Journal of Surgical Arts
ISSN NO: 1308-0709 Cerrahi Sanatlar Dergisi
Year 2018, Volume 11, Issue 2

Case report

Extraovarian primary retroperitoneal mucinous cystadenocarcinoma;Case report

Ekstraovarian yerleşimli primer retroperitoneal müsinöz kistadenokarsinom; Olgu sunumu.

Arslani Nuhi[1], Silvio Altarac[2], Nexhibe Nuhii[3]

Department of Abdominal and General Surgery[1], University Medical Center, Maribor, Ljubljanska 5, 2000 Maribor, Slovenia. Faculty of Medicine Maribor[2], University of Maribor, Slomskov trg 15, 2000 Maribor, Slovenia. Faculty of Medicine[3], University of Tetovo, Street Ilinden bb, 1200 Tetovo, Macedonia

Corresponding address: Dr. Arslani Nuhi, arslani.nuhi@gmail.com

J Surg Arts (Cer San D), 2018;11(2):39-42.

ABSTRACT

Extraovarian primary retroperitoneal mucinous cystadenocarcinoma is an exceptionally rare malignant tumor.

We report a case of a 71-year-old Caucaisan female with 2 week history of abdominal tenderness and mild pain in lower abdomen. Radiographic imaging showed an enormous cystic tumor of unknown origin in the left retroperitoneum. The patient was prepared for surgical removal of the cystic growth, presumably an ovarian tumor. Laparotomy revealed a large cystic tumor covered with omentum. The cyst was punctured, thus facilitating a complete removal of the tumor. Adjuvant hysterectomy with bilateral salpingo-oophorectomy was performed, due to growth's proximity to the left ovary. Histological examination unveiled that the removed tissue is an extraovarian retroperitoneal primary mucinous multicystic adenocarcinoma.

We present this case since primary retroperitoneal cystadenocarcinomas are diagnostically challenging and there is no available evidence based guidelines concerning treatment, especially neoadjuvant chemotherapy and adjuvant hysterectomy seem to be controversial.

Keywords: Cystadenocarcinoma, mucinous, extraovarian, retroperitoneal, neoplasm.

ÖZET

Over dışı retroperitoneal musinöz kistadekoarsinomlar oldukça nadir görülen malign tümörlerdir.

Beyaz, 71 yaşındaki bayan hasta, karın alt kısmında iki hafta önce başlayan karın ağrısı ve gerginlik şikayeti ile müracaat etti. Radyolojik görüntülemede sol altta orijini belirlenemeyen büyük bir kistik lezyon saptandı. Hasta muhtemel over kökenli bir lezyon öntanısı ile ameliyata hazırlandı. Laparotomide etrafı omentum ile çevrili büyük kistik tümör saptandı. Kist içeriği iğne ile aspire edilerek boşaltıldı. Lezyonun sol overe uzanması nedeniyle bilateral salpingo ooferektomi ve histerektomi eklendi. Histolojik inceleme sonucunda retroperitoneal kaynaklı primer müsinöz kistadenokarsinoma tanısı konuldu.

Burada nadir görülen primer retroperitoneal müsinöz kistadenokarsinom hastalığı ve kanıta dayalı tedavi modaliteleri tartışılmıştır.

Anahtar kelimeler: Kistadenokarsinoma, müsinöz, ekstraovarian, retroperitoneal, neoplazi.

INTRODUCTION

Mucinous tumors are a common histological type of gynecological neoplasms that occurs especially in the ovaries. They represent approximately 25% of all ovarian neoplasms (1). However, their occurrence in retroperitoneum is exceptional. These primary retroperitoneal mucinous (PRM) cystic tumors can be classified as cystadenoma, cystadenoma with borderline malignancy and cystadenocarcinoma. According to the English literature, PRM cystadenomas are the most common type with 48 reported cases, followed by 27 cases of PRM cystadenocarcinoma and only 10 cases with borderline malignancy (2). PRM tumors are mostly unilateral, between 7-30 cm in size, and have noticeable female predominance with approximately 88% (2,3). The median age at diagnosis is 42 years and ranges from 17-86 years (4)

Primary retroperitoneal tumors usually present with nonspecific abdominal pain, discomfort or fullness and a palpable abdominal mass. The extent of these lesions can produce obstructive symptoms by theexertion of pressure on adjacent organs. Preoperative diagnosis is often difficult because standard radiological imaging (computed tomography, magnetic resonance imaging, and sonography) is usually not adequate to distinguish their exact origin and nature. Definitive diagnosis is obtained after histologic examination.

