CASE REPORT

Mucocele of the appendix causing tubal factor infertility

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Objective: To report on the safety of conservative management of a young woman with pseudomyxoma peritonei of appendiceal origin desiring fertility preservation.

Design: Case report.

Setting: IVF Center at Tertiary Hospital.

Patient(s): A 32-year-old woman with primary infertility.

Intervention(s): Appendectomy, conservative second-look laparoscopy, and IVF.

Main Outcome Measure(s): Persistence of active mucinous lesions in the peritoneal cavity, presence of abnormalities at colonoscopy, and maternal/fetal complications from IVF.

Result(s): Normal second-look laparoscopy, no evidence of persistent disease, and successful intrauterine twin pregnancy following IVF.

Conclusion(s): Pseudomyxoma peritonei of appendiceal origin is rare in young female patients. Although traditional therapy in older patients often involves aggressive surgical therapy, this case report demonstrates that patients desiring fertility can be managed conservatively and safely and still maintain successful pregnancy.


Key Words: Mucinous cystadenoma, pseudomyxoma peritonei, disseminated peritoneal adenomucinosis, primary infertility, in vitro fertilization

We report the case of a 32-year-old healthy nulligravida Caucasian female who presented with a 3-year history of primary infertility. She reported menstrual cycles ranging between 26 and 42 days in length, but was confirmed to be periodically ovulatory by serum progesterone values. She had normal ovarian reserve testing by early follicular FSH and estradiol levels, and her partner’s semen analysis was normal. This patient underwent several ovulation induction cycles using Clomiphene Citrate, with confirmed evidence of ovulation but without achieving pregnancy. At this time, the decision was made to proceed with a hysterosalpingogram (HSG), which revealed a normal intrauterine cavity, delayed intraperitoneal spillage of contrast from the left fallopian tube with distal loculation, and a distal right hydrosalpinx with a question of delayed spillage of contrast versus loculation. Ultrasound examination confirmed the presence of a small (1.5 × 1.0 cm) right hydrosalpinx.

The patient’s gynecologic, medical, and surgical histories were otherwise unremarkable except for an ASCUS pap smear followed by a normal colposcopy. A review of systems was significant only for occasional right lower quadrant dull pain in the absence of other symptomatology. Her family history was significant for a father with hypertension and dyslipidemia and a mother with Grave’s disease. She denied any known family history of malignancy. Her husband’s medical, surgical, and family histories were likewise unremarkable. Her physical examination, including an abdominal and pelvic examination, was within normal limits. She had a body mass index of 28.4 kg/m². After extensive counseling and review of her HSG, a decision was made to perform a laparoscopic evaluation of her fallopian tubes and abdominal cavity with possible removal of her right hydrosalpinx.

Laparoscopic exploration (Fig. 1, top) revealed a small anteverted uterus with two small subserosal pedunculated posterior myomas. Both ovaries were free of adhesions, cysts, and endometriosis. The right fallopian tube was adherent to the pelvic side wall; although fimbria were visualized, its distal portion was mildly dilated. No distinct hydrosalpinx was visualized. The left fallopian tube appeared normal in caliber except for mild phimosis at the fimbriated end. Yellowish, gelatinous material covered the pelvic and abdominal...
cavities, including the anterior and posterior cul-de-sac, the uterus, and the ovarian fossas bilaterally. Pelvic washings with mucin were sampled and removed by suction for pathologic evaluation. Careful inspection revealed no evidence of endometriosis. Chromopertubation of the fallopian tubes revealed slow intraperitoneal spill from the left tube and localized spill from the right tube. Bilateral fimbrioplasty was performed, and subsequent chromopertubation revealed prompt spill bilaterally. The abdominal cavity was inspected more closely; the appendix was noted to be mildly dilated at its distal end without erythema or gross infection but appeared covered in gelatinous material. The remaining small and large bowel, liver, and stomach appeared normal except for mild midline omental adhesions to the anterior abdominal wall that were easily lysed. We performed an appendectomy and removed multiple areas of mucin-covered peritoneal adhesions. The patient experienced a routine postoperative course.

Pathologic evaluation of the appendix revealed a mucinous cystadenoma (5.0 × 0.8 cm) involving nearly the entire appendix. The resection margins were negative without evidence of appendiceal perforation, and there was one focus of fibrosis and nuclear atypia reported by pathology. Peritoneal adhesions revealed “mesothelial hyperplasia and mucin devoid of neoplastic epithelium” with no evidence of endometriosis or malignancy, and the washings were “negative for malignant cells.”

Postoperatively the patient was referred to a surgical oncologist. After extensive counseling on the unusual nature of her condition she underwent a colonoscopy with normal findings and proceeded to a second laparoscopic evaluation 4 months later. This second-look procedure revealed multiple white fibrotic lesions covering the pelvic structures, distal small bowel mesentery, and abdominal and pelvic peritoneal surfaces (Fig. 1, bottom), and biopsies were performed in these areas. At this time the abdominal cavity was grossly free of mucin. Pathologic evaluation of these white lesions revealed “fibromuscular and fibroadipose tissue with minute foci of acellular mucin and no epithelial cells.”

