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# How to distinguish: appendicitis or acute abdominal attack of FMF?

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**Summary**

Familial Mediterranean fever is an autosomal recessive inherited disease characterized by recurrent attacks of fever and pain secondary to polyserositis, mainly of the abdomen and joints. The symptoms are sudden onset, occur before the age of 20 in 90% of the patients and generally decrease spontaneously within 1-4 days. Because of similar clinical and laboratory settings, it is difficult to differentiate an attack of Familial Mediterranean fever from acute appendicitis and other acute abdominal conditions. Therefore preoperative evaluation is very important to prevent unnecessary surgical approach in Familial Mediterranean fever patients with abdominal attacks. In these circumstances, contrast enhanced abdominal tomography may be preferred if there is still doubt about the diagnosis. We report a case of perforated appendicitis and Familial Mediterranean fever whom we evaluated with contrast enhanced abdominal tomography.

**Key words:** Appendicitis, computed tomography, Mediterranean fever

**Özet**

**Apandisit mi, ailevi Akdeniz ateşi atağı mı: nasıl ayırt edebiliriz?**

Ailevi Akdeniz ateşi, tekrarlayan ateş ile özellikle karın ve eklemlerde, poliserözite ikincil ortaya çıkan ağrı ataklarıyla karakterize, otozomal resesif geçiş gösteren bir hastalıktır. Bulgular ani başlangıçlıdır, hastaların %90'ında 20 yaşından önce ortaya çıkar ve genellikle 1 ile 4 gün içinde kendiliğinden kaybolur. Benzer klinik ve laboratuvar bulguları nedeni ile ailevi Akdeniz ateşi atağını, akut apandisit ve diğer akut karın tablolarından ayırt etmek zor olabilir. Ailevi Akdeniz ateşinin akut atağı tanısı ile takip edilen hastalarda gereksiz cerrahi girişimleri önlemek açısından ameliyat öncesi değerlendirme önem kazanmaktadır. Bu koşullar altında, klinik tanının halen şüpheli olduğu durumda kontrastlı batın tomografisi tanının doğrulanması amacı ile tercih edilebilir. Bu yazıda kontrastlı batın tomografisi ile tanı konan, ailevi Akdeniz ateşi ve perfore apandisit tanılı bir hasta sunulmuştur.

**Anahtar kelimeler:** Apandisit, bilgisayarlı tomografi, Akdeniz ateşi

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## Introduction

Familial Mediterranean fever (FMF) is one of the most common causes of acute abdominal pain in children among the Mediterranean countries (1-3). Because of similar clinical and laboratory findings, it may be confused with appendicitis (2,4). There is no article about the role of computed tomography (CT) on differential diagnosis of acute abdominal attack of FMF and appendicitis in children. We herein report a case with FMF in whom we utilized CT for the diagnosis of appendicitis.

## Case

A 9-year-old male who was being followed for FMF at the Department of Pediatrics at our institution since 2000 was admitted with severe abdominal pain, bilious vomiting, dysuria and fever. On the third day of diagnosis and treatment of urinary tract infection, we consulted the patient because of his symptoms. On physical examination, body temperature was 38.4 °C, and diffuse abdominal rigidity, distention, rebound tenderness, reduced intestinal peristaltic sounds were detected. Leukocytosis, high C-reactive protein and high fibrinogen level were determined by laboratory tests. Also proteinuria and leukocyturia were present in urine analysis. Multiple mesenteric lymphadenopathies and minimally free fluid between intestinal segments were diagnosed by ultrasound (US) on the second day of symptomatic onset. Because of diagnostic dilemma due to obscure clinical and radiological findings, contrast enhanced abdominal tomography (CEAT) was performed to confirm these findings. The CEAT reported a blind ending, edematous and larger than 10 mm in

diameter tubular structure arising from medial side of cecum. There is a calcific opacity more than 7 mm in diameter, in the proximal luminal side of the tubular segment (Figure 1).

