Endometrial stromal nodule complicated by abdominal cocoon: Case report and literature review

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To investigate clinical characteristics, pathology, diagnosis and treatment of abdominal cocoon and Endometrial stromal nodule (ESN). Based on the clinical data of our case, domestic and foreign clinical data of ESN and abdominal cocoon were reviewed. The main manifestations of ESN included increased menstrual flow and vagina anomalous hemorrhage, or sometimes no obvious symptoms. Ultrasound showed hypoecho in the myometrium. ESN was diagnosed through surgical exploration and treated through excision of the nodule. Main manifestations of abdominal cocoon included repeated intestinal obstruction without clear reasons. X-ray showed a “cauliflower” sign characterized by complete or partial agglomeration of the small intestine at one area that could not be separated after fixation and compression. Abdominal cocoon was diagnosed through surgical exploration. Releasing intestinal adhesion serves the main treatment for abdominal cocoon is to restore the intestine function quickly. Abdominal cocoon and ESN are both rare illness, and their causes are inexplicit. Without specific clinical manifestations and diagnosis approaches, their diagnosis and treatment depend upon surgery.

Key word: Endometrial stromal nodule, abdominal cocoon, treatment.

INTRODUCTION

ESN is a rare benign endometrial mesenchymal tumor. Abdominal cocoon is also a rare clinical abdomen illness characterized by encasement of small intestine in a thick off-white fibrous membrane (Reynders et al., 2009; Qasaimeh et al., 2010). Our department admitted one case of ESN complicated by abdominal cocoon and is presented as follows.

Clinical data

A 45 year-old female patient was admitted to our hospital with main complaints of intermittent lower abdominal pain for 2 years and pain aggravation for 9 h. Two years earlier, she felt lower abdominal pain for several minutes and was relieved after some time. 9 h earlier, she felt left lower abdominal pain with nausea for 2 min at intervals of 10 min. No symptoms such as vomiting, vaginal bleeding, diarrhea, fever and low back pain were reported. The patient stopped exhaustion for 9 h. Abdominal ultrasound in a local hospital showed a mixed mass at 4 cm in diameter in the left appendix and a hypoechoic mass of about 7cm behind the uterus (considered to be fibroids and adenomyosis not excluded). She was initially diagnosed as having ovarian cyst torsion and recommended for treatment in a hospital at a higher level. She had no history of peritonitis and abdominal surgery. She had been pregnant seven times and gave birth to 2 full-term newborns. Physical examination found soft abdomen, middle and lower abdominal tenderness, and rebound tenderness especially around the belly button. No palpable mass, visible intestinal peristalsis or gastric peristaltic waves were reported. Bowel sounds were slightly active but not tinkling. Gynecological examination

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Abbreviations: ESN, Endometrial stromal nodule; cAMP, cyclic adenosine monophosphate; cGMP, cyclic guanosine monophosphate.
showed that the patient had given birth to at least one baby. Hypertrophy of uterus was observed. Anteverted uterus was noted with a similar size to pregnancy for 70 d.

The posterior wall bulged with hard quality and limited movement. There were no palpable masses in the appendix area. Doppler ultrasound in our hospital showed a hypoechoic mass of about 7cm behind the uterus (considered to be fibroids, and adenomyosis not excluded). Abdominal X ray found several visible fluid levels indicative of low intestinal obstruction. Following gastrointestinal decompression, senna medication and rehydration therapy, abdominal pain was relieved and stool and exhaustion were restored. Whole digestive tract imaging was performed to trace the cause of intestinal obstruction and showed slow small intestine movements. Uterine fibroids and intestinal obstruction of unknown origins were diagnosed preoperatively. Laparotomy under general anesthesia was done in joint efforts with the Department of General Surgery. We found that the abdominal cavity was closed by dense membrane-like structure during surgery. After carefully separating the adhesion layer, we found that the small intestine and its mesentery were encased by a 2 to 5 mm thick dense fat fiber which was like cocoon from the Treitz ligament to the ileocecal junction (Figure 1). The encasement was smooth and the peritoneal membrane was thickened with absence of the greater omentum. There was no peritoneal membrane adhesion above the transverse colon. When the membrane-like structure was incised, we noticed folding of the small intestine well vascularized and intestinal adhesions with slow peristalsis and normal color (Figure 2). In addition, we saw sigmoid colon twisting and adhering on the right. The pelvic adhesion was separated to expose the uterus.

The uterus was enlarged and a 6 cm nodule-like protuberance was noticed. The anterior and posterior walls of the uterus adhered to the intestine. The enlarged and prolonged left appendix (hydrosalpinx; and ovary not revealed due to encasement) adhered to the abdominal wall and the right appendix adhered to the right abdominal wall. Hysterectomy, left oophorectomy, and intestinal adhesiolysis were performed. After the intestinal adhesion was separated, the adhesions between intestinal tracts were resected, followed by smear of anti-adhesion drugs. Continuous decompression, enema with senna, and medications of somatostatin, and dexamethasone were administered to suppress digestive fluid secretion and digestive tract edema. The patient was instructed to do limited exercises at bedside. Rehydration was continued. At 4d post operation, the intestinal functional was recovered, and prokinetic drug was given to activate intestinal peristalsis. Postoperative pathology examination showed ESN and uterine sarcoma not excluded (Figure 3). The peritoneal peritoneum was formed by dense collagen fibers with local inflammation. Pathologist consultation considered ESN. Postoperative diagnosis included abdominal cocoon and ESN. The
patient was discharged at 8 days and followed regularly.

