Case Report

Benign multicystic peritoneal mesothelioma: A rare case presenting as pneumoperitoneum and pneumotosis intestinalis

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Introduction

Benign multicystic peritoneal mesothelioma (BMPM) is a rare peritoneal lesion that commonly presents as lower abdominal pain, palpable mass, or both. A case of BMPM presenting as pneumoperitoneum and pneumotosis intestinalis is described. Utilizing PubMed, a review of the literature was performed using the search words "benign multicystic peritoneal mesothelioma," "multilocular peritoneal inclusion cysts," "pneumoperitoneum," and "pneumotosis intestinalis." Although this lesion is typically benign, it has a high recurrence rate as well as the potential for malignant transformation; it is important to confirm the diagnosis in a minimally invasive manner, and to formulate a surgical treatment strategy to avoid unnecessary re-operations.

Case report

A 65-year-old female presented to the emergency room after a fall. The patient was given intravenous fluid resuscitation for hypotension after her initial vital signs were taken. A CT scan of her abdomen and pelvis was performed to evaluate the cause of her hypotension. The CT scan (Fig 1) indicated evidence of free intraperitoneal air; the surgical team was consulted.

Upon further questioning, the patient admitted to an episode of left lower quadrant (LLQ) pain approximately
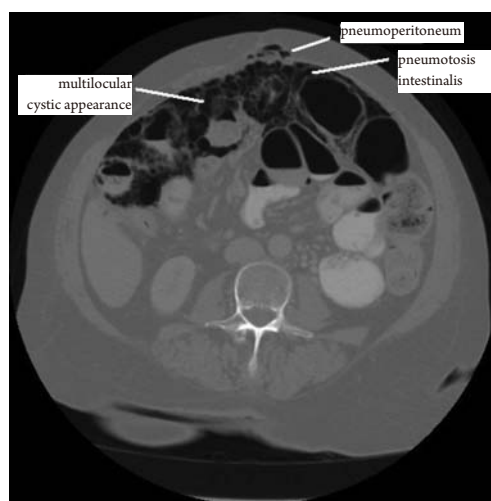

Figure 1 CT scan of the abdomen showing multicystic appearance, pneumoperitoneum and pneumotosis intestinalis.

one week prior and was now complaining of some LLQ pain. Her medical history was significant for atrial fibrillation and hypertension, as well as bilateral inguinal hernia repairs, umbilical hernia repair and surgeries on her right shoulder, bilateral knees, and bilateral hips. She denied alcohol use and stopped smoking over twenty years ago.

The patient was afebrile with a pulse of 77 and blood pressure of 104/67 after fluid resuscitation. Her chest was clear and her cardiac exam was unremarkable. The abdominal exam revealed some LLQ tenderness and her extremity exam showed palpable pulses bilaterally and evidence of surgical scars of her hips and knees. Initial laboratory data was within normal limits with a hemoglobin of 12, hematocrit of 36, creatinine of 0.9, and a white blood cell count of 8,000.

The patient initially refused explorative surgery and was treated with IV antibiotics and admitted to the intensive care unit for observation. The following day, the patient and

Figure 2 **Gross appearance of small bowel segment intra-operatively showing multilocular cysts covering the serosa and mesentery of the small bowel.**

her family agreed to an exploratory laparotomy to evaluate the etiology of the pneumoperitoneum. The surgical findings revealed cloudy peritoneal fluid and intraperitoneal air causing increased suspicion of perforation. A segment of small bowel, approximately 15 cm in length, with a gross appearance of pneumatosis intestinalis (Fig 2) was resected, and a primary anastomosis was performed.

A thorough inspection of the abdomen was performed with close attention to the duodenum and left colon to inspect for other sources of the free intraperitoneal air. No evidence of duodenal ulcer or diverticular disease was identified. There was no gross evidence of uterine inflammation or pelvic inflammatory disease. The liver capsule was normal without evidence of Fitz-Hugh Curtis syndrome. Following an incidental appendectomy, her abdomen was copiously irrigated and closed with retention sutures.

The patient's postoperative course was unremarkable. She was slowly advanced to a regular diet and was discharged to a rehabilitation facility for physical therapy 7 days after initial admission.

Two specimens were examined: 1) the appendix and 2) a portion of small intestine. The appendix measured 3.2 cm long by 0.6 cm in diameter with a small amount of adherent mesoappendiceal fat. The appendix serosa was tan, smooth, and shiny with focal vascular congestion. A fecalith was also noted in the distal lumen. The diagnosis was mild acute periappendicitis with no evidence of perforation. The small bowel resection consisted of a segment of small intestine 8 cm long by 4 cm in diameter with an attached 6 cm by 2 cm by 0.8 cm fragment of mesenteric fat. The serosal surface

was tan-white and shiny with a 6 cm by 5.5 cm by 1 cm multicystic subserosal lesion in the center. The specimen was opened revealing mucosa that was tan and glistening with the usual mucosal folds. There was no evidence of perforation throughout the specimen. The subserosal tissue was edematous with a discrete, complex, multiloculated, thin-walled cystic lesion. The cysts had thin fibrous walls lined by flattened mesothelial cells containing clear serous fluid. No immunohistochemistry was performed as this was an incidental finding with low suspicion. The final pathologic diagnosis of the small bowel partial resection was multicystic peritoneal mesothelioma with no evidence of perforation.

