CLINICAL IMAGE

PROGRESSIVE ABDOMINAL DISTENTION: A CASE OF PROGRESSIVE ABDOMINAL GROWTH IN A PREMENOPAUSAL WOMAN

Samantha Rikabi, OMS-IV; Lindsay Tjiattas-Saleski, DO, MBA, FACOEP

Edward Via College of Osteopathic Medicine-Carolinas, Spartanburg, SC

A 21-year-old G0P0 female with a past medical history of GERD and headaches presented to her primary care physician for abdominal distension and growth for the past 10 months. She developed an uncomfortable tightness in her abdomen, with evident suprapubic tenderness and progression of pain into the umbilical and epigastric regions. During the month before presentation, her symptoms worsened and she developed nausea, vomiting, fatigue, pelvic pain/tightness, abdominal distention, indigestion, and increasing abdominal girth (Figure 1). She noted early satiety, which she attributed to possible food intolerance, and tried multiple diets without improvement (gluten-free, keto, and lactose-free diets). She also noted left upper flank numbness when sitting up and driving.

In addition to the physical pain, she experienced emotional distress and embarrassment as people frequently asked whether she was pregnant. She no longer fit into her clothes and tried hiding the abdominal enlargement with oversized clothes. She did not note changes in bowel habits. She denied weight loss or gain according to her home scale; however, she did notice increasing abdominal girth.

Menarche started at age 12 and her cycles had been regular at 5 or 6 days long every 28 days with heavy cramping. She has a family history of heart failure, type 2 diabetes mellitus, and gastric carcinoma from her paternal grandmother. She took famotidine daily and ibuprofen as needed.

FIGURE 1:

Preop. Weight 216 lbs.

CORRESPONDENCE:
Lindsay Tjiattas-Saleski, DO, MBA, FACOEP
ltjiattassaleski@carolinas.vcom.edu

Copyright© 2022 by the American College of Osteopathic Family Physicians. All rights reserved.

QUESTIONS

1. What is the most likely cause of this patient's presentation?

a. Appendicitis

b. Celiac disease

c. Crohn's disease

d. Ectopic pregnancy

e. Ovarian mass

2. What is the most appropriate treatment for this patient?

a. Antibiotics, rest, and IV fluids

b. Corticosteroids and follow-up with rheumatologist

c. Dietary restrictions

d. Medication-induced abortion/surgery

e. Surgical intervention and removal of mass

ANSWERS

1. What is the diagnosis of this patient?

Correct answer:
E. Ovarian mass

The signs and symptoms exhibited by this patient are consistent with the development of a slow-growing mass—in this case, a mucinous cystadenoma. Ectopic pregnancy is unlikely due to continued regular menstrual cycle, extensive abdominal enlargement, lack of distinctly localized pain, and vaginal bleeding. Crohn's disease is a chronic, autoimmune inflammatory disease that occurs in genetically predisposed patients with symptoms including diarrhea, malabsorption, and abdominal distention.[1] This patient did not exhibit symptoms similar to these. Appendicitis presents as pain in the periumbilical and right lower abdominal areas, fever, and anorexia.[2] The symptoms of appendicitis typically start suddenly and progress over hours to days. Celiac disease is an inflammatory disease that affects the small intestine and demonstrates symptom regression on maintaining a gluten-free diet.[3] The patient thought she was experiencing food intolerance and thus experimented with multiple diets, including a gluten-free diet, but did not experience relief.

2. What is the appropriate treatment protocol for this patient?

Correct answer:
E. Surgical intervention and removal of mass

The appropriate treatment for ovarian mucinous cystadenoma is a unilateral salpingo-oophorectomy or ovarian cystectomy.[4-8] Clinical recurrence is uncommon after surgical intervention, but if it does occur, the tumor may not have been completely resected or there may be a new primary tumor.[7] Medication-induced abortion would be the appropriate choice for treating ectopic pregnancy.[9] Corticosteroids and follow-up with a rheumatologist is an appropriate treatment course for Crohn's disease. The main goal of treatment is to induce remission from the current symptom flare-up and prevent complications of the disease.[1] Antibiotics, rest, and IV fluids are the recommended treatment for uncomplicated appendicitis. Surgical intervention is recommended for more severe cases of appendicitis.[2] Celiac disease is treated by following a strict gluten-free diet for life. Patients must avoid foods that contain wheat, rye, barley, spelt, and more. Symptoms improve with adherence to the diet.[3]

