



An unusual cause of duodenal obstruction: Mesenteric fibromatosis in a patient with type I Mayer-Rokitansky-Kuster-Hauser syndrome

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ABSTRACT

Patients with mesenteric fibromatosis (MF) are clinically asymptomatic, with little or no focal symptoms until later in their course, at which time they complain of pain, abdominal discomfort, constipation, vomiting, abdominal mass, weight loss, and symptoms due to organ compression. Generally, it occurs as an abdominal mass but may also present in many different ways. In some cases, trauma, previous abdominal surgery, and hormonal stimulation (such as estrogen) may play a role in onset of this neoplasm. Patients with Mayer-Rokitansky-Kuster-Hauser syndrome present primary amenorrhea and may have some other anomalies, including hearing defects, heart defects, skeletal deformities, and genital neoplastic diseases. We diagnosed duodenal obstruction due to MF in a patient with type I Mayer-Rokitansky-Kuster-Hauser syndrome.

Keywords: Mesenteric fibromatosis, duodenal obstruction, metabolic alkalosis, Mayer-Rokitansky-Kuster-Hauser syndrome

INTRODUCTION

Mesenteric fibromatosis (MF) is a rare benign tumor of the abdominal cavity. MF usually presents as an abdominal mass, pain, or discomfort but can present in several other ways as well, including acute abdomen, bowel obstruction, bowel perforation, irreducible inguinal hernia, fistula, and obstructive jaundice (1-3). We diagnosed MF in a woman with Mayer-Rokitansky-Küster-Hauser syndrome who complained of recurrent severe hypokalemia and metabolic alkalosis due to duodenal obstruction.

CASE PRESENTATION

A 29-year-old female patient was admitted to our outpatient clinic with a complaint of gradually increased nausea and vomiting, abdominal pain, and severe weight

loss (about 32%) for 6 months, particularly for 10 days. She had no previous upper gastrointestinal symptoms until 6 months ago. The patient had a history of diagnosis of type I (isolated) Mayer-Rokitansky-Küster-Hauser syndrome in 2004. Surgical creation of a neovagina had been performed in the patient in 2009. She had been treated by estrogen preparation for 1 month in 2004. She was not taking any other medications, including laxatives, diuretics, or any drugs and alcohol.

On admission, she was severely cachectic and dehydrated. Body temperature: 36.7°C, systolic blood pressure: 90 mm Hg, diastolic blood pressure: 55 mm Hg, pulse rate: 115/minute, breath rate: 15/minute. Skin turgor and tonus were declined, along with dry tongue. We detected severe upper abdominal distention, and no bowel

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sounds were detected. The rectum was empty. She had a normal woman phenotype and had normal pubic hair. Signs of hyperandrogenism were not found.

Investigations showed glucose: 78 mg/dL, serum creatinine: 1.78 mg/dL, potassium: 2.6 mEq/L, sodium: 122 mEq/L, calcium: 9.5 mg/dL, magnesium: 0.92 mg/dL, and chloride: 81 mEq/L. Prothrombin time with an international normalized ratio was 2.1. Viral markers were negative. Complete blood count was normal. Spot urine analysis revealed 14 leukocytes and ketone positivity (+++). Thyroid hormone levels were normal. Arterial pH was 7.64, pCO₂ was 48 mm Hg, and HCO₃⁻ was 44 mEq/L. Her karyotype was 46, XX.

Diffuse areas of gas on plain abdominal radiographs were detected (Figure 1). Ultrasonography (Aplio-Toshiba, Tokyo, Japan) revealed a rudimentary uterus, cervix agenesis, and normal ovaries. There were dilatations in the stomach and duodenal segments. Esophagogastroduodenoscopy (Model GIF c-Q 20 Olympus Optical, Tokyo, Japan) showed stasis in the stomach and the first part of the duodenum, with normal mucosal findings, and luminal narrowing at the second part of the duodenum. Computed tomography (CT) (Siemens Medical Systems, Erlangen, Germany) of the abdomen showed characteristic dilatation on the stomach and duodenum (Figure 2). Colonoscopy (Pentax EC 3840 L video-colonoscopy, Japan) was normal. Cervix and upper vaginal aplasia was also detected.

The patient was treated with intravenous saline, vitamin K, frozen fresh plasma, and parenteral nutrition. The patient recovered modestly 72 hours after medical treatment. Then, exploratory laparotomy was performed. An obstruction region was detected at the level of the third part of the duodenum, and severe dilatation was observed in the stomach and duodenal segments before the obstruction. A solid mass with dimensions of 5x4x3 cm was identified at the root of the jejunal mesentery, infiltrating the aorta and small bowel wall. The mass, along with an ischemic segment of small bowel, was totally excised.