The first choice of treatment is radical surgery with total resection and careful inspection of possible primitive origins. Due to the rarity, there is no evidence of benefit from chemotherapy. Likewise, the role of adjuvant chemotherapy remains uncertain.

Case

A 71-year-old female with 2 week history of abdominal tenderness and mild pain in lower abdomen was referred to the department of gynecology. Additionally, the patient had noticed orthopnea, general malaise, and considerable weight loss of 8 kg in last year associated with food aversion. Clinical examination revealed palpable abdominal mass and thus, sonography was performed. Vaginal ultrasound revealed only basal segment of the growth, however, abdominal ultrasound showed approximately 30 cm large, cystic structure with proliferation in lower portions, which extended up to the xiphoid process. In addition, CT was performed. It disclosed a massive abdominal polymorphous tumor, which was partially solid, but mostly cystic with mixed dense and fatty content, along with some calcinations, as shown in Figure 1. Blood tests and serum tumor markers were within normal range.

Despite all diagnostic efforts, it was unable to determine the origin of the tumor growth, hence patient was planned for tumorectomy under the working hypothesis of a large ovarian tumor.

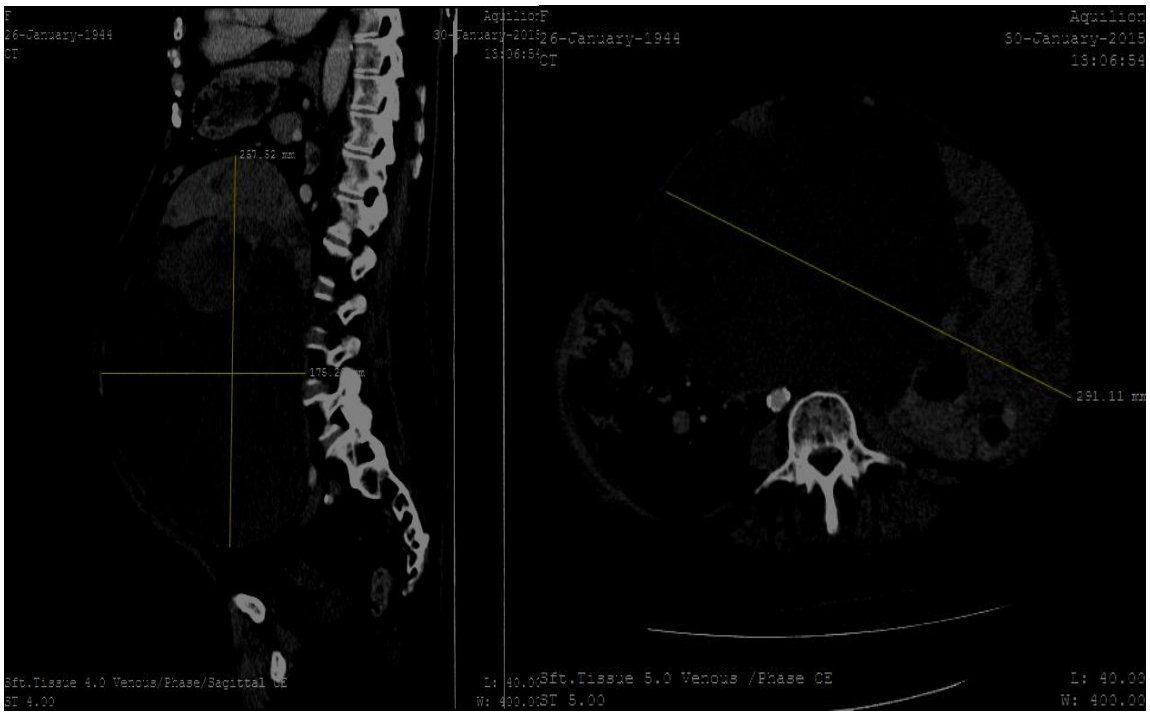

Figure 1: Sagittal and transverse CT scan in venous phase of the large abdominal polymorphous tumor.

Laparotomy revealed 30-40 cm large tumor covered with omentum. Surprisingly, uterus with adnexa, omentum, and intestinal surface were completely normal. The tumor was located in the left retroperitoneum, solitary and entirely separated from other structures. In relation to intraoperative findings, an abdominal surgeon was consulted during surgery. The growth was attached to the retroperitoneum subphrenically and laterally from mesocolon of the left intestine, which was pronated antemedially. Due

to the enormous size of the tumor, dissection was challenging (Figure 2). Operating team decided to puncture the cyst and managed to evacuate 3000 ml of brown fluid thus, facilitating successful removal of the tumor entirely. Due to growth's proximity to the left ovary, thesurgical team performed anadditional hysterectomy with bilateral salpingo-oophorectomy.