DISCUSSION

Ovarian neoplasms are classified into three categories based on tumor cell origin: stromal, germ cell, and epithelial with further subtypes as discussed in Table 1.[4,10,11] Epithelial tumors comprise approximately 60% of all ovarian tumors, with 40% of these being benign.[7] The two most common types of epithelial tumors are serous and mucinous cystadenomas, with their malignant counterparts being cystadenocarcinomas.[7] Mucinous cystadenomas are benign ovarian neoplasms of epithelial origin that comprise approximately 10% to 15% of all benign ovarian neoplasms.[4,7,11,12]

Table 1:
Subtypes of ovarian neoplasms

OVARIAN NEOPLASMS	
Stromal/sex cord tumors	Fibroma, granulosa-theca cell tumor, Leydig cell tumor, and Sertoli cell tumor
Germ cell tumors	Teratoma, dysgerminoma, endodermal sinus tumor, and choriocarcinoma
Epithelial tumors	Serous cystadenoma/cystadenocarcinoma, mucinous cystadenoma/cystadenocarcinoma, endometrioid tumors, clear cell tumor, and Brenner tumor

Mucinous cystadenomas are smooth tumors lined by a single layer of epithelial cells that secrete mucin.[11,13] The tumor size can range from a few centimeters in diameter to more than 30 cm.[7,11,12] There have been reports of tumors weighing up to 135 kg if there has been a delay in diagnosis.[12,13] The tumors are most likely to develop during the third to sixth decades, but they can occur in younger women (rarely less than 21 years of age).[4,5,7] They are unilateral in 95% of cases.[4,7,10,11,12]

The etiology of mucinous cystadenomas is currently unknown. There are associated risk factors, including obesity and tobacco use.[14,15] *Kirsten rat sarcoma viral oncogene (KRAS)* mutations have been documented in up to 58% of the cases.[5,7,8,14] *KRAS* is a proto-oncogene involved in the RAS/MAPK pathway responsible for cell proliferation.[8] *KRAS* mutations lead to an unregulated proliferation of cells and cause neoplasm formation. There are no current therapies available as *KRAS* has proved difficult to target with drug therapy; however, active research is being conducted to target *KRAS* mutations.[8]

Ovarian mucinous cystadenomas are generally asymptomatic in the early stages and are associated with nonspecific symptoms during growth.[6,7,11] Signs and symptoms experienced by patients commonly include pelvic pain, progressive abdominal distention, early satiety, heartburn, nausea, increased urinary frequency, urinary retention, and generalized discomfort.[4,7] The average tumor size upon discovery is typically 10 cm in diameter.[11] If not diagnosed early, the tumors can grow quite large, causing compressive or mass-associated symptoms as mentioned previously.[4,12,13] Complications of ovarian neoplasms can include torsions, hemorrhage, or rupture.[4,6,16] Rupture of cysts can lead to peritonitis, sepsis, and death from septic shock.[4,16] A rare but life-threatening phenomenon that can occur from a cystic rupture is called pseudomyxoma peritonei.[10,16-18] This involves widespread seeding of mucin-producing cells throughout the peritoneal cavity and can lead to bowel obstruction and peritonitis.[10,18]

The diagnostic workup for a patient with progressive abdominal enlargement and associated compressive symptoms is comprehensive and requires abdominal imaging.[6,13] A CT scan may be ordered first based on presenting symptoms such as abdominal distention, acid reflux, and nausea; however, the best initial evaluation of an adnexal mass is via ultrasound.[4,6,13] The pelvic ultrasound is not definitive and necessitates follow-up with a histopathological exam of the surgical specimen.[4,7] The histological findings of a mucinous cystadenoma will exhibit multiple cysts due to its multilocular nature and glands lined by simple nonstratified mucinous epithelium.[6,7,10,16] There will be no cytologic atypia or any mitotic figures present in the specimen consistent with a benign condition.[7] If the cysts do not contain septa, papilla, or solid components based on ultrasound results, the tumor can be closely monitored.[13] Surgical exploration should be considered if the mass changes in size or character.[13]

Mucinous cystadenomas can present with elevated tumor markers such as β-hCG, AFP, LDH, inhibin, CAE, CA-125, CA19-9.[4,5,11] Elevated tumor marker CA-125 has been documented in 80% of epithelial ovarian cancers and can be used for surveillance of disease recurrence.[5,10,11] Some guidelines recommend using CA-125 tumor marker for initial evaluation and management in women who present with symptoms suggestive of ovarian cancer; however, the use of tumor markers is not common practice.[5,10,11] If ovarian cancer is high on the differential, then the next step in management is an abdominal and pelvic ultrasound.[11]

