CASE REPORT

# Appendicitis in an Infant with Atypical Features

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## ABSTRACT

Acute appendicitis is an uncommon and challenging disease in infancy. Usually, the clinical presentation in neonates and infants is non-specific and varies depending on the age of the child and duration of the disease. Diagnosis of incomplete and atypical Kawasaki disease (KD) in infants is also a challenging aspect and there is no gold standard for this diagnosis and sometimes fever is the only symptom that could be found. Herein, we report a 6-month infant with a 7 days of fever and bilateral pleural effusion, elevated erythrocyte sedimentation rate, thrombocytosis, hypo-albominemia, normal abdominal ultrasound, and primary diagnosis of KD. Final diagnosis was perforated retrocecal appendicitis and abscess formation. Physicians should be aware of the vague signs and symptoms of acute appendicitis in neonates and infants and consider this diagnosis to prevent delayed diagnosis, inappropriate treatment, and consequent morbidity and mortality.

**Key Words:** *Appendicitis. Infant. Kawasaki disease. Retrocecal appendix. Perforation. Abscess.*

## INTRODUCTION

Acute appendicitis is an uncommon disease in infants and with challenging aspects that may cause the prompt and timely diagnosis very difficult.[1] The signs and symptoms usually are non-specific in this age group and the patient is not cooperative and able to express the exact cause of discomfort.[1]

The purpose of this case report is to present a rare case of acute appendicitis in a 6-month infant with primary diagnosis of atypical Kawasaki disease (KD). Clinicians should consider this diagnosis in infants to reduce morbidity and mortality.

## CASE REPORT

A previously healthy 6-month female infant with 7 days of fever and irritability was referred to Ali Asghar Children's Hospital with primary diagnosis of atypical KD. Her primary physical examination was normal except for fever. Primary laboratory data showed WBC = $3.4x10^3/mm^3$ (normal range: 6.0 - 13.5 x $10^3/mm^3$) with 14% neutrophil, 5% Band, 5% eosinophil, 74% lymphocyte and 2% monocyte; hemoglobin (Hb) = 10.6 g/dl, platelet 660,000/$mm^3$, ESR = 91 mm after one hour (normal < 15 mm/hour), quantitative C-reactive protein = 90 mg/L, reticulocyte count = 2%, serum albumin = 2.7 g/dl and serum total protein = 6.3 g/dl. CSF analysis result and other routine biochemical tests were within normal limits. Echocardiography showed a small atrial septal defect and patent foramen ovale but no evidence compatible with cardiac involvement of KD.

One day after admission, her vital signs became unstable and she was transferred to ICU following tachypnea, tachycardia and increased agitation. Abdominal and pelvic ultrasound was normal but X-ray (CXR) and ultrasonography showed bilateral pleural effusion. Repeated complete blood count (CBC) examination at this time revealed decreased WBC, neutrophil count and Hb (WBC = $2.1x10^3$ /$mm^3$ with 10% neutrophil and Hb = 7 g/dl).

On physical examination, the abdomen was distended and palpation of the abdomen revealed right-sided tenderness. As repeated abdominal and pelvic ultrasonography were reported to be normal, the patient was evaluated with abdominal computed tomography (CT), recommended by consulting surgeon. Meropenem, vancomycin and intravenous immunoglobulin (IVIG) were started empirically on the probability of severe sepsis and/or KD. Abdominal CT scan with contrast

