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Case Report

Delay in diagnosis of intestinal obstruction in a 17 year-old female with Familial Mediterranean fever: The first case report from Syria

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ABSTRACT

Familial Mediterranean fever (FMF) is an autosomal recessive auto-inflammatory disorder characterized mainly by brief recurrent episodes of peritonitis, pleuritis, and arthritis, usually with accompanying fever. Almost all patients with FMF experience abdominal episodes. Abdominal pain develops, and may progress to peritonitis. Intestinal obstruction secondary to adhesions may be observed in FMF patients but this is the first case from Syria.

A 17-year-old Syrian female patient presented to our hospital complaining of abdominal pain, frequent vomiting, weight loss and absolute constipation in the past ten days, with a confirmation of her infecting by FMF 3 years ago. The obstruction was treated conservatively and after 6 months we had to treat the obstruction by laparoscopic releasing of abdominal bands as a result of recurrence.

The patient was discharged and followed up for 6 months with excellent results. We herein report the first known case of FMF with small bowel obstruction in Syria with delayed in diagnosis. Physicians should be alert to this possible complication when FMF patients arrive at the emergency room.

1. Introduction

Familial Mediterranean fever (FMF) is a hereditary auto-inflammatory disorder characterized by recurrent bouts of fever and serosal inflammation. The geographic prevalence is most common in Turkish, Armenian, North African, Jewish, and Arab descent [1]. It has also been reported at a lower prevalence in many other populations such as in Greece, Italy, Japan, and China [2]. FMF is usually considered an autosomal recessive disease that results of mutations in the MEFV (Mediterranean Fever) gene, consisting of 10 exons located on chromosome 16p13.3. MEFVencodes a 781 amino acid (86kDa) protein (pyrin or marenostrin) which effectively regulates the transcription of intranuclear peptides involved in inflammation [3]. In 90% of FMF patients, the first attack occurs before 20 years of age [4]and comprises recurrent sudden episodes of fever, severe pain (due to serositis at one or more sites with peritonitis, pleuritis, and synovitis), lasting 1–3 days and resolving spontaneously. The diagnosis of FMF is clinically and supported by ethnic origin and family history. The genetic testing is just to confirm the diagnosis and exclude another diseases that mimic FMF. As long-term complication is secondary amyloidosis which is a major cause of morbidity [5], Another complications are small bowel obstruction thats results for recurrent peritonitis formation intra-abdominal adhesions. Delay in diagnosis and treatment is expected. Preventing acute attacks and minimize subclinical inflammation in between attacks, and to prevent the development and progression of amyloidosis is the target of therapy for familial Mediterranean fever. The cornerstone is colchicine which needs observation its therapeutic response for six months. In colchicine resistance cases,IL-1 inhibitors is the choice.

This case report has been reported in line with the SCARE criteria 2020 [9].

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2. Case presentation

A 17-year-old girl presented at the emergency department with crampy abdominal pain, vomiting and obstipation from 10 days. Her past medical history was only remarkable for Familial Mediterranean Fever (FMF). She was diagnosed with FMF 3 years ago without genetic testing, only with clinical findings. And it was managed with colchicine.

Abdominal examination reveled tenderness in the left hypochondriac region without rebound tenderness. Apart from that the abdomen was not distended. Bowel sounds were hypoactive. The rectum was empty in rectal examination.

Laboratory results were as follow: full blood count normal, electrolytes normal, urea and creatinine normal only c-reactive protein was 12. The abdominal x-ray showed small bowel air-fluid levels in the left side of the abdomen (Fig. 1). Provisional diagnosis of bowel obstruction was made and the patient was managed conservatively with IV fluids and nasogastric tube insertion. Our patient's condition improved completely and discharged later with complete remission.

After six months our patient presented once again with the same past compliant, which was managed conservatively by other doctors. But her condition continued to deteriorate, then she presented at our emergency department. Where the diagnosis of bowel obstruction was confirmed. An abdominal laparoscopic procedure was performed for releasing adhesions, she underwent laparotomy. On exploration, There were intra-abdominal adhesions in between liver and peritoneum. There were bands secondary to adhesions in the jejunum which we released them (Fig. 2), in addition to performing elective appendectomy to prevent recurrence (Fig. 3).

Eventually our patient had successive follow up until the time of discharge. The follow up has continued for 1 year and the results of our intervention were so satisfying.

