Meigs Syndrome: Case Report

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Síndrome de Meigs: Relato de Caso

Síndrome de Meigs: Informe de Caso

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ABSTRACT

Introduction: Meigs syndrome is a rare clinical condition, defined as the association of pleural effusion, ascites and ovarian fibroma, with resolution of symptoms after tumor resection. **Case report:** Female patient, 56 years old, with dry cough associated with hyporexia, weight loss and progressive dyspnea for one month. Chest X-ray and later chest tomography showed massive pleural effusion on the right. Thoracocentesis was performed with drainage of 2,500 ml of serous fluid, suggestive of exudate. On examination, a palpable mass was observed in the hypogastrium, with an upper limit in the umbilicus. Imaging exams show solid expansive formation of possible left ovarian origin and presence of ascitic fluid. The patient underwent total hysterectomy with bilateral salpingo-oophorectomy and resection of the pelvic mass. Intraoperatively, frozen section was suggestive of ovarian fibroma. Histopathological of the surgical specimen confirmed ovarian fibroma measuring 13.0 x 12.5 x 7.5 cm and cytopathological examination of the ascitic fluid was negative for neoplastic cells. The patient evolved in good general condition with resolution of the pleural effusion and ascites and continues without recurrence of symptoms. **Conclusion:** The definitive diagnosis is made by histological confirmation of ovarian fibroma and resolution of symptoms after removal of the tumor. Dyspnea may be the initial symptom and the CA-125 may be elevated. The prognosis is usually good and the chances of recurrence are minimal.

Key words: Meigs syndrome; fibroma; ascites; pleural effusion; surgical oncology.

RESUMO

Introdução: A síndrome de Meigs é uma condição clínica rara, definida como a associação de derrame pleural, ascite e fibroma ovariano, com resolução dos sintomas após a ressecção do tumor. **Relato do caso:** Paciente, sexo feminino, 56 anos, com tosse seca, associada à hiporexia, à perda de peso, à dispineia progressiva durante um mês. Radiografia de tórax e posteriormente tomografia de tórax mostraram derrame pleural volumoso à direita, sendo realizada toracocentese com drenagem de 2.500 ml de líquido seroso, sugestivo de exsudato. Ao exame, observou-se massa palpável em hipogástrio, com limite superior em cicatriz umbilical. Exames de imagem mostram formação expansiva sólida de possível origem ovariana esquerda e presença de líquido ascítico. A paciente foi submetida à histerectomia total com salpingo-ooforectomia bilateral e ressecção da massa pélvica. No intraoperatório, o exame por congelação foi sugestivo de fibroma ovariano. O histopatológico da peça cirúrgica confirmou fibroma ovariano medindo 13,0 x 12,5 x 7,5 cm e o exame citopatológico do líquido ascítico foi negativo para células neoplásicas. A paciente evoluiu em bom estado geral com resolução do derrame pleural e da ascite e segue sem recorrência dos sintomas. **Conclusão:** O diagnóstico definitivo é feito pela confirmação histológica de fibroma ovariano e resolução dos sintomas após a remoção da tumoração. A dispneia pode ser o sintoma inicial e o marcador tumoral CA-125 pode estar elevado. O prognóstico costuma ser bom e as chances de recidiva são mínimas.

Palavras-chave: síndrome de Meigs; fibroma; ascite; derrame pleural; oncologia cirúrgica.

