INTRODUCTION

Familial Mediterranean fever is one type of hereditary periodic fever syndrome. Abdominal symptoms accounts for 95% of all patients. When atypical pain fails to resolve spontaneously or persistent the surgeon should consider the diagnosis of intestinal obstruction due to adhesive bands.

Yalcinkayer et al. proposed another set of criteria for pediatric FMF but are limited to single ethnic group, were MEFV mutations in both alleles.[4]

FMF abdominal pain is very similar to a surgical acute abdomen. However, symptoms characteristically resolve spontaneously within 48–72 h. Abdominal pain can be localized or diffuse from the start. This pain may be due to peritoneal inflammation or due to adhesions. Self-resolving peritonitis is the most common clinical feature of FMF. However, repeated attacks of peritonitis may result in the formation of peritoneal adhesions that may cause adhesive bowel obstruction requiring urgent surgical intervention.[5]

Abdominal pain in FMF patients should be carefully evaluated according to the clinical features of the patient. It is estimated that 30–40% of patients underwent unnecessary surgical intervention such as appendectomy, cholecystectomy, and negative laparotomy.[3] In FMF patients, other causes of gastrointestinal manifestation are episodic abdominal pain, side effects of colchicine, gastrointestinal amyloidosis, vasculitis, inflammatory bowel disease, diarrhea, constipation, paralytic ileus, or mechanical ileus.[4,6] When atypical pain fails to resolve spontaneously or is persistent, the surgeon must consider the diagnosis of intestinal obstruction due to adhesive bands. Failing to recognize this may result in strangulation or gangrene of bowel. Therefore, this life-threatening surgical emergency should be kept in mind in the differential diagnosis of acute abdomen of FMF. There should not be any delay in exploration, if surgical diagnosis of intestinal obstruction is clearly established.[5]

CASE REPORT

A 6-year-old Egyptian male child with known case of Familial Mediterranean fever (FMF) presented to us with
L, hemoglobin 12.3 g/dL, platelet 350,000, procalcitonin 1.7, and C-reactive protein 48. Serum electrolytes showed hyponatremia with hypokalemia. Abdominal X-ray showed multiple fluid levels suggestive of intestinal obstruction. Ultrasound done showed dilated small bowel loops and no intra-abdominal collection. Computed tomography (CT) abdomen and pelvis showed diffusely distended small bowel loops with multiple fluid levels and mild bowel wall thickening. Disparity of bowel lumen diameter was noted at distal ileum (level of obstruction) with multiple mesenteric adenitis. There were no intra-abdominal collection and no radiological evidence of appendicitis or cholecystitis. Official CT report was that of intestinal obstruction.

Since the clinical condition did not improve over 24 h, he underwent explorative laparoscopy. Intraoperative findings showed three inflammatory adhesive bands between the ileal loops causing intestinal obstruction. One was at the level of the terminal ileum and others are at 10 and 15 cm proximally to the first one. These bands were released laparoscopically. The post-operative course was uneventful.

DISCUSSION

(A) One or more major criteria or 2 minor criteria, (B) 2 or more major criteria or 1 major plus 2 minor criteria, (C) 2 or more criteria.

FMF is a hereditary autosomal recessive autoinflammatory disorder characterized by recurring and self-limiting attack of febrile serosal inflammation.[1] It usually involves the peritoneal, synovial, and the pleural membrane. It is commonly found in Armenians, Turks, Arabs, Balkans, and Jews originating from North African countries.

Representative diagnostic criteria for FMF:[4]

A. Livneh FMF diagnostic criteria
   • Major criteria
     • Peritonitis (generalized)
     • Pleuritis (unilateral) or pericarditis
     • Monoarthritis (hip, knee, and ankle)
     • Fever alone
     • Minor criteria
     • Incomplete attacks affecting one or more sites
     • Abdomen
     • Chest
     • Joint
     • External leg pain
     • Response to colchicine.

B. Sohar criteria
   • Major criteria
     • Recurrent febrile episodes with serositis
     • Amyloidosis of amyloid A type without predisposes disease
     • Response to colchicine


- Minor criteria
- Recurrent febrile episodes
- Erysipeloid-like erythema
- Family history in the first-degree relative.

C. Yalcinkaya criteria for pediatric patients

Fever axillary temperature 38°C duration 6–72 h and more than three attacks:
- Abdominal pain duration 6–72 h and three attacks
- Chest pain duration 6–72 h and more than three attacks
- Arthritis duration 6–72 h attacks and oligoarthritis
- Family history of FMF.

REFERENCES
