Mature Cystic Teratoma Complicated with Pseudo - Meigs Syndrome and Elevated CA-125 Level in Puerperium: A Case Presentation and Review of the Literature

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This report presents a mature cystic teratoma complicated in puerperium with acute abdominal pain, diffuse ascites, hydrotorax and adnexial mass. A 42 years old multiparous patient, admitted to our clinic with abdominal distension, dyspnea, and diffuse abdominal pain. It was learnt that patient had normal spontaneous vaginal live birth 10 days earlier. There was a 6 cm sized cystic lesion with solid components originating from left adnexial area on pelvic ultrasonography. Besides, there was diffuse ascites. Abdominal CT revealed high probability for dermoid cyst or ovarian malignancy. CA-125 levels were increased. During laparotomy, 3000cc of ascites was drained and left salphingoopherectomy was performed. Frozen section result was benign. Exact pathology result was reported as mature cystic teratoma.

In puerperal period, benign ovarial pathologies should be kept in mind in patients presenting with diffuse ascites and ovarial mass.

Key Words: Mature cystic teratoma, Ascites, Pseudo-Meigs

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Introduction

New onset ascites in puerperium represents a very infrequent event.[1] Pseudo-Meig's syndrome is usually associated with ovarian cancer and other gynecologic malignancies.[2] An early diagnosis is extremely important, considering that some forms of ascites have benign causes and good prognosis, but others can be the expression of very aggressive diseases, potentially lethal if not early recognized and timely treated.[3] Elevation of CA-125 levels, ascites, hydrotorax are seen in this syndrome.[4] Interestingly, ascites and hydrotorax resolve with resection of the tumors.[5] In this report we described a case of benign mature cystic teratoma associated with ascites, hydrotorax, and elevation of CA-125 thereby mimicking an ovarian malignancy in puerperal period.

Case Report

A 42 year old woman was referred to the Selcuk University, Faculty of Meram Medicine, Department of Obstetrics and Gynecology, a tertiary care center, on postpartum 10th day with diagnosis of diffuse abdominal pain, adnexial mass and ascites. It was learnt that she had spontaneous vaginal birth 10 days earlier in another hospital and was discharged on postpartum first day without any complication. There was a history of ovarian cyst that was followed up throughout pregnancy. Her physical examination revealed expiratory wheezing, and a distended tender abdomen with positive fluid-wave transmission. In abdominal examination, there was diffuse distension and defense. In pelvic examination, cervical movements were painful, and there was vaginal discharge. No abdominal masses were palpable.

Laboratory investigations were normal, except for CA-125 levels that were elevated up to 113u/ml. CEA and CA19.9 values were within normal ranges and she had no other medical conditions. A posterioanterior plain chest radiography showed mild right pleural effusion. Transabdominal ultrasonography revealed a 63x62mm sized left adnexial mass and extensive ascites in abdomen. Contrast abdominal tomographic (CT) scan showed lacking lymphadenopathy, extensive ascites; a thin walled, 63x61mm mass with adipose density originating from the left ovary with high probability for dermoid cyst or ovarian cancer. Paracentesis was also performed and 3000cc serous fluid, consistent with an exudative process, was drained. Cytology from ascitic fluid was negative for malignancy.

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After preoperative preparations patient underwent exploratory laparotomy. The patient was noted to have a markedly distended abdomen, few liters of straw-colored ascites was evacuated upon entering the peritoneal cavity. The uterus was normal in size and a left 6x6 cm sized complex, multicystic mass, without evidence of external excrescence was noted. The right ovary appeared normal and there was no evidence of intraperitoneal spread of disease or retroperitoneal adenopathy. Frozen section of the left ovary was benign. A left salpingooophorectomy was performed. The final pathology was mature cystic teratoma. No areas of malignant transformation were identified on multiple sections of surgical specimen.

The patient was discharged home on sixth postoperative day. Her serum CA-125 levels decreased to normal levels in 2 weeks after surgery. And on control one month after operation, she had no evidence of pleural effusion and ascites.

Discussion

Pseudo Meigs syndrome is defined by the presence of a pleural effusion and ascites in association with a mature teratoma, leiomyoma, cystadenoma, or an ovarian malignancy.[5] And it can be combined with either benign or malignant neoplasms (Table 1).[5] In our case pseudo-Meigs syndrome was combined with a mature cystic teratoma.

Pseudo-meigs' syndrome in puerperium is very rare. In literature, there are 32 cases of pseudo-Meigs' syndrome associated with leiomyoma, and three other cases were associated with ovarian tumors.[6]

The etiology of the fluid accumulations remains unclear, although it appears to be related to lymphatic obstruction. The most likely pathogenesis of peritoneal and pleural effusions ascribes filtration of interstitial fluid in the peritoneum through the tumor capsule, and diffusion to the pleural space, usually at the right side, through diaphragmatic lymphatic vessels and apertures, as well as through intercellular gaps and small areas where muscular tissue of the diaphragm is replaced by areolar tissue.[7]

Gyang et al.[8] reported the case of a missed diagnosis of acute postpartum pancreatitis in a patient showing abdominal pain, pyrexia, anaemia and gross ascites few days after instrumental delivery, initially diagnosed as postpartum endometritis with peritonitis.