RESUMEN

Introducción: El síndrome de Meigs es una condición clínica rara, definida como la asociación de derrame pleural, ascitis y fibroma de ovario, con resolución de los síntomas después de la resección del tumor. **Informe del caso:** Paciente femenino de 56 años con tos seca asociada a hiporexia, pérdida de peso y disnea progresiva durante 1 mes de evolución. La radiografía de tórax y posterior tomografía de tórax mostró derrame pleural masivo en el lado derecho, se realizó toracocentesis con drenaje de 2.500 ml de líquido seroso, sugestivo de exudado. A la exploración se observa una masa palpable en hipogastrio, con límite superior en ombligo. Los exámenes de imagen muestran formación sólida expansiva de posible origen ovárico izquierdo y presencia de líquido ascítico. La paciente fue sometida a histerectomía total con salpingooforectomía bilateral y resección de la masa pélvica. Intraoperatoriamente, sección congelada sugestiva de fibroma de ovario. El histopatológico de la pieza quirúrgica confirmó fibroma de ovario de 13,0 x 12,5 x 7,5 cm y el examen citopatológico del líquido ascítico fue negativo para células neoplásicas. El paciente evolucionó en buen estado general con resolución del derrame pleural y ascitis y continúa sin recidiva de los síntomas. **Conclusión:** El diagnóstico definitivo se realiza mediante la confirmación histológica del fibroma de ovario y la resolución de los síntomas tras la extirpación del tumor. La disnea puede ser el síntoma inicial y el CA-125 puede estar elevado. El pronóstico suele ser bueno y las posibilidades de recurrencia son mínimas.

Palabras clave: síndrome de Meigs; fibroma; ascitis; derrame pleural; oncología quirúrgica.

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INTRODUCTION

Meigs syndrome is a rare clinical condition, defined as the association of pleural effusion, ascites and ovarian fibroma, with symptom resolution after tumor resection[1]. Currently, true Meigs syndrome is characterized by the presence of four clinical criteria: 1) primary fibroma tumor or fibroma-like tumor (tecoma, granulosa cell tumor or Brenner's tumor); 2) with ascites; 3) with pleural effusion; and 4) pleural and ascitic fluids disappear with tumor excision and do not accumulate again[2,3]. Other benign ovarian cysts (such as *Struma ovarii*, mucinous cystadenoma and teratomas), uterine fibroids and secondary metastatic tumors to the ovary, associated with ascites and/or hydrothorax, characterize pseudo-Meigs syndrome[3,4]. Pseudo Meigs syndrome, or Tjalma syndrome, is a rare manifestation of systemic lupus erythematosus (SLE), defined by the presence of ascites, pleural effusion and elevated levels of CA-125, after ruling out benign or malignant tumors[5].

The incidence of Meigs syndrome is difficult to estimate. It is believed that the incidence of ovarian sex cord stromal tumors is 0.20 per 100,000 women annually and, of these, only 1% had Meigs syndrome[1,4]. The presumptive diagnosis of Meigs syndrome is made clinically. The most common symptoms are dyspnea (32%) and abdominal distension (5%) [4,6]. Among the differential diagnoses, one should investigate malignant ovarian tumors, other types of cancer, including bowel and lung, nephrotic syndrome, congestive heart failure, liver cirrhosis and tuberculosis[4].

This study was approved by the Research Ethics Committee (REC) of the Walter Cantídio University Hospital under opinion number: 5,913,107 (CAAE: 67291023.2.0000.5045), according to Resolution nº. 466, of 2012, of the National Health Council[7].

CASE REPORT

Patient, female, 56 years old, brown, with a history of flu for one month. In September 2022, she had a dry cough without blood or mucus, associated with hyporexia, weight loss and progressive dyspnea. In October 2022, she went to the emergency service where the chest radiograph (Figure 1) and later the chest tomography showed voluminous pleural effusion on the right (volume estimated at 3,000 ml), determining deviation of the mediastinal structures to the left and volumetric reduction of the left lung, due to compressive effect. She underwent thoracentesis with drainage of 2,500 ml of serous fluid and a cycle of antibiotic therapy with ceftriaxone. Laboratory study of new thoracentesis showed result suggestive of exudate. Test for acid-alcohol resistant bacilli (AFB) and rapid molecular test negative for tuberculosis. In November, she was referred to the service due to the persistence of dyspnea. Thoracoscopy was performed with pleural biopsy and pleural drainage with outflow of 3,500 ml of citrus yellow fluid, with improvement of lung expandability and respiratory pattern. Pleural biopsy showed chronic and suppurative pleuritis associated with fibrosis, sclerosis and focal hemorrhage, with no signs of malignancy.

