Meigs' Syndrome: a case presentation and revision of the literature

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Meigs’s syndrome is defined as the presence of ascites and hydrothorax in association with a benign ovarian tumor. It is a rare clinical entity, which is also considered to be an uncommon complication of benign leiomyomas of the female genital tract. The case of a 33 year-old female patient who presented rapid weight loss and a quickly increasing abdominal circumference is described. Clinical and ultrasonographic studies revealed a mobile, semi-solid right adnexal tumor in the lower abdominal quadrants of 15 x 14-cm and ascites as well as hydrothorax of the left lung, confirmed by chest radiography.

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Abstract
Meigs’s syndrome is defined as the presence of ascites and hydrothorax in association with a benign ovarian tumor. It is a rare clinical entity, which is also considered to be an uncommon complication of benign leiomyomas of the female genital tract. The case of a 33 year-old female patient who presented rapid weight loss and a quickly increasing abdominal circumference is described. Clinical and ultrasonographic studies revealed a mobile, semi-solid right adnexal tumor in the lower abdominal quadrants of 15 x 14-cm and ascites as well as hydrothorax of the left lung, confirmed by chest radiography. Meigs’s syndrome was diagnosed and surgical intervention successfully removed the peritoneal effusion and the right ovarian tumor. The hydrothorax disappeared 7 days after the intervention. Cytomorphologic studies of both the tumor and ascitic fluid was void of malignancy. The pathogenesis of the pleural and ascitic fluids and the importance of CA-125 are discussed.

KEYWORDS: Meigs’s syndrome / ovarian tumor / ascites / hydrothorax / pleural effusion

Introduction
Meigs' syndrome is defined as the presence of ascites hydrothorax associated with a benign ovarian tumor that disappears after the removal of the tumor. Joe Vincent Meigs (1892-1963), a professor of the Harvard Medical School of Gynecology drew attention to the syndrome, however it had been described previously by numerous authors in the nineteenth century and beginning the twentieth. In 1937 Meigs and Cass alerted the medical profession on the importance of the syndrome. Finally, in the same year, Rhodes and Terrell denominated it Meigs' syndrome.

Meigs' syndrome is a strange clinical entity that is also considered to be an uncommon complication of leiomyomas of the female genital tract. The more frequently observed type of ovarian tumor it is the leiomyoma. It is accepted that the uterine tumors such as the fibromas, although extremely rare, can be associated to ascites and hydrothorax, as well as leiomyoma of the broad ligament, leiomyoblastoma of the colon and cancer of the ovary. This condition is denominated pseudo-Meigs' syndrome, a situation in which it is necessary to discard the origin metastatico or primitive he/she gives the tumor, by means of thoracoscopy or cytological exam, prior to surgical treatment.

The presumptive diagnosis of this pathology is basically clinical in spite of the priceless value that
imaging techniques have: ultrasonography, computerized axial tomography (CAT) and magnetic resonance imaging (MRI), in confirming the presence of ascites, pleural effusion and the characteristics of the ovarian tumor. Cytomorphology of the pleural and ascitic liquid is also used as well as the dosification of the serum levels of carcinogenic antigen-125 (CA-125) to discard the malignancy of the ovarian tumor that is associated with the syndrome. The definitive diagnosis is usually postoperative; although there have been reports of the possibility of performing adequate preoperative diagnosis.3-6,9

All the signs and symptoms of Meigs' syndrome disappear with the surgical extraction of the ovarian tumor by means of the abdominal surgery,4-6 although Bretelle et al. have recently reported what may be the first case of recurrence of Meigs' syndrome after an initial surgical treatment.10