In rare cases, puerperal ascites is a sign of systemic transudation due to generalized increased capillary permeability. Kanda et al.[9] described the case of a postpartum capillary leak syndrome in a 29-year-old healthy woman that suddenly developed hypovolemic shock, anasarca, pleural effusion and ascites in the postpartum period.

Puerperal cardiac ascites has been exceptionally reported in subjects with negative cardio-vascular history. A case of right atrial myxoma presenting as postpartum ascites and fever of unknown origin was described.[10] In another case reported in the literature, a peripartum cardiac failure occurred in a patient affected by Graves' disease.[11] Few cases of urinary ascites in the postpartum period, due to a bladder perforation during caesarean section or to a spontaneous rupture of the bladder, without history of recent trauma or instrumentation have been reported.[12,13]

The majority of ovarian tumors, associated with hydrothorax and ascites, have a diameter of more than 6 cm. The entity of effusions can be moderate or massive. The effusions generally derive from a transudative process, but can occasionally contain blood cells. Their connection with the pelvic tumor is demonstrated by their regression after neoplasm removal.[14] In our case tumor diameter was 6 cm sized and after removal, we have seen regression on ascites and hydrotorax.

There also reported 6 cases of pseudo-Meigs syndrome caused by secondary ovarian tumors from gastrointestinal cancers. The primary site was the colon or rectum in 5 and the stomach in 1. Two cases were due to Krukenberg tumors. Three patients with documented outcomes were alive 108, 52 and 12 months after resections, demonstrating that in these cases resection provide long-term palliation.[15]

In the literature, there are reported unique, of special interest, cases of pseudo-Meigs syndrome caused by rare pathological conditions (Table 2).[16]

In classical pseudo Meigs' syndrome surgical excision of pelvic mass induces immediate resolution of the ascites and normalization of the serum CA-125 level. In our patient, two weeks after the surgery, her CA-125 levels were decreased to normal range and after one month; there were no signs of ascites. We gave some examples of pseudomeigs' syndrome and pregnancy associated cases and CA-125 levels before and after surgery below (Table 3).[17,18] Recently, elevations in serum CA 125, usually associated with malignant serous ovarian carcinoma, had been reported in cases with Meigs syndrome secondary to ovarian fibromas.[19,20] Immunohistochemical staining for CA 125 was localized to the omentum and peritoneal surfaces rather than the fibromas, Immunohistochemical studies suggest that serum elevation of CA-125 is likely to be caused by mesothelial expression of the antigen rather than by the tumor.[21]

In conclusion; in a patient with pelvic mass, ascites and hydrotorax; pseudomeigs' syndrome must be considered in differential diagnosis. The mechanism of ascites is unclear but probably it is a result of lymphatic obstruction. Higher CA-125 levels are expected. After all, in this syndrome surgical excision of pelvic mass induces immediate resolution of the ascites and normalization of the serum CA-125 level.

Puerperal Dönemde Pseudo-Meigs Sendromu ve CA-125 Yüksekliği ile Komplike Olan Bir Matür Kistik Teratom Vakası: Olgu Sunumu

Bu vakada, puerperal dönemde akut karın, yaygın asit, hidrotoraks ve adneksiyel kitle ile komplike olan bir matür kistik teratom vakası sunulmuştur. 42 yaşında multipar hasta, abdominal distansiyon, dispne, yaygın karın ağrısı şikâyetiyle kliniğimize başvurdu. Hastanın 10 gün önce dış bir merkezde normal spontan vajinal yol ile doğum yaptığı öğrenildi. Yapılan pelvik ultrasonografik incelemesinde sol adneksiyal alandan kaynaklanan yaklaşık 6 cm çapında solid alanlar ihtiva eden kistik lezyon tespit edildi. Batında yaygın asit saptandı. Abdominal CT sonucunda, sol overde ince duvarlı, yağ dansiteli kitle (dermoid kist?, over tm?) tespit edildi. CA-125 seviyesinin yüksek olduğu tespit edildi. Operasyon esnasında batından yaklaşık 3000 cc serbest asit mai boşaltıldı ve sol ooferektomi yapıldı. Frozen incelemesi benign idi. Hastanın postoperatif dönemde semptomları hızla geriledi ve postoperatif 6. günde taburcu edildi. Kesin patoloji sonucu matür kistik teratom olarak bildirildi.

Puerperal dönemde, benign ovaryen patolojiler batında yaygın asiti ve ovarial kitlesi olan hastalarda ayırıcı tanıda akılda tutulmalıdır.

Anahtar Kelimeler: Matür kistik teratom, Asit, Pseudo-Meigs

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