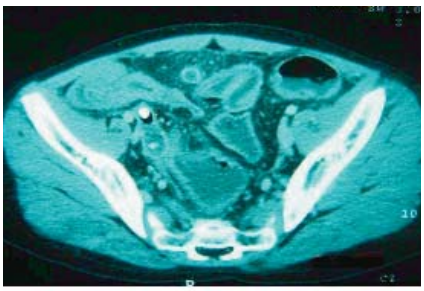

**Figure 1.** Computed tomographic appearance of the fecalith in the appendix vermiformis

The patient underwent the operation with the provisional diagnosis of perforated appendicitis. Exudative free fluid, multiple abscess focuses between intestinal segments and perforated appendix were observed at the exploration. Then appendectomy was performed. Ceftriaxone and metronidazole were administered postoperatively for 7 days. The patient was discharged at the postoperative 7th day uneventfully.

## Discussion

FMF is an autosomal recessive inherited disease characterized by recurrent attacks of fever and pain secondary to polyserositis, mainly of the abdomen and joints (3,4). FMF resulting from mutation in the gene, which is located in the short arm of chromosome 16, is one of the most common causes of acute non-surgical abdominal pain in children among the Mediterranean countries (1-3). Abdominal attacks are characterized by severe abdominal pain with fever and abdominal tenderness in 95% of patients (2,4-6). Abdominal pain with peritoneal signs is the first clinical presentation in 50% of the patients. Laboratory findings include leukocytosis, increased level of acute phase reactants (2,4,5). Approximately two-third of FMF patients undergo surgical intervention with the provisional diagnosis of appendicitis in any period of life and the appendix is found normal in most of the cases (4,7).

Acute appendicitis due to obstruction of the lumen by fecaliths, lymphoid hyperplasia, foreign bodies and parasites is the most common cause of emergency abdominal surgery in childhood (8). It is also characterized by lack of appetite, abdominal pain, vomiting, abdominal tenderness and rigidity. Laboratory findings are similar to laboratory findings, which are detected in FMF. If it is not diagnosed timely, this condition can manifest with some complications such as perforation, periappendicular abscess and peritonitis.

It may be difficult to differentiate FMF and other surgical acute abdominal conditions during abdominal attack, especially for acute appendicitis due to similar symptoms and laboratory findings (5,7). Because of nonspecific clinical and laboratory findings such as leukocytosis, proteinuria and leukocyturia, our case was followed-up with the diagnosis of urinary tract infection and FMF at the Department of Pediatrics. To hinder complication, urgent exploratory laparotomy is recommended by some centers in patients with acute abdominal attack of FMF (4,5). Some authors recommend elective laparoscopic appendectomy to prevent misdiagnosis and unnecessary emergency surgery (4,7). In this respect preoperative radiological evaluation gets importance utmost to prevent unnecessary surgical approach in FMF and may be helpful for accurate diagnosis and treatment.

Ultrasonography is the generally preferred diagnostic radiological tool because of availability. Although there are several clinical trials and articles about the value of ultrasonography in the diagnosis of acute abdominal conditions in children, it is not useful in some patients; such as when the appendix is perforated or retrocecal. In addition, some technical problems such as obesity, abdominal tenderness, increased bowel gas, and guarding are other difficulties (9,10). Also the accuracy of the diagnosis depends on the radiologist (8-10). In these complex cases appendiceal CEAT scan is more helpful than ultrasonography for evaluating patients with acute abdominal attack (9). It is fast, cost effective, has a high positive predictive value and identifies abscess and phlegmon better. In such cases the findings on CEAT scan are blind-ending tubular segment in connection with cecum with a diameter of more than 6 mm and thickened wall containing contrast material. Heterogenity and hypertrophy in surrounding adipose tissue are visible that indicate severe inflammation. Periappendicular fluid collection and intraabdominally free fluid can be seen in perforated cases. In some cases fecal impaction may be visible in the lumen of appendix (6,8). The diagnostic accuracy of CT has been reported to range between 93% to 98%. This rate was reported as 71% to 97% for US (8).

In our patient the clinical findings were similar to both of acute abdominal attack of FMF and appendicitis. Ultrasonographic findings were nonspecific and not helpful. So we decided to perform CEAT to make definitive diagnosis. Findings were almost similar with the literature findings, which are aforementioned.

As a conclusion, some FMF cases with acute abdominal attack may be confused with acute abdominal conditions, and some FMF cases actually have an acute abdominal condition, such as an acute appendicitis simultaneously as in our case. Although ultrasonography is a valuable diagnostic tool for the differential diagnosis of

abdominal attack in FMF and acute appendicitis, it is not effective in some cases who do not have specific ultrasonographic findings. In these cases CEAT may be very useful for the evaluation of acute abdominal conditions.

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