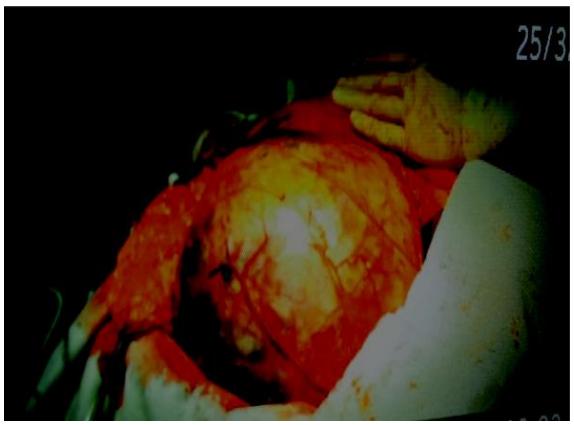

Figure 2: Exposed cystic tumor after laparotomy and removal of the omentum.

The excised emptied cystic growth weighed 490 g and measured 27 x 21 x 11,5 cm. Further histological examination has shown that the removed tissue could be extraovarian primary retroperitoneal teratoma with partial malignant transformation into invasive adenocarcinoma or metastatic adenocarcinoma with colorectal origin. In addition, CT and colonoscopy were performed, which revealed no colorectal pathology, thus discarding metastatic adenocarcinoma hypothesis. Two months after surgery a follow-up CT showed no residual pathology, with no abnormalities in liver nor lungs. Serum tumor markers were also within limits. The patient was presented at the oncological council, which abstained from any adjuvant chemotherapy.

Six months after surgery the patient's health condition gradually declined. A thorough examination revealed significantly elevated serum CEA (229 U/ml) and CA 125 (141 U/ml), additionally, CT indicated multiple round lesions in lungs and liver, all suspected metastases. The excised tissue was revised and described as an extraovarian retroperitoneal primary mucinous multicystic adenocarcinoma with partial necrosis. Immunohistochemical methods: HE stain, CEA+, CD68-, CK AE1/AE3+, B-Catenin +/-, CA 125 +/-, AMACR - (solitary cells +). Additionally, the case was once more reviewed at the oncological council, which refused any aggressive treatment and suggested palliative symptomatic care. The patient died nine months after operation with the advanced metastatic disease.

DISCUSSION

The retroperitoneal location is uncommon because of theabsence of epithelial cells in this area. Although the exact origin remains unclear, there are several theories explaining histogenesis of PRMC, including origination from teratoma, ectopic ovary, mucinous metaplasia of coelomatic epithelium, and enterogenous genesis (2,5,6). Hypothesis of heterotopic ovarian tissue etiology is based on the histological similarities, notwithstanding theoccurrence of malignancy in male patients and in afemale with normal ovaries. The second theory is based on ovarian or primary teratoma of retroperitoneum with displaced germ cells and mucinous epithelium obliterated all other components. Enterogenous genesis proposes intestinal duplication and is less persuasive. The hypothesis with gained acceptance is coelomic metaplasia, particularly invagination of the peritoneal mesothelium with subsequent mucinous metaplasia. During embriogenesis, embryonal coelomic epithetlium develops into peritoneal mesothelium, the germinal epithelium of the ovary, and the Müllerian duct. Peritoneal mesothelium retains the same potential for Müllerian differentiation as the epithelial tumor cells of the ovary. Therefore, PRMCAC share the same morphological and phenotypical characteristics as ovarian carcinomas (5,7-11).

Accurate preoperative diagnosis with standard imaging methods is troublesome. Computed tomo-graphy reveals the extension of the growth and mural calcifications, whereas magnetic resonance imaging can identify other structure involvement (2). Radiographic findings that suggest malignancy may include thickening and calcification of the cyst wall or mural nodules, further evidence of metastatic spread can be identified.

Benefit from aspiration cytology is poor. Neoplastic markers CEA, CA 19-9 and CA-125 are occasionally positive, despite they lack specificity (7). However, the values are usually normal. If increased, they can be used as anappraisal of responses to therapy or early regression detection (5). In our case, cytological examination of the punctured fluid did not provide any useful information, since it contained only necrotic cellular debris. In addition, serum tumor markers proved unreliable, since they rose only after metastatic dissemination.

Differential diagnoses include primary tumors, e.g. cystic teratoma, cystic mesothelioma, Müllerian cyst, epidermoid cyst, lymphocele, lymphangioma, pancreatic pseudocyst, and urinoma (2,3,6). Metastatic mucinous tumors originating from sites such as the ovaries, the intestines, and the pancreas must also be taken into consideration (12)

Adjuvant chemotherapy regarding our patient was abstained from since the excised tumor was at first considered to be an extraovarian primary retroperitoneal teratoma with no evidence of local or systemic invasion. At the time of second revision, the progression of the disease was so advanced, that adjuvant chemotherapy would be of little or no benefit at all. However, operating team in our case decided for additional hysterectomy with bilateral salpingo-

oophorectomy, due to growth's proximity to the left ovary.

