CASE REPORT

"Unresolving pneumonia" as the main manifestation of atypical Kawasaki disease

Y Uziel, P J Hashkes, E Kassem, G Gottesman, B Wolach

Two cases of atypical Kawasaki disease (KD) manifested as persistent lobar lung consolidation, prolonged fever, and active inflammatory laboratory markers unresponsive to antibiotic treatment are reported. One of the children developed a giant coronary aneurysm. Atypical KD should be considered in the differential diagnosis of young children with prolonged fever and lobar consolidation unresponsive to antibiotics.

A typical Kawasaki disease (KD) is associated with an increased risk of coronary artery abnormalities (CAA). Early diagnosis and initiation of intravenous immunoglobulin (IVIG) therapy is essential in significantly reducing this risk. Since the diagnosis of KD is based mainly on clinical findings, the paediatrician treating a child with prolonged fever without typical signs of KD needs an increased degree of suspicion for this diagnosis. In a large series of chest radiograph findings in children with KD, nearly 15% of the patients had pathological findings, mainly interstitial lung changes. We did not find reports in the English literature on lung consolidation as part of KD or as the presenting symptom of KD. We therefore report two cases of young children with “unresolving” lobar consolidation as the main presenting symptom of KD.

CASE 1

A 2.5 year old, previously healthy girl, was hospitalised with prolonged fever and “unresolving pneumonia”. She developed a temperature up to 39.5–40°C and was initially diagnosed as having bullous myringitis; she was treated with amoxycillin. On the fifth day of fever she was referred to the emergency department for further evaluation. The only positive physical examination findings were mild oropharyngeal erythema, small submandibular lymph nodes, and left bullous myringitis. There were no respiratory symptoms or signs, and the respiratory rate, oxygen saturation, and lung auscultation were normal.

Initial laboratory results were as follows: erythrocyte sedimentation rate (ESR) 125 mm/h, haemoglobin 10.6 g/l, white blood cell (WBC) count 7.8×10⁹/l, and platelet count 558×10⁹/l. Chest radiographs (fig 1A) performed as investigation for fever revealed consolidation in part of the left upper lobe. She was treated with various antibiotics, including ceftriaxone, azithromycin, and amoxycillin clavulonate for 10 days, but the fever persisted.

On the 16th day of her disease she was admitted to our paediatric department. Physical examination was unremarkable. She still had no respiratory signs. Laboratory results were as follows: ESR 125 mm/h, haemoglobin 8.4 g/l, WBC count 11.9×10⁹/l with a polymorphonuclear cell predominance, platelets 773×10⁹/l, aspartate aminotransferase 54 U/l, total protein 6.3 g/l, albumin 3 g/l, and urine sediment revealed 25 WBCs in a high powered field. Chest radiographs continued to show the same consolidation.

Investigations for infectious, malignant, and autoimmune diseases (cultures, serology for mycoplasma, Epstein-Barr virus (EBV), cytomegalovirus, hepatitis A, B, and C, brucella, chlamydia, rickettsia, Mantoux test, bone scan, abdominal ultrasound, bone marrow aspiration, antineutrophil cytoplasm antibodies, antinuclear factor, echocardiography) were negative.

Figure 1 Consolidation of part of the left upper lobe of the lung (A), with complete resolution on follow up (B).

Abbreviations: CAA, coronary artery abnormalities; EBV, Epstein-Barr virus; ESR, erythrocyte sedimentation rate; IVIG, intravenous immunoglobulin; KD, Kawasaki disease; WBC, white blood cell
negative. Since she had a quotidian pattern of fever with one spike per day (without rash), the diagnosis of systemic onset juvenile idiopathic arthritis was considered.

On the 20th day of the disease she developed arthritis of both knees, elbows, and ankles. The ESR increased to 180 mm/h, her haemoglobin decreased to 7.6 g/l, and platelets increased to 980×10^9/L. Prednisone 2 mg/kg/day was administered, resulting in an immediate defervescence. On the 22nd day of the disease, skin desquamation of the fingertips occurred and she was diagnosed with atypical KD. Two weeks later, a follow up chest radiograph (fig 1B), laboratory tests, and cardiac echocardiogram were normal. No CAA have been observed after two years of follow up.

**CASE 2**

A 2 year old, previously healthy boy was diagnosed with left lower and right middle lobar pneumonia based on consolidation on chest radiographs after five days of fever and leukocytosis. He had a cough; physical examination revealed mild tachypnoea (respiratory rate 60/min), oxygen saturation 94%, and lung crepitations on auscultation.

There was no clinical or radiologic response to various antibiotics. On the eighth day of his illness he developed severe diarrhoea, anasarca, and ascitis. Laboratory results were as follows: ESR 75 mm/h, haemoglobin 8.7 g/l, WBC count 34×10^9/L, platelets 403×10^9/L, and albumin 2.7 g/l. Echocardiography performed on the tenth day of disease revealed several large coronary aneurysms. He was treated with two courses of IVIG and intravenous pulse steroids until the fever abated. Follow up chest x-ray examination 20 days later was normal. After three years of follow up he is asymptomatic. However, a giant aneurysm in the left main coronary artery is still present, and he is currently treated with aspirin and warfarin.

**DISCUSSION**

We have described two young children with atypical KD manifested as prolonged fever, and “unresolved” lobar consolidation. Unfortunately one of the children was diagnosed only after the development of CAA. Between 15% and 25% of untreated patients with KD develop CAA. Early IVIG therapy within the first 10 days reduces this risk to less than 5%, and the risk of giant aneurysm development to less than 1%. Hence early diagnosis and treatment are crucial.

Atypical KD is defined as a fever of more than five days duration in the presence of less than four of the five classical clinical criteria or fever with detection of CAA on echocardiography. Only 20–60% of children with CAA associated with KD meet the classical diagnostic criteria. Reports of atypical KD in infants and older children describe these patients as being at greater risk of developing CAA. Therefore the diagnostic criteria for KD should serve only as a guideline, while in other cases a high degree of clinical suspicion is needed. Other clues to the diagnosis of KD include gastrointestinal (vomiting, diarrhoea, weight loss, hepatitis, gall bladder hydrops), musculoskeletal (arthralgia, arthritis), central nervous (headache, meningeal signs, cerebrospinal fluid pleocytosis), and genitourinary symptoms (urethritis, metanephrosis, sterile pyuria). Other cardiac abnormalities may include myocarditis and pericarditis. In our report we add lung involvement as another early organ presentation in KD. Hence early diagnosis and treatment are crucial.

Unresolving pneumonia in Kawasaki disease

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