Pulmonary Presentation of Atypical Kawasaki Disease

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To the Editor: A 19-mo-old boy presented with fever, cough, rhinorrhoea and respiratory distress for 2 d. On examination the child was interactive and alert, febrile (101 °f) with tachypnoea, had chest indrawing and diffuse crepitations in bilateral chest regions. The child did not have rash, conjunctival congestion, lymphadenopathy or peripheral edema. Systemic examination was unremarkable. The oxygen saturation on room air was 84% and 97% with 40% oxygen supplementation. Chest roentgenogram showed bilateral hyperinflation with diffuse reticulonodular infiltrates (RNI) (Fig. 1a). Polymerase chain reaction for SARS-CoV-2, influenza, respiratory syncytial and cytomegalovirus were negative. The investigation done on day 4 and day 8 of illness are given in Table 1 which showed worsening trends of the markers of inflammation. Evaluation for tuberculosis, scrub typhus, urine and blood cultures were all non-contributory. Because of persistent distress and fever, echocardiogram was performed on day 8 which revealed aneurysm of left main coronary artery (3.7 mm; 5.62 Z score) and dilatation of left anterior descending artery (2.2 mm; 2.48 Z score) with the laboratory markers of Kawasaki disease (KD) were in worsening trend. He became afebrile and distress-free within 36 h of intravenous immunoglobulin (IVIG) therapy and aspirin at 50 mg/kg/d for 2 d followed by 3 mg/kg/d (anti-platelet dose). Roentgenogram done 5 d after IVIG showed clearing of RNI (Fig. 1b). There was

no periungual peeling or nail changes at week 2 of follow-up with normal systemic examination. Echocardiogram after 8 wk showed normal coronary arterial dimensions and anti-platelet dose of aspirin was stopped at 8 wk. The general and systemic examination was unremarkable at this point.

The diagnosis of incomplete KD has three sequential steps [1]. Our index case never followed the sequence and only had coronary artery abnormalities (CAA) without compatible clinical and laboratory feature which was perplexing to us. Occasional cases of mild CAA (2.5–5 Z score) have been reported in non-KD febrile illness but moderate CAA have never been [2–4]. The presence of moderate CAA rules out non-KD febrile illness in our case.

The incidence of pulmonary presentation of KD has been 1.83% in a retrospective cohort [5]. Bronchopneumonia was the commonest pulmonary manifestation, though pneumothorax, pleural effusion and empyema have been observed. The first clinical sign of KD was observed after a mean duration of 14.5 d from onset of clinical symptoms. Unresolved fever and distress with abnormal laboratory markers only aroused the suspicion of KD in this cohort [5]. Diagnosing KD in a child with isolated pulmonary presentation is always challenging when classical features of KD are absent. Worsening trends of hemogram, liver function test and inflammatory markers in a child with unresolving and unexplained fever and respiratory distress is a pointer of incomplete KD. It may be too late to diagnose and treat KD after the diagnostic cut-off point for laboratory values are reached, especially whenever the child has a compatible clinical setting.

Persistent fever with unexplained distress ≥ 7 d with RNI in roentgenogram should arouse a suspicion of incomplete KD. Worsening trends of inflammatory markers, even when not meeting the diagnostic cut-off, might be the earliest marker for KD in these children with isolated pulmonary presentation.

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Fig. 1 Chest roentgenogram (a) on day of admission showing bilateral hyperinflation with diffuse dense reticulonodular infiltrates, (b) done 5 d after intravenous immunoglobulin administration showing persistent bilateral hyperinflation but with clearance of reticulonodular infiltrates

Table 1 Laboratory reports on day 4 and 8 of illness

Investigation	Day 4 of illness	Day 8 of illness
Hemoglobin (g/dL)	9.6	9.1
Mean corpuscular volume (fl)	61	56
Total leucocyte count ($\times 10^3/\mu\text{L}$)	16.5	18.7
Platelet count ($\times 10^3/\mu\text{L}$)	312	402
Erythrocyte sedimentation rate (mm/h)	28	35
C-reactive protein (mg/dL)	0.9	1.2
Urine microscopy	No leucocytes	No leucocytes
Serum albumin (g/dL)	3.5	3.1
Aspartate transaminase (U/l)	31	34

Compliance with Ethical Standards

Conflict of Interest None.

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