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Case Report

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Congenital Trans Mesenteric Hernia (CTMH) in a Female Patient with Familial Mediterranean fever (FMF): Case Report and Review of the Literature

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Abstract

Congenital Trans mesenteric Hernia (CTMH) is a rare entity of internal hernia that occur as a congenital defect in children due to an error in the rotation of the midget, presenting usually with an abdominal pain, fever and bilious vomiting. However, if left untreated, disastrous outcomes such as acute bowel obstruction and strangulation can take place. Its diagnosis is sometimes challenging to physicians and requires exploratory laparotomy to confirm the presence of a hernia defect. CT scan prior to surgery is seldom needed and may not have a benefit to guide for the exact diagnosis. For treatment, surgical repair, either open or laparoscopic, is the procedure of choice to manage such a condition. We report a case of CTMH in a 3.5–year-old female patient who presented to our hospital with abdominal pain and inconsolable crying.

Keywords: Internal Hernia, Para duodenal Hernia, Congenital Defect, Small Bowel Obstruction, Surgical Repair

Introduction

Congenital trans-mesenteric hernia (CTMH); a herniation of viscera through the mesentery, is a rare type of hernia in which the mesenteric defect is usually 2-3 cm in dimeter and infrequently seen in newborns. Infants with CTMH usually present with intestinal obstruction. However, this rare entity is not considered in the differential diagnosis of bowel obstruction [1, 2].

Furthermore, it represents approximately 5-10 % of all types of internal hernias in children and it are sometimes associated with high morbidity and mortality. The diagnosis of such infants who usually have non-specific symptoms is a bit challenging [2-4].

The initial diagnosis of CTMH usually resembles that of acute intestinal obstructions and a delay in diagnosing this serious condition may increase mortality. The definitive diagnosis of CTMH is basically made during surgery [5, 6]. Computed Tomography (CT) scan is frequently used and can be useful for diagnosis, but an accurate pre-operative diagnosis is rarely made [7].

In regard to treatment, surgical options; either open or laparoscopic, are considered the cornerstone for repair of the hernia defect [8].

Case Report

A 3.5 years old, female child presented to our clinic; at Pediatric University Hospital, Damascus. The child was doing well until age of eight months old, when she started to complain of a frequent abdominal pain, bilious vomiting a once, inconsolable crying, and constipation.

At that time, she sought a medical advice and variety of laboratory tests, Ultrasound (US) and stand abdominal X-ray (AXR) were performed and were free of any clear pathology. Thereafter, the symptoms got improved with only some analgesics. In the next day, the patient came back with inconsolable crying and fever (38.5°C, rectal) and thus was admitted to Pediatric Hospital for further investigations. There was no history of weight loss, anorexia, or surgical history. She was not taking any drugs with no drug allergies yet. The physical examination revealed tenderness in all abdominal areas. Pertinent laboratory values included Complete Blood Count (CBC), urinalysis and kidney function tests, liver function tests, electrolytic and C - reactive protein, amylase and CSF (Cerebrospinal Fluid) were normal. The upper gastrointestinal endoscopy was done and was normal as well.

During the last two years, the child continued complaining of an abdominal pain, constipation (treated by Glycerin Suppositories) and frequent bilious vomiting. The parents stated that their child took some positions in an attempt to relieve the pain such as sleeping in a sitting position. At that period of time, DNA analysis (Northen blotting) and examining a possibility of (FMF) mutation was required and Familial Mediterranean Fever was identified (treated by Colchicine).

On October 22nd 2017, the 3.5 years old child came again to the hospital with abdominal pain and bilious vomiting despite many medications she was taking. A physical examination was unremarkable except for a mass in the left area of the abdomen which raised suspicions towards Intussusception. Therefore, we repeated the laboratory testing, radiological examination (Ultrasound; fecalith in colon, AXR; was normal) and rectal water soluble enema (Gastrografin); megacolon and material evacuated slowly. A pediatric surgeon

finally decided for the patient to undergo an exploratory laparotomy. However, an internal hernia in the mesenteric colon was identified.

Intraoperative, a large hernia defect in the mesenteric colon containing small bowel was recognized, this defect was located at the left upper area of the abdomen posterior to the large bowel with evident adhesions [Figure 1]. Repair of the hernia and placing the small intestine in their original place was successfully done. Follow-up of the patient showed remarkable improvement with no further complaints [Figure 2].

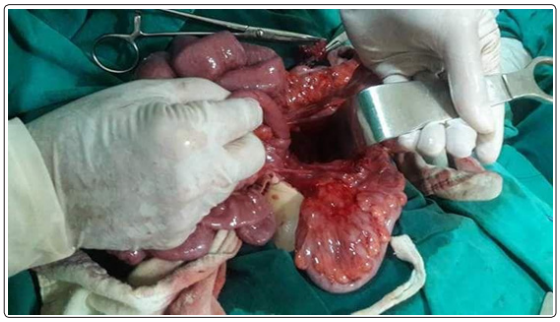

Figure 1: Large defect in mesenteric colon was containing small bowel which pushed back in

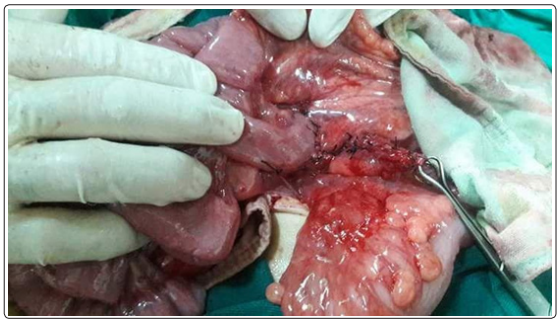

Figure 2: Repair of the hernia done with sutures (stitches)

Discussion

Congenital trans-mesenteric hernia (CTMH) is a protrusion of the viscera through defect between transverse colon and descending colon; the left upper area of the abdominal cavity; as in our condition. It mainly affects children; caused by congenital mechanisms such as error in the rotation of the midgut, or in adults; after surgery, inflammation and trauma [8, 9].

Although internal hernia is not a common condition in the Middle East, it represents 5-10 % of all causes of internal hernia in children; it is quite uncommon in western areas either [2].

Following the embryonic formation of an intestinal loop, congenital defect occurs in areas of mesentery that are thin or non-vascular [10].

Preoperative diagnosis is difficult because clinical symptoms may range from simple complaints like bilious vomiting to catastrophic presentations like acute-onset obstruction [11]. The vast majority of conditions present with epigastria discomfort, periumbilical pain, abdominal distention, bile vomiting; and infrequently symptoms of bowel obstruction [1, 11]. Usually, hernia types are named according to location, but paraduodenal hernia is classified as the most common type of internal hernias [7]. However, diagnosis of splenic flexure hernia preoperative is difficult and there could be a need for a CT scan or an exploratory laparotomy [12].

It has been reported that the diagnosis of meso-colic hernia including: 1. loop of small bowel contained in the hernia sac. 2. Abnormal location of small intestine support that can be seen though a CT scan. However, in our case we did not conduct CT as it was not available. So, we had to do an exploratory laparotomy [7].

In this case, the condition imitated Intussusception or Ischemia in colon and so exploratory laparotomy was required [10].

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