Small Bowel Obstruction by a Carcinoid Tumor in a Patient with Familial Mediterranean Fever

Isack Gabizon MD[1], Hili Giladi MD[1], Amnon Ovnat MD[2], Oleg Dukhno MD[2], Daniel Flusser MD[1,3] and Shaul Sukenik MD[1,3]

Departments of [1]Internal Medicine D and [2]Surgery B, and [3]Rheumatology Unit, Soroka University Medical Center and Faculty of Health Sciences, Ben-Gurion University of the Negev, Beer Sheva, Israel

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Acute small bowel obstruction is relatively rare in patients suffering from familial Mediterranean fever. Most cases occur in childhood and are due to adhesion formation.

We report a unique case of acute small bowel obstruction in an FMF patient that was caused by a carcinoid tumor of the small intestine. There was a significant delay in reaching the correct diagnosis because the symptoms of acute bowel obstruction were mistaken for those of classical abdominal FMF. To our knowledge this is the first reported case of coexisting FMF and carcinoid tumor.

Patient Description

A 55 year old white Israeli woman, born in Egypt, was diagnosed in 1960 at the age of 10 as suffering from FMF. The diagnosis was based on recurrent attacks of abdominal pain and fever, which began at the age of 9. Typical attacks lasted for 24–72 hours and recurred at 1–2 month intervals. Some episodes were associated with left ankle pain and swelling. The clinical manifestations of each episode were high fever and diffuse peritoneal irritation. Associated laboratory findings included an elevated sedimentation rate, leukocytosis, and high serum fibrinogen levels.

At the age of 22 the patient married and subsequently gave birth to three boys over the next 10 years. Colchicine therapy, which was initiated in 1976 when she was 26, reduced the frequency and severity of the attacks to one every few months. During pregnancy she was advised to discontinue colchicine therapy.

The patient was relatively well until January 2004 when she started to complain of diffuse and persistent abdominal pain without fever or nausea. There was no change in her bowel habits. She insisted that her pain was completely different from the pain she suffered during attacks of FMF. A laparoscopic cholecystectomy was performed, but her symptoms persisted.

Six months later, in July 2004, she was admitted to the Internal Medicine department because of abdominal pain, nausea and vomiting, and severe watery diarrhea, which started one week prior to admission. On admission her temperature was normal. Physical examination revealed a soft abdomen with no sign of distension or peritoneal irritation. Auscultation of the abdomen was normal and the liver and spleen were not palpable. The rectal examination was normal.

Routine laboratory tests, including complete blood count, kidney and liver function tests, and urinalysis, were all normal. Repeated stool and urine cultures were sterile. A gynecologic examination was normal. Plain abdominal radiography and abdominal ultrasonography were normal. The patient refused to undergo a sigmoidoscopy. The dose of colchicine was increased from 1 to 2 mg/day and the diarrhea, vomiting and abdominal pain subsided gradually. She was discharged from the hospital 6 days after admission.

On the same day that she was discharged from hospital the patient returned to the emergency room because her abdominal pain recurred and she began to suffer from massive vomiting. On readmission she was afebrile, her pulse was 120 beats/minute with a regular rhythm, and blood pressure was 110/70 mmHg. The physical examination, including examination of the abdomen, was normal. A plain abdominal film was normal. A few hours after admission the patient's condition began to deteriorate. Her abdomen became distended with increased metallic peristalsis sounds. The leukocyte count increased to 16,620/mm^3 from 8,720/mm^3 a few hours earlier, with a marked shift to the left. At this point a repeat abdominal plain film revealed dilated small intestinal loops with air-fluid levels and no air in the large intestine, compatible with a small bowel obstruction. Abdominal ultrasonograpy was attempted but could not be completed because of severe recurrent vomiting.

An emergency laparotomy was done that same day, which revealed a large amount of ascitic fluid and an obstructing intraluminal mass at the ileocecal junction. A right hemicolectomy and terminal ileectomy were performed. The pathologic diagnosis was an insular-type, low grade, carcinoid tumor that invaded all layers of the intestinal wall and penetrated into the fatty and perineural tissue. There was no evidence of tumor tissue at the surgical margins, or in the regional lymphatics.

