normal.

Management and Outcome

Considering a possible diagnosis of cellulitis of the neck, she was started on intravenous ceftriaxone and flucloxacillin. Initial investigations [Table 1] revealed anemia, a high total leukocyte count (TLC), and raised C-reactive protein (CRP). The throat swab did not yield any pathogen, and the blood culture was sterile. The child

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developed stridor on day 2, and considering a possibility of acute laryngotracheobronchitis, he received a single dose of intravenous dexamethasone along with adrenaline nebulization. Because of dysphagia and stridor being present in the background of neck cellulitis, magnetic resonance imaging (MRI) [Figure 1] of the neck was performed, which showed a diffuse increase in signal intensity of subcutaneous tissue on fat-suppressed images with multiple enlarged lymph nodes bilaterally. T2 fat suppressed (T2FS) screening of the anterior chest showed edema of the mediastinum with enlarged anterior mediastinal lymph nodes, suggesting anterior mediastinitis.

Fever subsided on day 3 of hospitalization, stridor subsided, dysphagia improved, and the child started accepting oral feeds. In view of the symptomatic improvement, otorhinolaryngology opinion was deferred. The neck swelling gradually subsided, but fever recurred on day 5, with an increase in TLC and platelets. Along with persistent fever, TLC and platelets continued to rise, with the CRP almost doubling from 67.3 mg/l to 126 mg/l from days 6–8. Furthermore, on day 8, erythema and edema of bacillus Calmette–Guérin (BCG) site were noticed, suggesting BCG reactivation. Our patient initially presented as neck cellulitis and anterior mediastinitis was unresponsive to antibiotics, continued to have high TLC and CRP with progressively increasing platelets. BCG reactivation and echo cardiography showing coronary artery aneurysm finally clinched the diagnosis of KD. A two-dimensional (2D) echocardiography was advised which showed small left main coronary artery (LMCA) aneurysm (+2.9 Z). Thus, the diagnosis was revised to atypical KD with LMCA small aneurysm presenting as neck cellulitis and anterior mediastinitis.

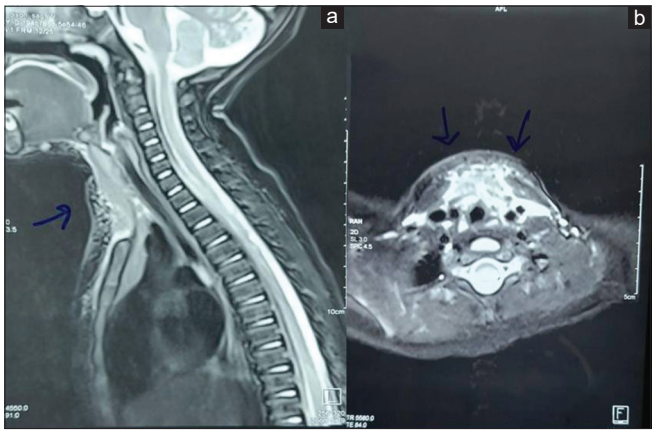

Figure 1: (a and b) Magnetic resonance imaging neck showing edema of the anterior mediastinum, enlarged lymph nodes, and cellulitis suggestive of anterior mediastinitis. The arrows show the cellulitis of the anterior chest wall

The child was started on intravenous immunoglobulin (IVIg) 2 g/kg over 16 hours over along with aspirin 5 mg/kg as per institutional protocol on day 8 of admission. The child became afebrile within 24 hours of completion of IVIg therapy. Repeat echocardiography after 2 days showed an increase in the size of LMCA aneurysm (+6.9 Z). Considering the presence of a medium-sized aneurysm, treatment was intensified with infliximab (10 mg/kg single dose) on day 10. On day 12, LMCA aneurysm showed some regression (+5.8 Z), but CRP remained high (67.7 mg/l). Hence, oral prednisolone 2 mg/kg was started, and the child was discharged on prednisolone and aspirin, with plan to taper off prednisolone over 15 days. Progressive regression in the size of LMCA aneurysm was seen after 15 days (+4.6 Z) with complete regression by 2 months.

