Familial Mediterranean Fever in the Arabian Peninsula : Case Reports and Review of Literature

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ABSTRACT

Familial Mediterranean Fever (FMF) is an inherited disease which manifests with attacks of fever and serosal inflammation, and is complicated in 26% of patients with secondary amyloidosis.

In this paper the author reports a series of five patients, including the first Bahraini with this disease; and reviews the clinical course, pathology, theories of pathogenesis and treatment of this disease.

Familial Mediterranean Fever is a disease characterised by paroxysmal attacks of fever, abdominal, pleuritic or joint pain. The typical attack lasts between 24 and 48 hours and resolves spontaneously, only to recur again after an upredictable time. The disease is often confused with acute abdominal surgical conditions, and therefore, is mistreated as such. FMF was first reported by Siegal in 1945[1]. In this article the author reports the first case from the Arabian Peninsula, summarises her personal experience with this disease between the period 1984–1986 and reviews the literature on the clinical presentation, theories regarding the aetiology and the treatment.

CASE NO. 1

A 37-year old Bahraini female presented with a nine-year history of recurrent attacks of abdominal pain and fever. Her typical attack starts with periumbilical pain which gradually spreads to involve the whole abdomen. The pain is always associated with fever and vomiting. The symptoms last for two to three days and recur about twice a month. She has undergone repeated investigations for her symptoms with barium enemas, upper GI series, ultrasonography of the abdomen and serum amylase determinations which were not diagnostic. She was advised appendectomy during these attacks by many physicians but she refused insisting that the symptoms would soon resolve. Her parents are first cousins; the family is originally from Saudi Arabia. There was no family history of a similar condition. Her physical examination during a typical attack showed T = 38.4°C. She appeared ill and in distress due to abdominal pain. The abdomen was slightly distended with diminished bowel sounds. Diffuse tenderness rebound and guarding were elicited upon palpation of the abdomen.

Flat and upright films of the abdomen showed dilated small bowel loops with air-fluid levels. The patient was observed in the hospital and had total resolution of her fever and abdominal pain and tenderness in 24 hours. The diagnosis of FMF was suspected. Two weeks later when the patient was free of symptoms metaraminol provocative test was carried out according to the method of Barakat et al.[2] Three hours after the infusion of 10 mg of metaraminol in 500 cc normal saline, the patient had reproduction of her typical attack. She was diagnosed as having FMF and was started on colchicine 0.5 mg orally three times daily. She has been followed up for one year with only one recurrence of her attack.

CASE NO. 2

A 28-year old female from Alexandria, Egypt, presented with history of recurrent attacks of abdominal pain, pleuritic chest pain, pain of both knees, skin rash and fever since the age of 9 years. The attack usually starts suddenly with pain in the right lower quadrant of the abdomen which then spreads

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to the whole abdomen. Pleuritic chest pain accompanies the abdominal pain. Some attacks were associated with pain and swelling of the knees and an erythematous skin rash over the lower extremities. All attacks were associated with fever. The patient was admitted repeatedly to hospitals for investigations and during two hospitalisations she was operated on. The first operation, at the age of 12 years, was an appendectomy and the second one was a right hemicolectomy at the age of 22 years, for what she was told was bowel obstruction. The abdominal pain remained unchanged after these surgeries, and in addition diarrhoea developed after the right hemicolectomy. The attacks lasted three days and recurred three to four times per month. She denied family history of a similar illness and her parents are not related.

Physical examination during an attack revealed T = 38.4°C. She appeared ill and in distress due to abdominal pain and left pleuritic pain. There was a left pleural friction rub anteriorly. Abdominal examination showed scars of her previous surgeries, the bowel sounds were diminished. Upon palpation of the abdomen rigidity, direct and rebound tenderness was elicited all over, especially over the right lower quadrant.

Films of the abdomen revealed distension of the small and large bowel loops. A chest film revealed a small left pleural effusion. The patient was diagnosed as having FMF and she was placed on colchicine 0.5 mg three times daily. She had three recurrences in the past year while on colchicine.

CASE NO. 3

A 32-year old male Palestinian Arab presented with a twelve-year history of attacks of fever, abdominal pain, and pain of the hip and knee joints. The abdominal pain would usually start in the right lower quadrant and spread to the whole abdomen. Fever preceded his abdominal pain and symptoms lasted one to three days. He had one to two attacks per month. His elder brother had identical symptoms, his parents are first cousins.

Physical examination during an attack revealed T = 38.8°C. He appeared ill and distressed due to abdominal pain. The abdomen was slightly distended with diminished bowel sounds. Rigidity, direct and rebound tenderness were elicited upon palpation of the abdomen.

Films of the abdomen showed distended small and large bowel. The fever and the physical examination findings subsided in 48 hours. Two weeks later the patient underwent metaraminol provocative test with precipitation of his attack seven hours after metaraminol infusion. He has been on treatment with calchicine 0.5 mg three times daily and has had no recurrence of attacks after one year of follow-up.

CASE NO. 4

The 37-year old brother of case No. 3 had identical history of 15 years duration and had an appendectomy during one of his attacks. This patient refused to have metaraminol challenge but accepted being treated with colchicine. He has been on 0.5 mg of colchicine three times daily with one recurrence of attack during ten months of follow-up

CASE NO. 5

A 68-year old Palestinian Arab male presented with nocturnal diarrhoea, leg weakness and postural hypotension of eighteen months duration. He gave a history of recurrent attacks of right hip pain associated with fever and abdominal pain since the age of nineteen years. He stated that these attacks occurred about three times per month and lasted three to four days. He had many investigations for his attacks and no diagnosis was reached.

Physical examination revealed supine BP = 100/60 mm Hg, erect BP = 70/50 mm Hg. Supine pulse = 70/minute, erect pulse 70/minute. The patient appeared pale, emaciated and could not stand without support. His abdominal examination was normal but he had decreased anal sphincter tone. Neurological examination revealed bilateral foot drop, severe muscle wasting, absent deep tendon reflexes and absent vibration, pin-prick and deep pain sensations below the knee. Autonomic testing using ECG monitoring revealed absence of autonomic response during postural changes, valsulva or hand-grip manoeuvres.

Colonoscopic rectal biopsy showed the presence of amyloid deposition. The patient was diagnosed as having amloidosis secondary to FMF. He was treated with colchicine 0.5 mg three times a day with