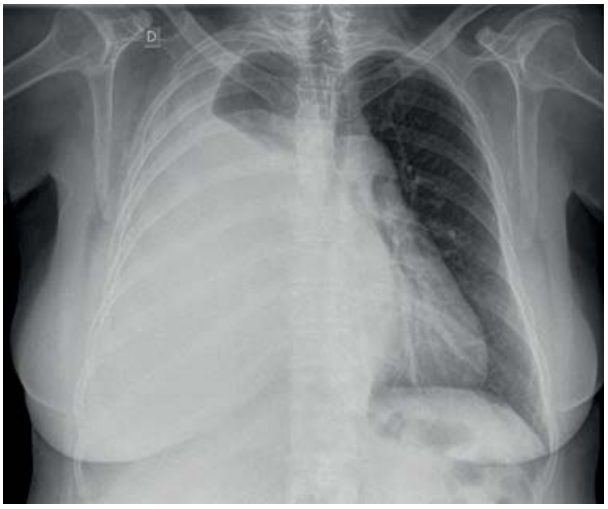

Figure 1. Posteroanterior chest X-ray showing extensive pleural effusion on the right

During hospitalization, a painful abdomen was observed on deep palpation in the lower abdomen associated with the palpable mass in the hypogastrium, with an upper limit in umbilical scar. Abdominal ultrasound showed a moderate amount of peri-hepatic free fluid and in the lower abdominal cavity, in addition to a hypoechoic mass with heterogeneous echotexture, with regular contours and defined limits, located in the lower abdomen on the right in the pelvis, measuring about 13.1 x 8.8 cm. Computed tomography showed a large expansive lesion centered on the supravesical pelvic region measuring 14.1 x 8.6 x 12.1 cm with an estimated volume of 770 cm^3, indeterminate as to anatomical origin (Figure 2). Then, magnetic resonance imaging indicated a largesolid expansive formation, with heterogeneous contrast enhancement, located in the middle abdominopelvic transition, superiorly to the urinary bladder and anteriorly to the uterus, of possible left ovarian origin, without invasion of adjacent structures, measuring about 14.7 x 14.6 x 9.0 cm, in addition to small volume ascites (Figure 3). Tumor markers CA-125: > 500; CA 19-9: 2.5 and CEA: 0.2 were requested, and surgical approach of the tumor was scheduled.

In December 2022, she underwent total hysterectomy with bilateral salpingo-oophorectomy and resection of

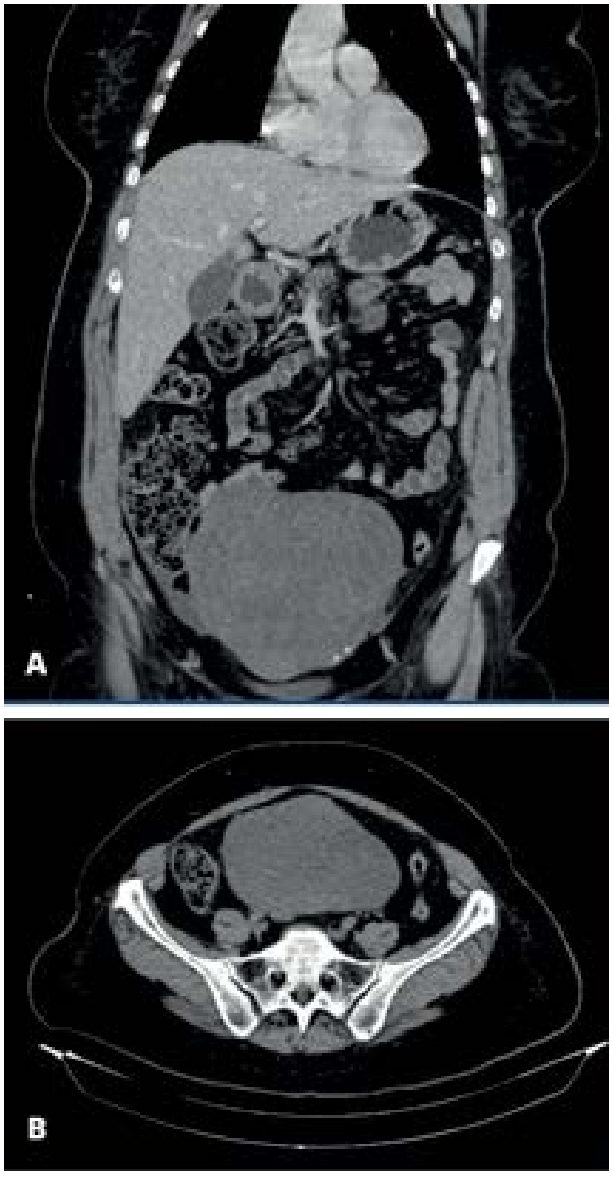

Figure 2. Computed tomography images. A) Coronal section showing solid, hypovascular tumor with some isolated peripheral foci of calcification. B) Axial cut showing bulky lesion in the pelvic region