Macroscopic examination of the specimen revealed a 5.5x4.5x2.8-cm, firm, and tan-gray serosal mass with irregular margins, which was infiltrating the adipose tissue and bowel wall but not related to the mucosa. Microscopic examination showed a neoplasm with hypocellular appearance, consisting of spindle cells in long fascicles that were spreading to the muscularis propria of the small bowel. Tumor cells showed infiltration to the adipose tissue, had pale eosinophilic cytoplasm, and were embedded in a collagen network interrupted by fibrotic sections, but mitosis was not seen (Figure 3). Masson's trichrome staining revealed a large amount of green fibers compatible with collagen (Figure 4). Immunohistochemical analysis showed that the tumor cells expressed vimentin and actin (Figure 5) but not S100, desmin, CD34, CD117, estrogen,



Figure 1. Diffuse gases on plain abdominal radiographs (300x298)

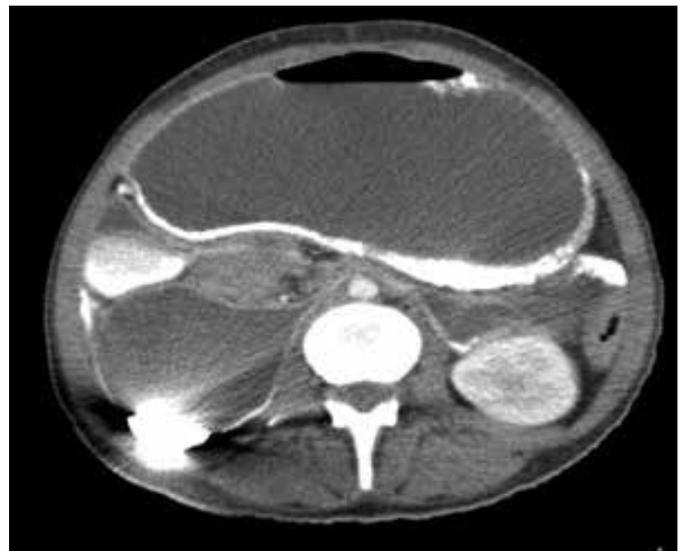


Figure 2. Characteristic dilatation in stomach and duodenum

and progesterone. Microscopic findings and immunohistochemical features were suggestive of MF.

Our patient completely recovered and was discharged after 10 days of the operation. Follow-up visits at 3 and 8 months after the surgery revealed normal findings.

DISCUSSION

The fibromatosis can affect both superficial and deep parts of the body. Superficial fibromatosis involves the face and neck (fibromatosis coli), palms (Dupuytren's contracture), feet (Ledderhose's disease), penis (Peyronie's disease), shoulder, thigh, buttock, and trunk (4). MF is a very rare tumor of the abdomen. MF can be observed in the abdominal cavity with a proportion of 5-10%, particularly in the mesenteric and retroperitoneal region

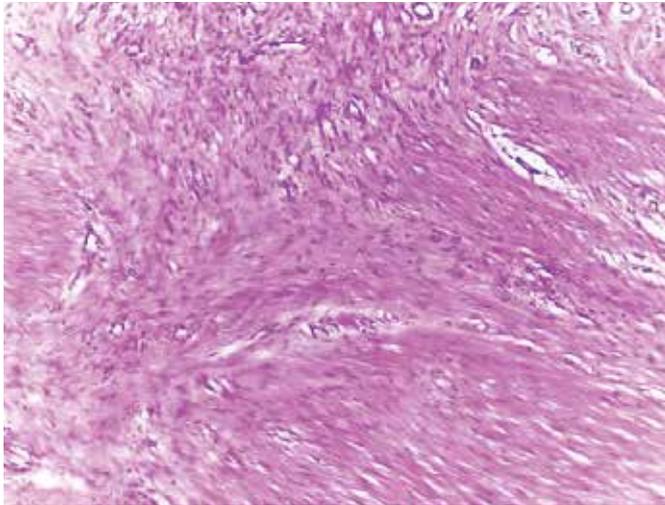


Figure 3. Spindle cells growing in fascicles, with eosinophilic cytoplasm and plump nuclei (HE, x100)