3. Discussion

Both familial Mediterranean fever and bowel obstruction are mimic acute abdominal condition. Familial Mediterranean fever (FMF) is an autosomal recessive auto-inflammatory disorder and it leads to serositis such as, peritoneum, pleura and synovial bursa of joint. It is a rare disorder of the world and it is very common in people of Sephardic (non-Ashkenazi) Jewish, Armenian, Arab and Turkish heritage. The incidence rate in the Mediterranean region is 1 in 200 of the general population [6].It was estimated in the Japan about 500 patient and recently the number of patient is increased [7].FMF is distinguished by relatively short attack fever that range from 1 to 3 days associated with serositis, synovitis or skin rash. In some patients, attacks begin in infancy or very early childhood, but 80–90% of attacks occurs at age 20 and the interval between attacks ranges from days to years. The symptoms of acute attack are Nausea, vomiting and abdominal pain and are similar to intestinal obstruction. Tunca M et al., had been reported that appendectomy forms the most common surgical procedure for 19% of patients with FMF [8].In both cases of intestinal obstruction and Mediterranean fever, the sedimentation rate and C-reactive protein are increased. Mediterranean fever is distinguished from bowel obstruction in that it subsides within 12–24 hours, while bowel obstruction deteriorates. Peritonitis is a common symptom of the disease and recurrent episodes may lead to peritoneal adhesions. This increase the risk of intestinal obstruction. Intra-abdominal adhesions secondary to recurrent peritonitis are common in FMF, but comparing this case to the previous cases in the medical literature, the delayed in diagnosis of intestinal obstruction is a extreme serious and rare case. Although the patient was taking colchicine, it did not prevent adhesions from occurring. As a result, the diagnosis of bowel obstruction with Mediterranean fever will be difficult. The importance of this case comes because it is the first case in Syria and because its diagnosis was delayed for 10 days, during this period the patient suffered from weight loss, vomiting and complete constipation. A plain abdominal X-ray was performed and it was managed it by conservative treatment of bowel obstruction. After 6 months obstruction bowel recurred where it was performed plain abdominal x-ray and exploratory laparotomy. It was detected adhesion and intestinal obstruction. Laparoscopic treatment was chosen to remove the adhesions and obstruction and it was performed elective

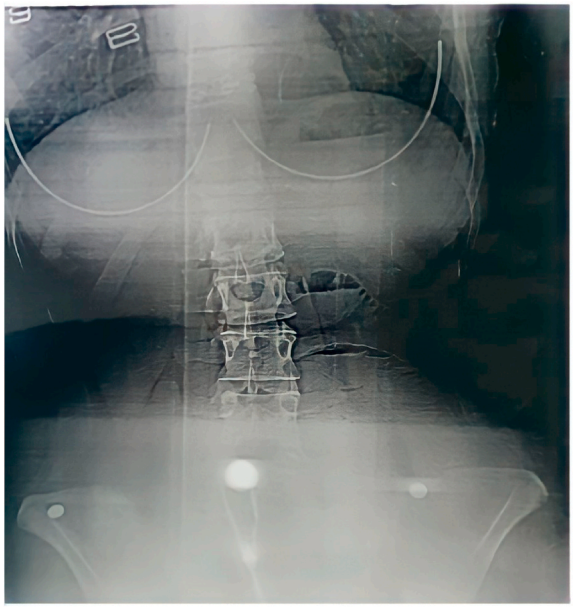

Fig. 1. The abdominal x-ray showed small bowel air-fluid levels in the left side of the abdomen.

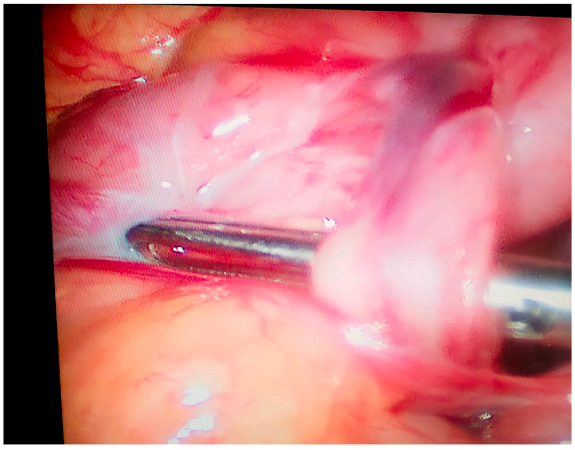

Fig. 2. Laparoscopic release abdominal bands.

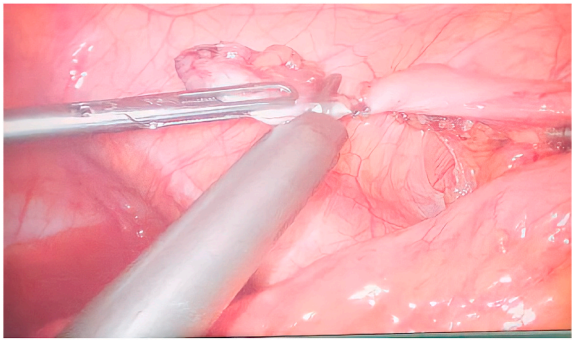

Fig. 3. The picture during performing elective appendectomy.

appendectomy during operation. The case has been followed up for one year and the patient's condition was good without any complication.

4. Conclusion

The combination of familial Mediterranean fever with intestinal obstruction is a reality in a few cases, so doctors, especially Syrians (because it is the first case in Syria), must follow up on Mediterranean fever patients when there are symptoms of constipation and abdominal pain, and carefully investigate the presence of intestinal obstruction Diagnostic because it is considered an emergency situation that requires immediate treatment, and most importantly, avoiding delays in diagnosing obstruction.

Ethical approval

This case reports didn't require review by Ethics committee, Aleppo university hospital, Aleppo university, Faculty of medicine, Aleppo-Syria.

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Author contribution

Kusay Ayoub: contributed in study concept and design, data collection. Samir kanjo:contributed i writing the paper. Sarya Swed:contributed in writing the paper. Yamane Chawa:contributed in writing the paper. Mahmoud Alhamadeh Alswij:contributed in writing the paper. Nihad Mahli:contributed in reviewing the paper.

Consent for publication

Written informed consent was obtained from the patient for publication of these two case reports and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Registration of research studies

Not applicable.

Guarantor

Kusay Ayoub.

Declaration of competing interest

All authors declared no conflict of interest.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2021.103011.

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