Tumors of the ovary that present with diameters greater than 10 cm are referred to as giant ovarian masses.[6] The standard treatment is a unilateral salpingo-oophorectomy with intraoperative pathological evaluation.[4,6,7] Specific surgical techniques include drainage of

cystic fluid, cystectomy, oophorectomy, and/or hysterectomy.[4,12] Oophorectomy is the preferred and most common method because it has the lowest rate for local recurrence.[4-8,18] A minimally invasive laparoscopic approach is ideal, but open laparotomy procedures are more commonly practiced.[4,12] Laparoscopy has been shown to decrease recovery time and morbidity when compared with a laparotomy.[4,12] However, opting to perform a laparoscopy depends on several factors such as tumor size, susceptibility to rupture, and surgeon level of comfort.[4,12]

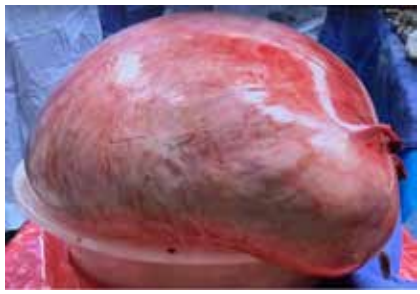
The prognosis of mucinous cystadenomas is excellent, with a five-year survival rate of 98% after surgical excision.[7,18] There are documented cases where patients prolong seeking health care for various reasons including access to health care, willingness to seek treatment, economic status, level of education, and emotional components like fear and anxiety.[4] If left untreated, the mass grows exponentially, affecting the patient's quality of life and increasing the risk of rupture.[4,6,16] Although there are no documented cases of intrabdominal rupture, continued growth without intervention can cause the cyst wall to thin.[12] Thinning of the cystic wall further complicates surgical removal and enhances the risk of rupture.[12] In these situations, the less common technique of intraoperative surgical aspiration before removal is the preferred method.[12] Surgical aspiration is not commonly done and is performed under special circumstances.[6] The preferred method is the removal of the tumor intact since aspiration of cysts before excision is associated with increased risks of recurrence, infections, bleeding, cystic rupture, peritoneal adhesions, or possible dissemination of malignant cells.[4,6,7,18]

The patient in this case sought care from her primary care physician after the development of abdominal distension and progressive compressive symptoms. She delayed seeking treatment for several reasons and thus presented with a large mass weighing 32.6 lbs. and measuring 42 cm in greatest diameter (Figure 2). She was immediately referred to an obstetrician-gynecologist for further evaluation and an abdominal and pelvic ultrasound was performed. The ultrasound revealed a large cystic mass with multiple septations measuring an estimated 20–30 cm. The mass was suspected to be a neoplasm originating from the left ovary. Four days after her initial presentation, she had an open left salpingo-oophorectomy via midline laparotomy incision from xiphoid to pubis (Figure 3). The final pathologic report revealed that the tumor was negative for atypia or malignancy and confirmed the diagnosis. The patient is recovering well from the procedure (Figure 4).

CONCLUSION

Ovarian mucinous cystadenomas are benign tumors originating from surface epithelium of the ovary and are diagnosed at an average age of 40–49.[4,7,11,18] They are large, multiloculated, cystic masses containing mucinous fluid that can grow unregulated until medical intervention is implemented.[7,10] These tumors typically require surgical excision.[4,5,12] The prognosis is exceptional, with a five-year survival rate of 98%.[7,18] Local recurrence is minimized with oophorectomy, and only rare cases of malignant transformation have been documented.[4,7,18] Early detection and treatment provide the best prognosis and improvement in quality of life.[4]

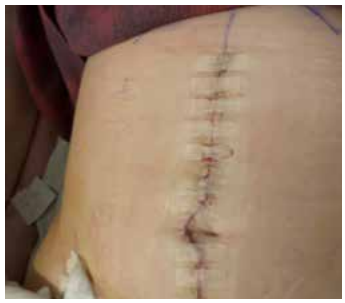
FIGURE 2:



Per pathology report: "Final pathologic diagnosis: mucinous cystadenoma, 42 cm in greatest dimension, negative for atypia or malignancy, histologically unremarkable fallopian tube."

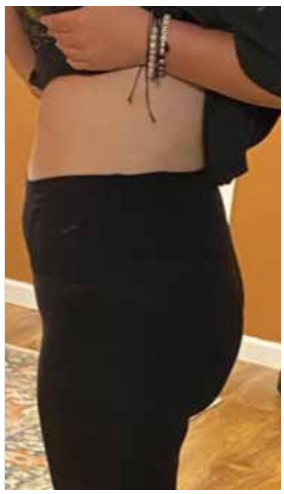
Gross description: "32.6 pounds, 42x30x29-cm fluctuant intact cystic ovary with an associated 30x.04-cm elongated fallopian surface. The largest cyst contains yellow-red watery fluid and additional smaller cysts contain translucent mucoid fluid. No significant solid or papillary are as identified."