Discussion

Approximately 150 cases of benign multicystic peritoneal mesothelioma, with various presentations have been reported since it was first described by Mennemeyer and Smith in 1979 (3-12). Upon extensive literature review, no report of BMPM presenting with pneumoperitoneum and pneumotosis intestinalis was identified. This disease is quite rare (0.15/100,000 annually) which makes its diagnosis, treatment, origin, and pathogenesis a unique clinical challenge (3).

Benign multicystic peritoneal mesothelioma lesions usually occur in the peritoneum along the pelvic cul de sac, uterus, and rectum, but may occasionally involve the round ligament, small intestine, spleen, liver, kidney, previous scars, or the appendix (2,1,3,4). Unlike malignant mesothelioma, BMPM has not been shown to have an association with asbestos exposure. In as many as half of the cases, lesions have recurred within a few months to years after resection (1). Although it is considered benign, rare cases have been reported to proceed to malignant transformation (5).

BMPM, also referred to as multilocular inclusion cysts, occurs most frequently in young to middle-aged premenopausal women (1,2). Rarely, it occurs in males (10,14). The disease has been considered to be either a hyperplastic reactive lesion or a benign neoplasm. Due to its reported association with previous abdominal surgery and endometriosis, some authors support the notion of BMPM being a non-neoplastic reactive lesion (2), however, recurrence after partial resection and malignant transformation resulting in death has been well documented over the years (5).

The lesions typically appear as single or multiple small, thin-walled, translucent, unilocular cysts that may be attached or free in the peritoneal cavity (1). Extraperitoneal locations such as the pleura, spermatic cord, and

pericardium have been rarely reported (2). Grossly the cysts are most often seen attached and growing on the surfaces of the pelvic cul de sac, uterus, and rectum in a multilocular mass. The cystic fluid varies from yellow to watery or gelatinous in consistency with the cytology showing sheets of benign monomorphous mesothelial cells (2,1). On microscopic examination BMPM cysts are lined by a single layer of flattened to cuboidal mesothelial cells which occasionally have a "hob-nail" appearance. In up to one third of the cases, the lining of the cells can undergo adenomatoid or squamous metaplasia (1,2).

Although pneumoperitoneum and pneumatosis intestinalis have a wide variety of differential diagnoses ranging from benign to life threatening, these conditions have never been reported as associated with benign multicystic mesothelioma. The differential diagnosis of BMPM includes a variety of malignant and benign lesions that present as cystic or multicystic abdominal masses. Cystic lymphangioma, cystic adenomatoid tumors, cystic mesonephric duct remnants, endometriosis, mullerian cysts involving the retroperitoneum, and cystic forms of endosalpingiosis are several of the benign lesions that should be considered in the differential (11). Multilocular cystic lymphangiomas are the most commonly confused lesions with BMPM. Unlike BMPM, cystic lymphangiomas usually occur in male children in extrapelvic regions. They are usually found localized to the small bowel, omentum, mesocolon, or retroperitoneum and contain chylous contents. Unlike BMPM, they also have mural lymphoid aggregates and smooth muscle unlike (1,11). Malignant lesions to consider are malignant mesothelioma and serous tumors that involve the peritoneum.

BMPM usually presents with vague lower abdominal pain, mass, or both, but is also commonly diagnosed incidentally upon laparotomy for other surgeries (1). The patient may also present with obstructive symptoms such as nausea, bloating, or vomiting. Despite its relatively benign process some patients may present with an acute abdomen (11). CT scans may be diagnostically beneficial but, as in this case, can also indicate a more acute need for surgery as actually necessary. Pre-operative fine needle core biopsies have been reported to be of some benefit in the differential diagnosis of BMPM (11,16). Cytologic features of peritoneal washings in cases of BMPM have shown the washings to be hypercellular with a population of mesothelial and squamous metaplastic cells (6). Ultimately, the diagnosis is usually made by the pathologist after surgical resection has been performed.

Due to its rarity, BMPM treatment options remain an area of controversy and there is no streamlined treatment plan. Currently aggressive surgical resection is the mainstay of treatment with palliative debulking and reoperation for recurrence (15,11,5). With up to 50 percent recurrence rates and its malignant potential, debulking surgery does not appear to be the most acceptable treatment option for these patients. Patients may suffer from poorly controlled chronic abdominal and pelvic pain (15). Uncertain results have been reported with patients receiving adjuvant chemotherapy and/or radiation therapy (5). Other approaches such as sclerosive therapy with tetracycline, continuous hyperthermic peritoneal perfusion with cisplatin, and antiestrogenic drugs have been suggested (11). The optimal treatment may be cytoreductive surgery with peritonectomy combined with perioperative intraperitoneal chemotherapy to eliminate all gross and microscopic disease (5). The goal of this treatment regimen is to reduce the likelihood of progression or recurrence.

Although the prognosis for BMPM is very good, aggressive approaches to this disease should be considered. Patients have a high likelihood of recurrence and repeat surgeries are common. The intention of this report is to increase the awareness of this disease entity and to consider it whenever the patient's presentation does not match that of the working diagnosis. This patient presented without peritoneal signs despite a CT scan that suggested a more severe pathology. Before jumping into an exploratory laparotomy based on imaging findings, surgeons should trust our physical exam and pursue a more definitive diagnosis. With a definitive diagnosis we can approach the surgical issue in the most appropriate manner. In this case, a diagnosis could have been made by a minimally invasive technique such as a needle biopsy or a diagnostic laparoscopy. Once a definitive diagnosis of BMPM is made, then a single surgery should be the goal to eliminate all gross and microscopic disease.

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