Negative prognostic factors for the high risk of recurrence are presumed rupture of cystic mass, extracapsular invasion in adjacent organs, metastatic dissemination, and histopathological features of aggressive sarcoma-like or anaplastic components. Considering the rarity and short follow-up of PRMCAC, prognosis remains uncertain (3,5,13).

Considering its rarity, adequately aggressive treatment remains ambiguous. Also the benefits of comprehensive surgery with total hysterectomy, bilateral salpingo-oophorectomy and lymphadenectomy are not clearly defined. Most authors agree on tumor excision alone in thedesire to preserve fertility where ovaries and uterus appear uninvolved. It is generally accepted that if the uterus and ovaries are macroscopically normal, removal is not justified (3). Chemotherapy is favored in tumor invasion to adjacent structures, sarcoma or anaplastic nodules, likewise as in spontaneous or intraoperative rupture of the lesion (3,14) Despite aggressive adjuvant chemotherapy, biological behavior of some tumors is rapidly fatal (4).

REFERENCES

1. Crum CP. The female genital tract. In: Kumar V, Abbas AK, Fausto N (Eds). Robbins and Cotran pathologic basis of disease, 7th ed., Elsevier Saunders, Philadelphia, 2005, pp; 1059-117.
2. Knezevic S, Ignjatovic I, Lukic S, Matic S, Dugalic V, Knezevic D, et al. Primary retroperitoneal mucinous cystadenoma: A case report. World J Gastroenterol 2015;21(17): 5427-31.
3. Ramia JM, De la Plaza R, Arteaga V, et al. Primary Retroperitoneal Mucinous Cystadenoma with Sacromatous Carcinoma in Mural Nodule. Glob J Surg. 2013;1(1): 1-3.
4. Gilani SNS, Carr NJ, Mustajab A, Moran BJ. Primary retroperitoneal cystic mucinous carcinoma - case report and review of the literature. Case Rep Clin Pathol. 2015;2(3): 84-9.
5. Zanoni D, Sikokis A, Bersanelli M, Gnetti L, Buti S. Retroperitoneal cystoadenocarcinoma: a case report with a literature review. Acta Biomed. 2015;86(1): 97-105.
6. Thamboo TP, Sim R, Tan S-Y, Yap W-M. Primary retroperitoneal mucinous cystadenocarcinoma in a male patient. J Clin Pathol. 2006;59(6): 655-7.
7. Kamiyama H, Shimazu A, Makin Y, Ichikawa R, Hobo T, Arima S, et al. Report of a case: Retroperitoneal mucinous cystadenocarcinoma with rapid progression. Int J Surg Case Rep. 2015; doi: 10.1016/j.ijscr. 2015.04.004.
8. Chung DH, Lee SH. A case of primary retroperitoneal mucinous cystadenocarcinoma treated with fertility-sparing surgery. Korean J Obstet Gynecol. 2012;55(6): 424-8.
9. Chiang CH, Su CT, Chen LK, Chen CJ, Wu CC. Primary retroperitoneal mucinous cystadenocarcinoma in a twenty-nine year old pregnant woman. J Radiol Sci. 2012;37(4): 163-7.
10. Barka M, Mallat F, Mabrouk MB, Mrad C, Ahmed KB, Hmida W, et al. Primary retroperitoneal benign mucinous cystadenoma in a male. Int J Case Rep Images 2015;6(4): 198-202.
11. Elnemr A, Yonemura Y, Shinbo M, Nishino E. Primary retroperitoneal mullerian adenocarcinoma. Rare Tumors. 2010; doi:10.4081/rt. 2010.e6
12. Feng JF, Liu H, Chen DB. Primary retroperitoneal mucinous cystadenocarcinoma in a male patient: a rare case report. Hippokratia 2013;17(3): 271-3.
13. Heelan Gladden AA, Wohlauer M, McManus MC, Gajdos C. A Primary Retroperitoneal Mucinous Tumor. Case Rep Surg. 2015; doi: 10.1155/2015/157613
14. Hanhan HM, Gungorduk K, Ozdemir iA, Gokcu M, Sanci M, Ayaz D, Özeren M. Primary retroperitoneal mucinous cystadeno-carcinoma during pregnancy. J Obstet Gynaecol. 2014; 34(6):535-8.