DISCUSSION

Endometrial stromal nodule

According to the current World Health Organization classification (2003) for female genital tract tumors, the endometrial stromal tumors were categorized into ESN, low-grade endometrial stromal sarcoma, and undifferentiated endometrial sarcoma (Reynders et al., 2009). ESN is the least common of the endometrial stromal tumors. They are rare neoplasms which are diagnosed in most instances by light microscopy (Fdili et al., 2011). ESN is a benign tumor composed of well-differentiated endometrial stromal cells arranged as a well-circumscribed nodule with smooth, non-invasive margins (Mekni et al., 2004). Nodules may be in the endometrium or the muscles. It is thought to be associated with ovarian endocrine hormone (Tavassoli and Devile, 2003).

Clinical manifestations

Clinically, ESN has clinical manifestations including increase menstrual flow, irregular vaginal hemorrhage and anemia, or sometimes no obvious symptoms. ESN is often misdiagnosed as uterine myoma. Ultrasound often shows hypoechoic uterus (diagnosed as uterine myoma).

In our hospital, the patient had no obvious preoperative symptoms. Ultrasound showed a 6 cm nodule-like protuberance suggestive uterine myoma (glandular myoma not excluded). ESN can hardly be diagnosed preoperatively as its symptoms are not specific. Its diagnosis always depends on postoperative pathology examination. In this case, the patient was misdiagnosed as having uterine myoma prior to surgery, but ESN was confirmed by pathology.

Diagnosis

There are no established diagnosis criteria for ESN as it lacks specific clinical manifestations. Preoperative diagnosis is difficult and the misdiagnosis rate is high. It was previously misdiagnosed as ectopic or interstitial hyperplasia or uterine myoma. Its diagnosis is often dependent on pathology examination. Macroscopic findings show isolated nodule in the intramural uterine muscle. Its boundaries are clear with bulging growth. The section of the nodule compressing the surrounding normal smooth muscle tissue is yellow. Being soft, it lacks the spiral shape structures of uterine myoma. 60% cases of ESN are located in the muscle intramural; 33% in the inner membrane and muscle wall; and 7% only in the inner membrane. In our case, a 6 cm nodule was distinguished from surrounding structures without spiral shape structures at the section. Microscopically, benign tumor cells were distributed evenly, similar to normal proliferation endometrial stroma cells. The cells were
spindle shaped with little cytoplasm and no mitotic figures. Visible small blood vessels resembling endometrial spiral small arteries were distributed among these cells. Tumor cells were diffusely aligned around these small blood vessels. There was no vascular tumor necrosis. Vascular thrombi and hyaline degeneration, clear tumor boundaries, compression interface with neighboring uterine muscular layer were reported. However, muscular infiltration, endometrial and vasculature infiltration were noted (Mekni et al., 2004).

Treatment

ESN is a benign tumor. Resection of the nodule is proper treatment. Some studies, however, consider that ESN has muscular layer infiltration and potentially malignant features. Some patients have metastasis to the lung and brain. Unless the tumor is maturely differentiated and exhibits distinct benign characteristics, its nature and prognosis cannot be determined via biopsies other than comprehensive examination of tumor specimens (Chew and Oliva, 2010). This patient in our case was initially misdiagnosed as uterine myoma. Hysterectomy was performed after communicating with the patient and family members and the outcome was favorable.

Abdominal cocoon

Etiology

There have been multiple theories regarding the etiology of abdominal cocoon. Abdominal cocoon is also called regional fibrotic encapsulation of the small intestine or sclerosing encapsulating peritonitis. The abdominal cocoon is a rare cause of intestinal obstruction. This rare condition, in which the small intestine is encased in a thick fibrous membrane, has been reported predominantly in females (Reynders and Van der Stighelen, 2009). Sahoo et al. (1996) thought that abdominal cocoon could be either primary or secondary. Primary abdominal cocoon has no clear etiology, occurring in patients without a history of abdominal surgery or trauma (Bas et al., 2008). This disorder may coexist with congenital malformations such as the absence of the greater omentum, uterus or appendices, or abnormal development, or the right sigmoid colon torsion. Some researchers consider that it is correlated with embryonic development abnormalities. Others hold that it is the continuum of the greater omentum down the transverse distortion. Meconium obstruction is an autosomal recessive disease with pancreatic cystic fibrosis that can cause exocrine pancreatic insufficiency and gastrointestinal endocrine mucosa disease, leading to fecal block in the middle and lower ileum. The intestinal wall is susceptible to perforation causing meconium peritonitis. Fibrinous exudates may cause intestinal tract adhesions fixed to the posterior abdominal wall.