Case Presentation

A 33 year-old female patient, G2 P2, with history of good health who was remitted to the gynecology department of our Center for her admission presenting weight loss of approximately 10 kilograms in 2 months and a rapidly increasing abdominal circumference. The patient didn't present any other symptoms. On clinical examination a large abdominal tumor was found occupying the lower abdominal quadrants up to the navel, with certain mobility provoking pain in the patient. The presence of liquid in the abdominal cavity was clinically demonstrated. On gynecological examination, a uterus of normal form, size and consistency, the left adnexum was without alteration on examination. The right adnexum presented a mobile, semi-solid tumor with a smooth surface measuring approximately 12 x 14-cm, unadhered to neighboring organs. Slight pain was caused on its mobilization. The presumptive diagnosis on admission was giant cyst of the right ovary and ascites. A complete hematological study was carried out, with results within normal parameters. The sonographic study of the superior hemiabdomen showed the presence of supradiaphragmatic liquid (corresponding with hydrothorax), liquid in the abdominal cavity, as well as a right adnexial tumor of 16 x 18-cm. On radiological examination, pleural effusion is observed. Paracentesis and thoracocentesis were carried out for the extraction of liquid for cytomorphicologic study that was negative of malignancy for both samples. The patient began to present dyspnea and slight affectation of her general well being, reason for which surgical intervention was decided on with the presumptive diagnosis of Meigs' syndrome. Findings on laparotomy were: 4000cc of ascitic effusion, a large tumor of the left ovary measuring 18 x 20-centimeters. Exeresis of the tumor and total abdominal hysterectomy with omentectomy were performed. The cytomorphicologic study of the ascitic effusion and the tumor confirmed the absence of malignancy. The study also identified the tumor as ovarian fibroma. Postoperative progress was favorable, with remission of the hydrothorax by the seventh postoperative day. The patient was discharged from our Center after 20 days.

Discussion

The pleurae are a structure of mesodermic origin that consists of two layers, denominated visceral and parietal layers. Both pleural layers unite in the base of the lung, leaving a space between each called the pleural cavity. A continuous process exists of filtration of liquid from the capillary vessels to the space subpleural, and give there to the cavity pleural. This process depends on the balance of hydrostatic and coloidosmotic pressures in both spaces, according to Starling's law, giving place to the presence of a minimal quantity of liquid in the physiologic virtual cavity.11 The pleural effusion in the cases of ovarian tumors, usually corresponds to an exudate because the liquid moves from the peritoneal cavity to the pleural cavity through diaphragmatic defects or lymphatic channels. It is generally located in the right and can be massive on occasions, with biochemical or cellular unspecific characteristics of the liquid. Statistically, 70% of para-ascitic effusions are on the right, 15% on the left and 15% are bilateral.11,12

A possible pathogenesis for the formation of the pleural and peritoneal effusions in the case of ovarian tumors can be explained by the filtration of interstitial liquid to the peritoneum through the ovarian tumor capsule. This liquid can also diffuse to the space pleural through the diaphragmatic lymphatic vessels in the Bochdalek's foramen or through diaphragmatic defects.11

The cytological exam of the ascitic and pleural liquid in patients with ovarian tumors should be
performed in order to differentiate between reactive processes and tumor spread. Although the detection of malignant cells is a marker of malignant disease and a sign of poor prognosis, the benign effusions don't affect neither the stage of the disease nor the prognosis of the patient. Some authors emphasize that an ovarian mass with pleural and abdominal effusion doesn't always represent an advanced stage malignancy, not even in presence of high serum levels of CA-125.

In the evaluation of the presented patient, we did not use this tumor marker. However, we believe that the CA-125 deserves special attention for its role in the preoperative evaluation of women with pelvic masses. CA-125 was originally described as tumor marker for ovarian cancer and it is used in the screening of this pathology. At present its elevation (above 35 mIU/mL) is associates to several benign conditions as: endometriosis, leiomyoma, pregnancy, pelvic inflammatory disease, as well as diverse malignancies. Several authors have pointed out that CA-125 serum levels rise in Meigs´ syndrome and they suggest suspicion of the syndrome in the presence of any important pleural effusion, even high serum levels of CA-125, a negative cytological study of the ascitic liquid and the absence of peritoneal implants at CAT-scan. Other researchers argue for the systematic use of CA-125 in the preoperative evaluation of women with pelvic masses to allow for the differentiation of the malignant ones from the benign.

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