Discussion

Kawasaki disease, predominantly an acute-onset vasculitis of medium vessels, primarily affecting children below 5 years of age, though known in the literature for over 50 years, still puzzles pediatricians with its ever-evolving atypical manifestations. The exact cause remains unknown, with no single pathognomonic clinical or laboratory finding for

Table 1: Serial investigations of the index child

Investigations/treatment	Day 1	Day 3	Day 6	Day 8	Day 10	Day 12	After 2 weeks	After 8 weeks
Hb (g/dL)	9.3	8.9	9.0	8.6		8.1		
TLC (/mm^3)	12,000	1,3980	18,700	18,500		14,300		
DLC (%)	N52L41	N37L59	N75L45	N61L32		N24L70		
Platelets (/mm^3)	446,000	556,000	683,000	873,000		992,000		
CRP (mg/L)	109.2	53.6	67.3	126		67.7		
Na/K (mg/dL)	138/5.05							
Urea/creatinine (mg/dL)	19/0.23							
TP/albumin (mg/dl)	6.6/3.9							
AST/ALT (IU/L)	70/74							
Bilirubin (total/direct) (mg/dl)	0.38/0.2							
Echocardiography				LMCA + 2.93Z	LMCA + 6.91Z	LMCA + 5.8 Z	LMCA + 4.18Z	LMCA + 2.07Z
Therapy				IVIg + Aspirin	Infliximab	Prednisolone		

Hb: Hemoglobin, TLC: Total leukocyte count, DLC: Differential count, CRP: C-reactive protein, Na: Sodium, K: Potassium, TP: Total protein; AST/ALT: Aspartate aminotransferase/alanine transaminase, LMCA: Left main coronary artery, IVIg: Intravenous immunoglobulin

making the diagnosis. The standard diagnostic criteria of KD are based on a set of symptoms and signs developed by the American Heart Association (AHA)[1] or the Japanese Kawasaki Disease Research Committee.[2] Atypical KD was used for patients who did not meet the classical criteria but had coronary complications, but recently, it was proposed to use atypical KD interchangeably with incomplete KD, regardless of coronary complications. The incidence of incomplete KD ranges from 15% to 36.2%, being relatively more common in children at extremes of age (<1 year old or >5 years).[3] No other factor has been found to be associated with the incidence of incomplete KD.

As per AHA, a diagnosis of incomplete KD is possible in children with fever and two principal features (according to the Japanese criteria, three principal symptoms), along with six additional laboratory and echocardiographic criteria. To diagnose atypical KD, a patient should have more than three laboratory abnormalities, which include anemia for age, sterile pyuria, hypoalbuminemia (≤3 gm/dl), leukocytosis (≥15000/mm^3), and elevation of alanine aminotransferase (ALT) and platelet cell count.[4,5] In our case, we found anemia, leukocytosis, and elevated platelet count, though sterile pyuria, hypoalbuminemia, and elevated ALT features were absent.

Other findings which support the diagnosis of atypical KD are inflammation at the BCG inoculation site (found in our case), anterior uveitis, elevated levels of brain natriuretic peptide (BNP) and N-terminal pro-BNP, hyponatremia, elevation of left ventricular mass, and diastolic dysfunction of the left ventricle.[6-8] Hua *et al*.[9] reported that in patients <6 months old, total fever duration of ≥8 days, delayed diagnosis, and albumin ≤3.5 mg/dl were independent risk factors for coronary artery lesions in KD. Mediastinal lymphadenopathy is a rare finding associated with KD.[10] Our patient initially presented as neck cellulitis and anterior mediastinitis was unresponsive to antibiotics, continued to have high TLC and CRP with progressively increasing platelets. BCG reactivation and echo cardiography showing coronary artery aneurysm finally clinched the diagnosis of KD.

Conclusion

The case creates awareness among pediatricians to consider the possibility of incomplete KD in children who have had an unresolving, unexplained fever, unresponsive to antibiotics for more than 5 days, meeting some of the clinical criteria of KD, especially with clues like BCG reactivation. Incomplete KD can easily be missed, if not suspected, and delayed diagnosis may result in a higher risk of developing coronary artery lesions. Treatment needs to be intensified in the presence of coronary aneurysms.

Lessons learnt

- Index of suspicion should be high for incomplete KD in young children presenting with unexplained fever for more than 5 days associated with some evidence of systemic inflammation
- Incomplete and atypical presentations are common in infantile KD, and a 2D echocardiography should be performed at the earliest suspicion
- Treatment needs to be intensified in the presence of coronary aneurysms.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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