The intestinal tract encased by fibrous tissues also may form abdominal cocoon (Navani et al., 1995). Secondary abdominal cocoon may be caused by infections, such as retrograde infection. Foo et al. (1978) reported that abdominal cocoon in women was associated with retrograde infection in the female reproductive tract. Retrograde virus infection (such as echovirus, Ke Saji virus or adenovirus) during menstruation causes subacute primary peritonitis. Fibrous exudates encase the small intestine. Our case was complicated by sigmoid colon torsion. As the patient had no history of abdominal
surgery, peritonitis or infertility, the case may be caused by secondary retrograde infection after pregnancy. Secondary abdominal cocoon may result from stimulation of foreign substances, such as abdominal trauma, tuberculosis, nephritis, cancer, heart failure and ascites, leading to multiple serositis with increased fibrinous exudates and fibrous capsule for intestinal tracts (Liu et al., 2009). Medications also contribute to secondary abdominal cocoon. Chemotherapy drugs such as cisplatin, via intraperitoneal injection after diluted by normal saline, can cause chemical peritonitis peritoneal stimulation in addition to killing detached cancer cells and cancer cells of small foci. When the peritoneal exudates are absorbed, they leave behind fibrinous membrane of deposits. β-blockers such as propranolol decrease the cyclic adenosine monophosphate (cAMP) and cyclic guanosine monophosphate (cGMP) ratio that the controls normal cell growth, resulting in overgrowth of collagen fibers and consequent peritoneal fibrosis. Abdominal cocoon has geographic features. It is distributed in the tropical and subtropical regions other than the temperate region. This may be related to the climate and sweating.

Clinical manifestations

Abdominal cocoon is characterized by abdominal masses accompanied by abdominal pain, bloating, nausea, vomiting and other intestinal symptoms. X-ray barium meal examination shows a “cauliflower sign” Part of or the whole small intestine is concentrated in an area and fixed and difficult to separate after compression (Navani et al., 1995). Ultrasound demonstrates that the intestine tract is dilated apparently with peristalsis and masses. CT scan indicates a large bowel shadow (Gurleyik et al., 2010). In our case, the patient had a history of repeated intestinal obstruction without obvious causes and no palpable abdominal masses; and digestive imaging showed slow peristalsis.

Diagnosis

Though clinical manifestations of abdominal cocoon are not specific, they are mainly related to slow intestinal peristalsis due to fibrinous encasement of the small intestine. It presents with recurrent episodes of intestinal obstruction such as bloating, abdominal pain, nausea and vomiting. Physical examination shows bulging belly, visible intestinal peristalsis, bowel sounds hyperthyroidism, and fixed or movable abdominal masses (Wei et al., 2009). Without specific diagnosis indicators, preoperative diagnosis is impossible for abdominal cocoon. It is often identified during surgery. In our case, abdominal cocoon was found during hysterectomy. The small intestine is covered by a gray stiff thickened fibrinous membrane similar to cocoon. Other adjacent organs such as liver, stomach and part of colon are also covered by this membrane, while the greater omentum is absent. Macroscopically, the cocoon is 1 to 10 mm thick, gray or off-white. It can contain one single layer or multiple layers. It is also easy to be separated from the intestinal wall. Microscopically, it consists of dense collagen fibers with or without local inflammation. The disease could be diagnosed according to one or two of the following criteria: (1) above-mentioned imaging findings; (2) repeated intestinal obstruction and no history of abdominal surgery; and (3) a history of trauma, tuberculosis, inflammation, cancer or ascites.

Treatment

Abdominal cocoon is treated through surgical release by means of blunt separation, similar to that for adhesive intestinal obstruction. The encasement can start from the beginning part of the jejunum and end at the end of the ileum with two sides attached to the mesenteric root. When multiple abdominal organs are encased, generally, we do not need to remove the whole mass or cut the intestinal loops except for cases with intestinal strangulation or necrosis. Efforts should be on membrane excision and adhesion separation. Wide adhesions cannot be excised completely to prevent from damaging blood vessels and the intestinal tract that causes intestinal fistula or intestinal necrosis. In addition, the surface of the intestinal tract can be flushed with chitosan to avoid other adhesions. Since the intestinal tract is folded and fixed in the procedure, decompression, enema, and intestinal edema medication should be administered to inhibit the secretion of digestive juice. Early resumption of peristalsis is very important. Early activities should be appropriate and medications may be given to inhibit fibroblast growth. In our case, anti-adhesion medications were given to smear the intestinal tract during surgery. Continuous decompression, somatostatin and dexamethasone were administered. Electrocytes and fluids were infused. At 4d post operation, flatus was resumed. Prokinetic drug was given to activate intestinal peristalsis. The patient recovered well later.

In conclusion, ESN are abdominal cocoon are two rare diseases without defined causes and specific clinical manifestations. No specific diagnosis criteria are available for the two cases; and their diagnosis is dependent on surgical exploration. The two cases may not be correlated. We report these two cases and review relevant literature in our study. As the misdiagnosis rate is low for these diseases, a better understanding of them in clinical practice may help to improve the diagnosis prior to surgery.

REFERENCES


