

# An unusual complication of familial Mediterranean fever: Intestinal volvulus and necrosis

Ailevi Akdeniz Ateşi'nin ender bir komplikasyonu: İntestinal volvulus ve nekroz

Gülay A. TİRELİ, Murat ÜNAL, Serdar SANDER

Department of Pediatric Surgery, Bakırköy Maternity and Children's Hospital, Istanbul

*Familial Mediterranean fever is an autosomal recessive disease characterized by recurring inflammatory attacks of synovial membranes. More than 95% of patients show peritoneal involvement which mimics acute abdomen and can sometimes cause unnecessary surgical intervention. The authors present two patients with the diagnosis of familial Mediterranean fever who underwent surgery because of rare abdominal complication of the disease. Two patients with the diagnosis of familial Mediterranean fever underwent laparotomy, and segmental small bowel resection was done because of the necrosis. Adhesive intestinal obstruction with associated bowel strangulation and volvulus is a rare complication of familial Mediterranean fever, and this life-threatening emergency must be kept in mind.*

**Key words:** Familial Mediterranean fever, adhesive intestinal obstruction, volvulus

*Ailevi Akdeniz Ateşi, sinovyal membranların tekrarlayan inflamatuvar atakları ile karakterize otozomal resesif geçişli bir hastalıktır. Hastaların %95' inden fazlasında zaman zaman gereksiz cerrahi girişime yol açan akut batın ataklarına yol açan peritoneal tutulum gözlenir. Yazarlar hastalığın peritoneal tutulumuna bağlı ender gelişen bir komplikasyon nedeni ile ameliyat edilmiş iki olgu sunmuşlardır. Ailevi Akdeniz Ateşi tanılı iki hastada akut batın tablosu ile laparotomi yapılmış ve ince barsak nekrozu saptanarak rezeksiyona gidilmiştir. Adheziv intestinal tıkanıklık ve buna bağlı intestinal volvulus Ailevi Akdeniz Ateşi'nin ender görülen bir komplikasyonudur ve bu önemli komplikasyon akılda tutulmalıdır.*

**Anahtar kelimeler:** Ailevi Akdeniz Ateşi, adheziv intestinal obstrüksiyon, volvulus

## INTRODUCTION

Familial Mediterranean fever (FMF) is an autosomal recessive disease characterized by recurring inflammatory attacks of synovial membranes such as pericardium, peritoneum, and joints. More than 95% of patients show peritoneal involvement (1) which mimics acute abdomen and can sometimes cause unnecessary surgical intervention.

## CASE REPORTS

### Case 1

A 12-year-old boy was admitted to the hospital with a complaint of colicky abdominal pain and bilious vomiting of two-days' duration. He had a history of recurring sterile pericarditis, abdominal pain, anemia, and hepatosplenomegaly and was diagnosed to have FMF. He had received colchici-

ne treatment for three months. Genetic research revealed homozygotes M694V-gene mutations on 10th exon of the MEFV gene. At the recent admission, physical examination revealed slight hepatosplenomegaly, a mass below the umbilicus, generalized abdominal tenderness, and rebound tenderness. Plain abdominal X-ray showed multiple air-fluid levels and ultrasound examination revealed intestinal distention and moderate intraabdominal fluid.

The patient underwent laparotomy, which revealed extensive intraabdominal adhesions, one of which resulted in ileal volvulus and necrosis 20 cm in length. Primary resection and anastomosis and bridectomy were performed. The patient was discharged uneventfully.

**Address for correspondence:** Gülay A. TİRELİ  
Zuhuratbaba Sarsılmaz Sokak Uğur Apt. No:6/1,  
34740, Bakırköy, Istanbul, Turkey  
Fax: +90 212 571 47 90  
E-mail: gultireli@yahoo.com

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## Case 2

A 13-year-old boy who was diagnosed to have FMF and who received colchicine treatment for five years admitted to the hospital with a complaint of abdominal pain and bilious vomiting of three-days' duration. Extensive abdominal tenderness was detected. Laboratory studies revealed leukocytosis. There were no specific findings in abdominal X-ray; abdominal ascites was found on ultrasound. Patient was operated and a necrotic ileal segment 15 cm in length was found due to obstructive peritoneal adhesions. The necrotic segment was removed and anastomosis was performed. Patient was discharged at 5th postoperative day uneventfully.

## DISCUSSION

There are two clinical features of associated peritoneal involvement in FMF. Benign abdominal pain attacks which mimic "acute abdomen" are the most common form and sometimes lead to unnecessary abdominal explorations. According to the large series by Schwabe et al. (3), of the 100 cases, 55 underwent 82 operations, of which only 12 (14%) revealed abnormal abdominal findings. During these explorations, usually benign peritoneal adhesions and some amount of sterile exudate within the abdominal cavity are found. The other and more serious abdominal form is the obstructing intestinal adhesions due to recurrent peritonitis. In that case, the peritoneum shows acute inflammation consisting of sterile exudate containing fibrin and polymorphonuclear cells in the peritoneal cavity, and the organization of this exudate results in primary peritoneal adhesions (1). In rare condi-

tions, these adhesions might lead to strangulation or volvulus and bowel necrosis. It is very important to differentiate the two clinical pictures because of the serious result of the adhesive intestinal obstruction. In a report by Michaeli et al. (4), exact diagnosis of the bowel strangulation could be made on postmortem examination. All characteristics of the benign attacks of peritonitis such as high fever, pain, abdominal distention, rebound tenderness, rigidity, and absence of peristalsis are also found in the course of adhesive intestinal obstruction. But signs and symptoms of the benign attacks end within 12-24 hours of course while the clinical picture deteriorates progressively in the other (1, 4, 5). The presence of a mass, as seen in our first patient, might be determined, and must alert the physician to intestinal strangulation (4). The English literature contains a small number of cases on this rare complication of FMF, and the reported cases are briefed in Table 1.

**Table 1.** Summary of the FMF patients with adhesive intestinal obstruction and/or strangulation/volvulus as reported in English literature

Author	Year	Number of patients	Death
Sohar <sup>1</sup>	1967	4	2
Schwabe <sup>3</sup>	1974	1	-
Tal <sup>6</sup>	1980	3	1
Ciftci <sup>5</sup>	1995	11	-

In conclusion, adhesive intestinal obstruction with associated bowel strangulation and volvulus is a rare complication of FMF. This life-threatening emergency must be kept in mind in children with FMF and abdominal pain attacks.

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