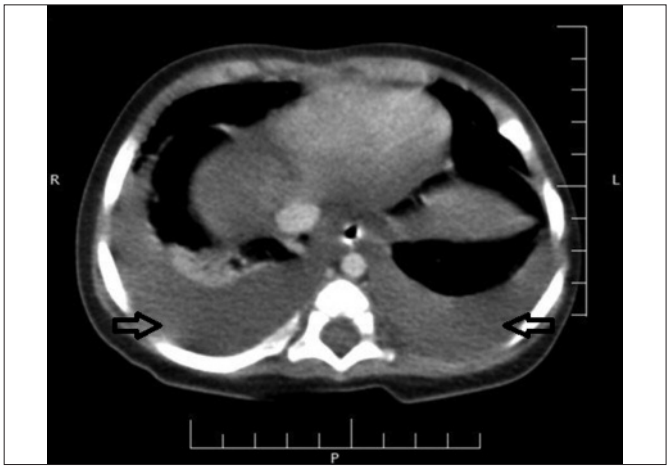


**Figure 1:** Bilateral pleural effusion with sub segmental collapse of the lower lobe of the right lung.

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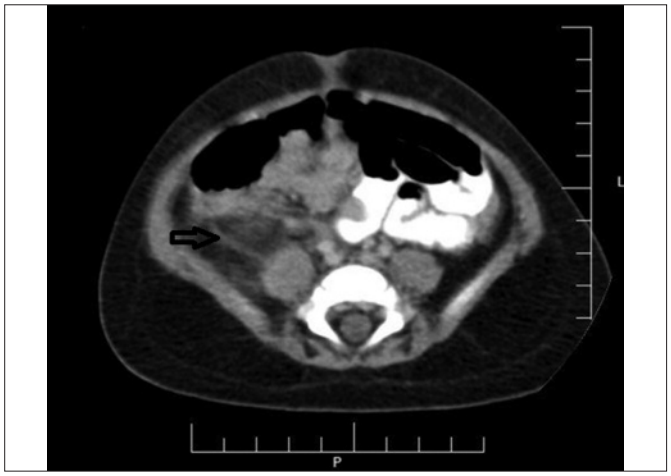

**Figure 2:** Fat stranding in right lower quadrant of the abdomen adjacent the bowel loops.

showed mild liver enlargement, bilateral moderate pleural effusion with maximum anteroposterior (AP) diameter of 20 mm on both sides (Figure 1) and fat stranding in RLQ adjacent to the bowel loops (Figure 2).

The patient underwent surgery and a perforated retrocecal appendix and retroperitoneal abscess were found that was managed with appendectomy and abscess drainage. The pleural effusion disappeared spontaneously on ultrasound after 2 days. She was discharged after receiving intravenous antibiotics for a week. Pathological evaluation confirmed the peroperative diagnosis. Postoperative follow-up after 8 months did not show any significant complication.

## DISCUSSION

Acute appendicitis is an uncommon disease in infants and neonates. Less than 2% of all appendicitis cases occur in infants and it is even more uncommon in neonates.[2]

The clinical presentations of acute appendicitis in neonates and infants might be different from adults and vary depending on the age and duration of the disease.[2]

In infantile appendicitis, fever, irritability, vomiting, diarrhea, and grunting have all been described as presenting signs and symptoms.[1]

These non-specific manifestations in addition to the lack of specific laboratory tests have complicated the diagnosis of acute appendicitis and made it a major challenge in pediatric age group.[2] It could be the reason of 80 - 100% perforation rate of appendix in children under 3 years of age.[2]

Unfortunately, the diagnosis of incomplete and atypical KD in infants is also a challenging situation, much like infantile appendicitis, and there is no gold standard for this diagnosis.[3]

The presenting signs and symptoms of the infants with KD do not usually fulfill the criteria needed for the diagnosis of this disease; and sometimes fever is the only symptom that could be found. According to the most recent recommendations of American Heart Association, infants younger than 6 months with more than 7 days of fever without another explanation should be evaluated for KD. The patient should undergo laboratory testing and if evidence of systemic inflammation is found, an echocardiography should be performed even if the infant has no clinical criteria.[4] This patient was febrile for 7 days with increased inflammatory markers such as ESR, CRP and platelet count without any explanation at that time, therefore, KD was a plausible differential diagnosis. However, the patient had progressive leukopenia and neutropenia which are considered atypical findings in this disease.[3]

There are rare reports of KD accompanying true appendicitis.[5] As far as the authors are aware, this is the first report of a true acute perforated appendicitis with primary diagnosis of atypical KD.

The role of imaging in the diagnosis of acute appendicitis in infants and children, in whom the physical examination and laboratory data are frequently vague, is critical. Graded compression sonographic technique to diagnose acute appendicitis has shown high sensitivity and specificity in several studies, but the rarity of appendicitis in infants is an impediment to determine its precise sensitivity and specificity in this age group.

Ultrasound examination is operator dependent. In addition, an aberrant location of the appendix, such as a retrocecal position, and appendiceal perforation might be the causes of non-visualization of the appendix.[6] So normal ultrasound examination result could not rule out the diagnosis of acute appendicitis as in this case.

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