Mucinous cystadenoma of the appendix presenting as an umbilical hernia: A case report

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Abstract. Mucinous cystadenoma of the appendix is a rare condition that develops as a result of proliferation of mucin-secreting cells in an occluded appendix. Mucinous cystadenoma of the appendix presenting as an umbilical hernia is a rare clinical entity. The most common causes of this condition are known to be ascites, hepatitis and cirrhosis; however, the patient in the present study, was diagnosed as hepatitis- and cirrhosis-negative, with no history of chronic coughing or constipation. The aim of the present study was to report a rare case of mucinous cystadenoma of the appendix presenting as an umbilical hernia in a 66-year-old female patient. The patient had a 6-month history of a reducible mass in the umbilical region and was diagnosed with umbilical hernia. Computed tomography and ultrasonography were performed and revealed massive ascites. Ultimately, a laparoscopic appendectomy was performed and borderline mucinous appendiceal cystadenoma of low malignant potential was confirmed. In addition, the present study discussed the association between mucinous cystadenoma of the appendix and umbilical hernia, as well as the diagnostic process and treatment strategies.

Introduction

Umbilical hernia is a common surgical complication, with ~10% of all primary hernias comprised of umbilical and epigastric hernias (1). According to the European Hernia Society classification, abdominal wall hernias located 3 cm above to 3 cm below the umbilicus are defined as umbilical hernia (2). In adults, intra-abdominal hypertension is a major cause of umbilical hernia (3). Predisposing factors for intra-abdominal hypertension include obesity, pregnancy or ascites. There are two primary repair options for umbilical hernias: Suture and mesh. Currently, due to low recurrence rates, mesh reinforcement is recommended (4). Mucocele of the appendix is a descriptive term that implies a dilated appendiceal lumen caused by abnormal accumulation of mucus (5). Mucinous cystadenoma is the most common cause of mucoceles of the appendix (63-84%) (6), and can generate a large amount of ascites. An appendiceal mucocele is a rare condition that is observed in 0.2-0.6% of all appendectomy specimens (7). Mucinous cystadenoma of the appendix is a rare clinical entity with symptoms that can vary, which poses a diagnostic challenge (8). Appendectomy is advised for focal or diffuse mucosal hyperplasia and cystadenoma when the appendiceal base is intact. Cecal resection is performed for cystadenoma with a large base and right colectomy is recommended for cystadenocarcinoma (9). In the present case, the increased abdominal pressure followed by the accumulation of ascites contributed to the occurrence of an umbilical hernia.

Case report

A 66-year-old female was admitted to the Third Central Hospital of Tianjin (Tianjin, China) on 3 September 2014, presenting with a 6-month history of a reducible mass in the umbilical region. A tentative diagnosis of an umbilical hernia was formed. The patient had no history of chronic coughing or constipation. Local examination revealed a large umbilical hernia (3 cm in diameter) when standing. Further examination revealed a distended and soft abdomen with no signs of tenderness or rebound tenderness, and no palpable mass or presence of shifting dullness. The patient was confirmed to be negative for hepatitis and cirrhosis. Tumor marker test results were as follows: Cancer antigen (CA)72-4, 28.98 U/ml (normal range, <8.20 U/ml); CA-125, 40.21 U/ml (normal range, <35.00 U/ml); and carcinoembryonic antigen (CEA), 11.55 ng/ml (normal range, <5.00 ng/ml). Ultrasonography showed massive ascites and disclosed no abnormalities in the liver, gallbladder, pancreas and spleen. Increased levels of CA72-4, CA-125, CEA may indicate the existence of coeliac or gynecological diseases. Computed tomography (CT) of the abdomen using a SOMATOM Definition Flash CT scanner (Siemens Healthcare AG, Munich, Germany) showed the following: i) An umbilical hernia with abdominopelvic effusion (Fig. 1); ii) diffuse peritoneal and omental thickening and calcification (Fig. 2); and iii) a...
cystic mass arising from the cecum, with calcification observed on the inside (Fig. 3). Due to the increased levels of tumor markers, abdominopelvic effusion and the imaging results, a laparoscopic exploration was performed to identify the primary cause of the hernia. During surgery, a yellow, jelly-like fluid filling the abdominal cavity and a swollen appendix wrapped with mucus and calcification were observed. Densely calcified foci were interspersed among the omentum, mesentery and lower abdominal wall. No uterine or ovarian abnormalities were found. Next, a laparoscopic appendectomy was performed. The intraoperative histopathological diagnosis was that of mucinous cystadenoma of the appendix with malignant potential. The postoperative pathological features included a single layer of tall mucus-secreting adenoma cells, large amounts of mucin filling the lumen and scant mucinous epithelium with low-grade dysplasia presented in mucinous pools. However, no invasive lesions were observed. Therefore, the post-operative pathology results diagnosed a borderline mucinous appendiceal cystadenoma of low malignant potential. No complications were observed in the post-operative period, and the patient had no recurrent disease at 1 year follow-up. The present study was approved by the Ethics Committee of the Third Central Hospital of Tianjin and written informed consent was obtained from the patient and her family.

Discussion

In adults, intra-abdominal hypertension is a major cause of umbilical hernia (3). The possible causes of intra-abdominal hypertension include obesity, pregnancy or ascites. In the present case, it was believed that the main cause of the umbilical hernia was ascites. The increased levels of tumor markers and the CT scan findings led to the consideration of the existence of ovarian cancer or mucinous cystadenoma; the diagnosis of mucinous cystadenoma of the appendix during laparoscopic exploration was unexpected.

An appendiceal mucocele is a rare condition that is observed in 0.2-0.6% of all appendectomy specimens (7). Four histological types of appendiceal mucocele exist: i) Retention cysts, ii) mucosal hyperplasia, iii) mucinous cystadenoma; and iv) mucinous cystadenocarcinoma (10,11). Mucinous cystadenoma is a rare cystic neoplasm of the vermiform appendix, which characteristic villous adenomatous changes of the appendiceal epithelium that are associated with marked distension of the appendiceal lumen with mucin (12). Among these four types, mucinous cystadenocarcinoma has the least developed etiology, and the clinical progression of the disease has not yet been determined. The clinical symptoms for mucinous cystadenocarcinoma may include right lower abdominal pain, palpable abdominal masses, weight loss, nausea, vomiting, gastrointestinal bleeding and signs of intestinal intussusception (13); however, in the present case, the disease presented as an umbilical hernia. In the present study, ultrasonography did not provide useful information, while the CT scan indicated peritoneal pseudomyxoma. Peritoneal pseudomyxoma is a peritoneal or retroperitoneal accumulation of a gelatinous substance secondary to the rupture of a mucinous appendiceal lesion (14), thus it occasionally combines mucinous ascites and peritoneal implants, in which case the prognosis is considerably poorer, synonymous with recurrent peritoneal involvement, which is known as a gelatinous disease of the peritoneum (15). Pre-operatively, the high risk of an appendiceal malignant tumor was considered in the present case; however, the intraoperative histopathological diagnosis was of a mucinous cystadenoma of the appendix, with malignant potential. An
appendectomy was the only surgical procedure performed, since it is sufficient for the treatment of such cases.

In conclusion, patients with appendiceal mucoceles present with a range of clinical symptoms. In the present case, the patient presented with an umbilical hernia. An accurate diagnosis is essential for the selection of the appropriate surgical procedure. The present showed that laparoscopic exploration is a useful procedure for the diagnosis of this intractable condition, and an appendectomy may be performed at the same time if indicated. Although rare, appendiceal mucoceles should be considered during the diagnosis of an umbilical hernia.

References