FIGURE 3:



Postsurgical incision.

FIGURE 4:



Two weeks postop. Weight 178 lbs.

REFERENCES

1. Baumgart DC. The diagnosis and treatment of Crohn's disease and ulcerative colitis. *Dtsch Arztebl Int*. 2009;106(8):123–133. doi:10.3238/arztebl.2009.0123
2. Hardin DM Jr. Acute appendicitis: review and update. *Am Fam Physician*. 1999;60(7):2027–2034. PMID: 10569505.
3. Schuppan D, Zimmer KP. The diagnosis and treatment of celiac disease. *Dtsch Arztebl Int*. 2013;110(49):835–846. doi:10. 3238/arztebl.2013.0835
4. Akhras LN, Akhras LN, Faroog S, AlSebary L. A 27-kg giant ovarian mucinous cystadenoma in a 72-year-old postmenopausal patient: a case report. *Am J Case Rep*. 2019;20:1606–1060. doi:10.12659/AJCR.917490
5. Cowan RA, Haber EN, Faucz FR, Stratakis CA, Gomez-Lobo V. Mucinous cystadenoma in children and adolescents. *J Pediatr Adolesc Gynecol*. 2017;30(4):495–498. doi:10.1016/j.jpag.2017.02.001
6. Gwanzura C, Muyotcha AF, Magwali T, Chirenje ZM, Madziyire G. Giant mucinous cystadenoma: a case report. *J Med Case Reports*. 2019;13(1):181. doi:10.1186/s13256-019-2102-z
7. Limaiem F, Lekkala MR, Mlika M. Ovarian cystadenoma. In: StatPearls [Internet]. StatPearls Publishing; 2022 Jan–. PMID: 30725635.
8. Somagutta MR, Luvsannyam E, Jain MS, et al. A rare case of massive ovarian mucinous cystadenoma with postmenopausal bleeding. *Cureus*. 2020;12(9):e10198. doi:10.7759/cureus.10198
9. Murray H, Baakdah H, Bardell T, Tulandi T. Diagnosis and treatment of ectopic pregnancy. *CMAJ*. 2005;173(8):905–912. doi:10.1503/cmaj.050222
10. Magowan B, Owen P, Thomson A. Ovarian neoplasms. In: Magowan B, Owen P, Thomson A, eds. *Clinical Obstetrics & Gynaecology*. 4th ed. Elsevier; 2019:115–121.
11. Moyon MA, León DA, Aguayo WG, et al. Giant ovarian mucinous cystadenoma, a challenging situation in resource-limited countries. *J Surg Case Rep*. 2019;12:rjz366. doi:10.1093/jscr/rjz366
12. Moon, AS, DeAngelis AM, Fairbairn M, et al. Removal of 132-pound ovarian mucinous cystadenoma: a case report. *SAGE Open Med Case Rep*. 2020;8. doi:10.1177/2050313X20906738
13. Lavie O. Benign disorders of the ovaries & oviducts. In: DeCherney AH, Nathan L, Laufer N, Roman AS, eds. *Current Diagnosis & Treatment: Obstetrics & Gynecology*. 12th ed. McGraw Hill; 2019. Accessed November 3, 2022. https://accessmedicine.mhmedical.com/content.aspx?bookid=2559&sectionid=206964809
14. Craen AM, Lebowitz D, Amico K, Ganti L. Mucinous cystadenoma causing abdominal distension: a case report. *Cureus*. 2018;10(11):e3657. doi:10.7759/cureus.3657
15. Jordan SJ, Green AC, Whiteman DC, Webb PM. Risk factors for benign serous and mucinous epithelial ovarian tumors. *Obstet Gynecol*. 2007;109(3):647–654. doi:10.1097/01.AOG.0000254159.75977.fa
16. Kamel RM. A massive ovarian mucinous cystadenoma: a case report. *Reprod Biol Endocrinol*. 2010;8:24. doi:10.1186/1477-7827-8-24
17. Gorli B. A rare case of ruptured mucinous cystadenoma causing pseudomyxoma peritonei. *Pediat Therapeut*. 2012;2:6. Poster presented at: Second International Conference on Pediatrics & Gynecology; September 24–26, 2012. Accessed November 3, 2022. https://www.longdom.org/proceedings/a-rare-case-of-ruptured-mucinous-cystadenoma-causing-pseudomyxoma-peritonei-22124.html
18. Marko J, Marko KI, Pachigolla SL, Crothers BA, Mattu R, Wolfman DJ. Mucinous neoplasms of the ovary: radiologic-pathologic correlation. *RadioGraphics*. 2019;39(4):982–997. doi:10.1148/rg.2019180221