The patient recovered from surgery without complication and did not require chemotherapy. At 1 year follow-up after surgery she is doing well except for her FMF attacks, which occur every few months. Genetic testing revealed the patient to be homozygotic for the M694V mutation.

FMF = familial Mediterranean fever

Comment

Mor et al. [1] published a detailed and extensive review of the gastrointestinal manifestations of FMF. Abdominal pain is by far the most frequent symptom, occurring in almost all patients during at least some of their attacks. Diarrhea associated with abdominal pain, the chief complaint in our patient, has been described in only about 20% of FMF patients during attacks [1]. Nausea and vomiting, which also were prominent in our patient, are even less common in FMF.

Evidence of acute peritonitis is the most common finding on physical examination during an FMF attack. The differential diagnosis of acute peritonitis is extensive, and other common entities such as acute appendicitis, acute cholecystitis, acute pancreatitis, acute diverticulitis, and pelvic inflammatory disease must always be considered.

Inflammatory bowel disease is more common and severe in non-Ashkenazi* Jewish patients with FMF [2] and can also cause peritoneal irritation. Crohn's disease was identified as relatively prevalent in FMF patients [3].

Special consideration should be given to vasculitides such as Henoch-Schönlein purpura, polyarteritis nodosa and Behcet's disease, which can induce severe abdominal pain, nausea, vomiting, diarrhea, and even fever. Complications such as bleeding, bowel infarction and perforation can cause diffuse peritonitis.

Amyloidosis of the gastrointestinal tract can cause intractable diarrhea and severe malnutrition. Rare cases of ischemic colitis and perforation or obstruction of the large intestine due to amyloid deposition have been reported.

Although signs and symptoms of acute peritonitis are the most common finding during an FMF attack, intestinal obstruction is rare. Still, the possibility of its occurrence should not be underestimated. Adhesions, which form as a result of recurrent sterile exudation into the peritoneal cavity, are usually the cause of small bowel obstruction. This complication is more prevalent in children, and has been reported to occur in patients treated with colchicine.

There are no reports in the literature to date of a tumor-induced obstruction of the small bowel in FMF. Carcinoid tumors, while rare, are the most common gastrointestinal neuroendocrine tumor. They can arise in any part of the gastrointestinal tract. Those that occur in the small intestine are commonly located in the ileum within 60 cm of the ileocecal valve [4]. The majority of patients are asymptomatic and the tumor is an incidental finding. Symptoms of the carcinoid syndrome, such as diarrhea and flushing, are infrequent, occurring in less than 10% of patients with small bowel carcinoid. They usually start in the fifth or sixth decade of life. The most prevalent initial symptom, vague abdominal pain, as seen in our patient, occurs in approximately 40% of the patients. Intermittent obstruction is seen in 25% of patients and may be caused by the intraluminal mass or mesenteric kinking and distortion due to tumor invasion [5].

In the case presented here, the patient insisted that the abdominal pain and nausea, which began in January 2004, were different from the pain that she felt during her typical FMF attacks. In our opinion these symptoms were the initial clinical manifestations of the tumor. The laparoscopic cholecystectomy was unwarranted.

We believe that the coexistence of these two conditions is totally coincidental, and that the delay in the diagnosis of the carcinoid tumor resulted from a misinterpretation of the abdominal symptoms. Physicians should always listen carefully to patients and bear in mind that FMF patients can develop concomitant diseases. This is particularly important to remember when the presenting symptoms are similar to those that are typical of FMF.

* Ashkenazi refers to Jews of East European origin, in contrast to Sephardi, which denotes Jews of Middle Eastern or North African descent.

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Correspondence: Dr. S. Sukenik, Dept. of Medicine D, Soroka University Medical Center, P.O. Box 151, Beer Sheva 84101, Israel.
Phone: (972-8) 640-0601
Fax: (972-8) 627-2836
email: ssukenik@bgumail.bgu.ac.il