To address the patient’s primary desire for fertility, the decision was made in consultation with her surgical oncologist to first proceed with in vitro fertilization (IVF) with subsequent consideration for more definitive management of her mucinous cystadenoma. After GnRH agonist down-regulation, her ovaries were stimulated with injectable gonadotropins. Oocyte retrieval yielded 21 total oocytes, and 9 out of 15 mature oocytes fertilized. She underwent a two-embryo blastocyst transfer 5 days after oocyte retrieval and subsequently achieved a viable twin pregnancy. She delivered by
vaginal delivery at full term without maternal or fetal complications.

After delivery, the patient was again counseled regarding therapeutic options for her cystadenoma. In consultation with both a general surgeon and surgical oncologist, she was given options that included radiologic observation only, periodic surgical reexploration with pathologic examination, and radical surgical treatment. With an overall lack of knowledge of the disease course in her age group, her favorable histologic type, and minimal scope of disease, she was counseled that her prognosis would likely be better than patients included in the literature. She was also counseled that the existing literature does not provide an answer as to the benefit of additional surgery in a patient with disease as minimal as hers. Concordant with her desire for possible future childbearing, she elected to undergo periodic radiologic (CT scan) and possible surgical monitoring.

DISCUSSION

Pseudomyxoma peritonei (PMP) is a rare condition involving the accumulation of mucin throughout the abdominal and pelvic cavities. Pseudomyxoma peritonei is now more appropriately classified as disseminated peritoneal adenomucinosis (DPAM) (primary tumor adenoma, minimal to moderate atypia, and no significant mitoses), peritoneal mucinous carcinomatosis (PMCA) (primary tumor carcinoma, marked atypia, signet ring cells), or intermediate in nature (1). Pseudomyxoma peritonei is most often associated with an appendiceal mucoecele (or obstructive dilation), largely from appendiceal cystadenomas or cystadenocarcinomas, but in women has ovarian origins as well. Cytologically, specimens often consist of mesothelial cells, fibroblasts, and mucin (2, 3). Approximately half of all mucocoeles produce no recognizable symptoms, and may be incidental surgical findings. If symptomatic, the most common presentation is that of abdominal pain. A CT scan may be useful in diagnosing PMP before surgical intervention with characteristic low attenuation ascites and coating of visceral surfaces (4), but this requires a high index of suspicion even in the presence of a known tumor and certainly in cases where the finding is incidental. Pseudomyxoma peritonei can theoretically be prevented at the time of appendectomy by ensuring that the specimen is sent intact with no spillage of cells intraperitoneally, as approximately 1% of all appendiceal specimens reveal a benign tumor, and PMP may develop as long as 10 years from the initial tumor presentation (5). Although progression of disease is rapid in the case of PMCA with peritoneal studding, almost no information is known about the natural history of DPAM in the presence of minimal and isolated disease.

Most mucinous appendiceal tumors appear in older men and women, with median age at diagnosis over 50 years of age. Reports in younger women are rarer. However, reports of cystic right adnexal masses thought to be ovarian in origin were eventually surgically confirmed to be appendiceal mucinous cystadenomas (6). The literature reveals five cases where appendiceal tumors appear specifically associated with infertility in women. Two malignant cases were reported in 1987 (7). The first published case with subsequent fertility was reported in 1992, in a woman presenting with abdominal pain and gastrointestinal symptoms (8). The first reported pregnancy in a case treated initially by surgical exploration and chemotherapy with cisplatin was reported in 1995 (9). Recently, a women presenting with primary infertility thought to have a right hydrosalpinx on pelvic ultrasound was confirmed to have appendiceal mucinous cystadenoma with PMP (10). In very rare cases, patients found to have PMP and apparent mucocoeles of the appendix were actually diagnosed on final histology with endometriosis infiltrating and obstructing the appendix. Only one of these patients presented with infertility (11–13).

Historically, treatment for PMP consisted of aggressive cytoreductive surgical debulking including right hemicolectomy and extensive pelvic deperitonealization followed by intraperitoneal chemotherapy. When stratified retrospectively by histology, this yielded 5-year disease-free survival rates of approximately 80% for DPAM and 25% for PMCA. This latter number has been significantly improved if adequate debulking was possible (14, 15). This radical surgical practice has more recently been challenged in favor of less aggressive surgical management, reserving colectomy for cases where appendiceal surgical margins are positive, lymph nodes show involvement, or when chemotherapy is contraindicated (16, 17). However, there are no data on which to base the extent of peritoneal surgery. Additionally, survival data are almost exclusively based on cases where the underlying histology is PMCA or else multifocal DPAM in older patients. There is little basis on which to provide prognostic, surgical, or survival data for young patients with solitary, benign DPAM.

In this case report, the patient elected to undergo fertility treatment with IVF after initial surgery with the goal of achieving pregnancy before completing treatment for her mucinous cystadenoma. This patient completed her pregnancy without any known complications to herself or to her children. In the absence of evidence for malignancy, there is a role for conservative management in young women when fertility remains desirable. Additionally, in a patient with abnormal HSG findings and no history suggesting an etiology of tubal damage, laparoscopy may help identify women with important nongynecologic disease.

REFERENCES


