CLINICAL VIGNETTE

A Patient with Enlarging Painful Abdomen

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Case Presentation

A 65-year-old female transferred care from another medical system to UCLA for increasing abdominal discomfort as well as poor appetite and ten pound weight loss.

Six months prior she was evaluated at an outside institution. A 20 cm cystic structure was found on abdominal ultrasound. Abdominal CT scan confirmed a large cystic structure in the right hemiabdomen measuring up to 20 cm, with simple anechoic appearance and unclear origin. She underwent ultrasound guided cyst aspiration with removal of two liters of clear fluid and partial amelioration of her abdominal discomfort. Cell count included 14 nucleated cells, 98 monocytes, and no mesothelial cells, Fluid LDH was 13, creatinine 0.5, and CEA in the fluid was 96.3. Gram stain was negative and no albumin or total protein was reported.

On exam she was underweight with BMI of 16. Her abdomen was soft, mildly distended, nontender, with no appreciated ascites. Review of her prior labs included normal amylase, lipase, CBC, CMP, celiac serology, iron, ferritin, B12, CA 19-9, TSH, Vitamin D. The CEA was slightly elevated at 3.9 (normal <3.1). Her past medical history includes fibromyalgia, Raynaud disease, hypothyroidism, SIADH and migraine with aura with left sided facial paresthesias. She has prior cholecystectomy for cholelithiasis. Her current medications included hyoscyamine, levothyroxine, meloxicam, omeprazole, sumatriptan, nizatidine, estradiol vaginal cream, estradiol tablet, and norethindrone.

Because the prior CT abdomen demonstrated the cyst abutting the ascending colon with mildly thickened wall, colonoscopy was done and showed a normal colon and terminal ileum, a few diverticula and small benign polyp in the rectal vault. Upper endoscopy revealed mild gastritis.

MRI of the abdomen (three months after the cyst was drained) reports a 7.1 x 7.4 x 11.4 cm cystic mass, with a thin internal septation without thickened components or mural nodularity.

Patient was referred to an oncological surgeon for evaluation and removal of the cystic mass. The surgery was done laparoscopically with a left sided approach, and a large cystic mass arising from the mesentery of the right colon near the hepatic flexure was removed.

Pathology reported mucinous cystadenoma of the mesentery (3.2 cm in size). The remaining tissue showed features of a mesothelial inclusion cyst, with reactive lymphocytic aggregates present within the cyst wall. The pathology was negative for atypia, borderline features and malignancy, and there are no features of lymphangioma. However portions of the cyst showed surrounding ovarian-like stroma, and a mesothelial lining consistent with a mesothelial inclusion cyst.

Immunohistochemical results were positive for CK7, negative for CK20 and CDX2, ER and PR positive, PAX8 partial positive, AE1/AE3 and calretinin positive.

After surgery the patient reported improved appetite, and decreased abdominal discomfort.

Discussion

Mucinous cystic neoplasms are rare tumors that arise in the ovary and various extra-ovarian sites. These include kidneys, pancreas, gallbladder, lungs, liver, appendix, mesentery, and mesocolon, with an estimated incidence of 1 in 27,000 to 1 in 250,000.[1,2] Mucinous cystadenomas most commonly occur in women up to the sixth decades of life, and rarely occur in extremes of age.[3] These tumors contain mucin producing cells thought to develop from inclusions of mesothelial cells or from ectopic ovarian nidus with subsequent mucinous metaplastic transformation of the lining cells to a cystadenoma promoted by estrogen receptors.[4,5] The current classification is based on histopathological features, and includes the 6 following groups: (1) cysts of lymphatic origin: simple lymphatic cyst and lymphangioma; (2) cysts of mesothelial origin: simple mesothelial cyst, benign cystic mesothelioma, and malignant cystic mesothelioma; (3) cysts of enteric origin: enteric cyst and enteric duplication cyst; (4) cysts of urogenital origin; (5) mature cystic teratoma (dermoid cysts), and (6) pseudocysts: infectious and traumatic cysts.[6]

Mucinous cysts of mesentery are also graded as benign, borderline, and malignant neoplasms.[1]

Immunohistochemistry is valuable to identify these neoplasms, with the epithelial cells having an immunophenotype of CK7 and CEA positivity and the stromal cells expressing ER and PR.[7] Incompletely excised and neoplasms which are borderline and beyond need close follow up as recurrences are known.[7]

Presentation of mesenteric cysts is heterogenous, with some found incidentally on imaging, and others presenting with symptoms including abdominal pain, anorexia, nausea, vomiting, or changes in bowel habits.

CT and abdominal ultrasound are indicated for diagnosis. However, these tumors can be difficult to differentiate as cystic masses versus solid tumors on pre-operative imaging.

Most mesenteric cyst are removed surgically either via laparoscope or open procedure. Cyst aspiration has high rates of recurrence,[8] as with this patient.

Complications of the mesenteric cyst include rupture, torsion, infection, malignant transformation or intestinal obstruction.[8]

Conclusion

Mucinous cystadenomas are rare tumors originating in the ovary and extra-ovarian sites. They are difficult to diagnose, and prompt diagnosis and treatment decreases the risk of complications, such as rupture, torsion, infection, malignant transformation or intestinal obstruction.

Most mesenteric cyst are removed either via laparoscopic or open procedure, as cyst aspiration has high rates of recurrence.

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