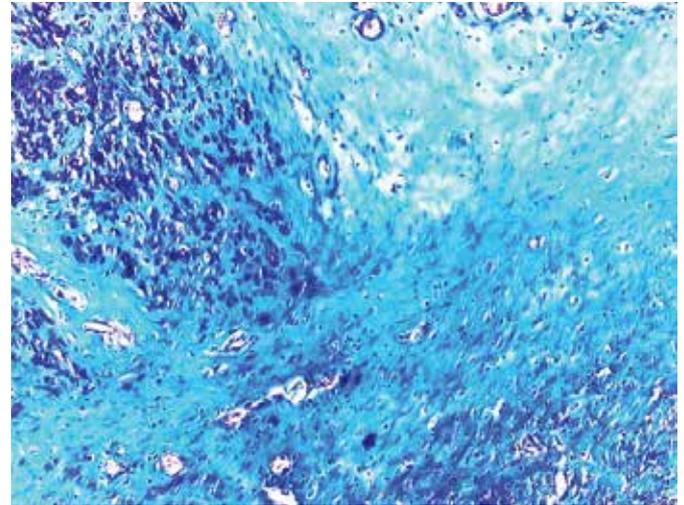


Figure 4. The bundle of collagen fibers stained green (Masson's trichrome, x100)

(5). It usually presents as an abdominal mass, intestinal obstruction, pain, or discomfort (1). Only three MF patients with a duodenum obstruction have been reported up to now (6-8). The patient had been followed empirically before its clinical status started to get serious. Before admission, the patient's clinical status was attributed to duodenal ulcer or acute gastritis.

Mesenteric fibromatosis is also known as deep fibromatosis, aggressive fibromatosis, and desmoid tumors. It usually has a benign character; however, it can present as a large mass or infiltrate surrounding structures (9). The etiopathogenesis of MF is unknown; various factors have been implicated by different authors. Trauma, especially operative trauma, may contribute to the formation of these tumors (10). The operation area and the region where MF was detected were not associated with each other. Growth of these tumors is slowest in young girls and reaches a peak at menopause, suggesting that estrogen may act as a growth factor. Further, estradiol receptors have been demonstrated in tumor tissue in amounts higher than control tissue (11). It is not possible to say whether the estrogen use for 1 month, which was approximately 8 years ago, was important or not in the development of MF. On the other hand, we could not find estrogen or progesterin receptors in the tumor tissue.

In some familial Mayer-Rokitansky-Kuster Hauser cases, heart, skeletal, and earing problems have been reported. On the other hand, neoplastic problems have been reported to be sporadic in Mayer-Rokitansky-Kuster Hauser syndrome, mostly originated from genital organs, especially in prepubertal age, which is interesting (ovarian teratoma, endometrial sinus tumor, ovarian yolk-sac tumor) (12). Our patient is the first Mayer-Rokitansky-Kuster-Hauser case ever reported to date with the diagnosis of MF.

Our patient is the first Mayer Mayer-Rokitansky-Kuster-Hauser case ever reported with the diagnosis of MF to date. It must be

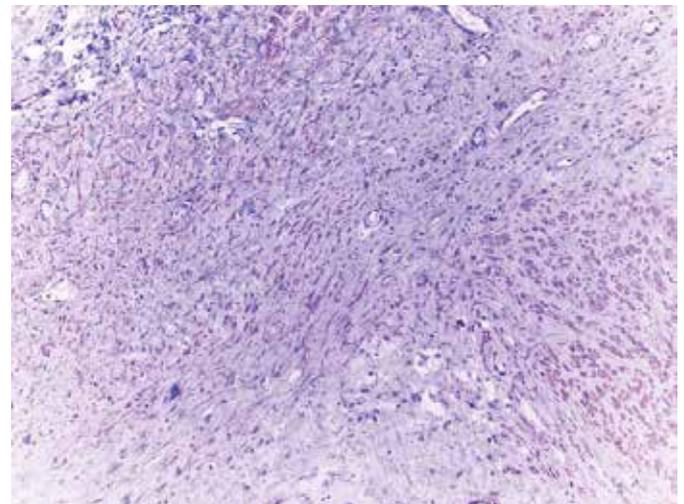


Figure 5. Immunohistochemical stain for smooth muscle actin (x100)

kept in mind that subjects with this syndrome may suffer from intraabdominal neoplastic problems other than in the genital region. MF possibility should be considered when evaluating the differential diagnosis of patients with duodenal obstruction.

Ethics Committee Approval: Ethics committee approval was received for this study.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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