the pelvic mass. This procedure was performed through a median infraumbilical incision, in which the presence of moderate ascitic fluid with citrus appearance, large right adnexal lesion of solid content, absence of hepatic or peritoneal implants and other organs without changes were observed. Intraoperative freezing examination was suggestive of fibroma. The histopathology of the surgical specimen confirmed ovarian fibroma measuring 13.0 x 12.5 x 7.5 cm and the cytopathological examination of the ascitic fluid was negative for neoplastic cells. The patient progressed in good general condition with resolution of pleural effusion and ascites and continues without recurrence of symptoms.

DISCUSSION

Ovarian fibroma is a benign tumor that arises from the stromal component of the ovary. It is composed of spindle-shaped or oval fibroblasts similar to collagen-producing cells. Among sex cord stromal tumors, ovarian fibroma is the most common tumor, representing 4% of all ovarian tumors and occurs most commonly in women in the fourth decade of life[8]. They usually present as palpable pelvic or adnexal masses with an average size of 8 cm, however, they can be asymptomatic, as about one third of tumors can be less than 3cm[8,9]. They are rarely associated with the production of hormones such as estrogen. Larger tumors may cause pelvic pain or acute abdominal pain due to ovarian torsion. In general, they present as hypoechoic masses on ultrasound, demonstrate homogeneous delayed enhancement on tomography, hyposignal on T1 and T2 images, delayed enhancement on post-contrast magnetic resonance imaging[9].

The pathophysiology behind the formation of pleural effusion and ascites in Meigs syndrome has not yet been fully understood. Several hypotheses have been suggested to explain this phenomenon[10]. It is believed that peritoneal

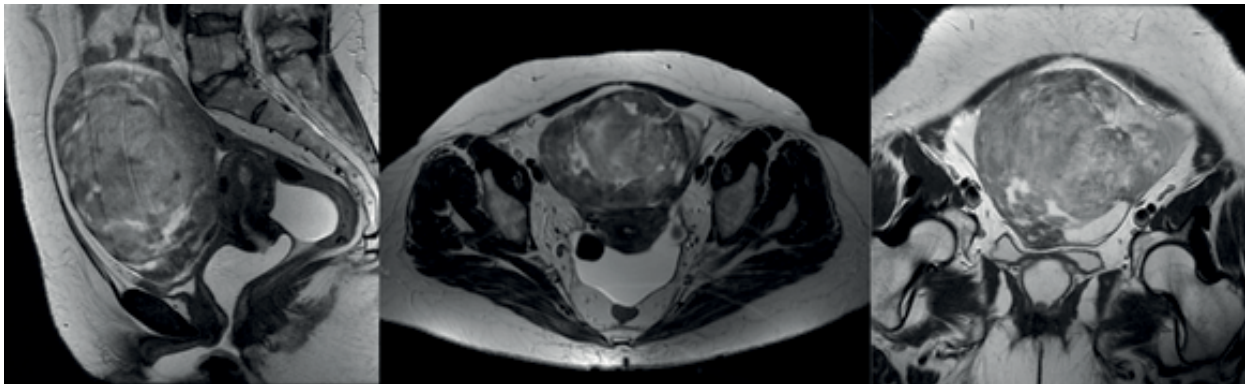

Figure 3. Coronal and axial T2 magnetic resonance images showing a large solid lesion, located in the abdominal-pelvic transition, superiorly to the urinary bladder and anteriorly to the uterus without invasion of adjacent structures

fluid formation may be linked to increased vascular permeability and capillary leakage mediated by the release of inflammatory cytokines and growth factors[11]. In some patients, high levels of vascular endothelial growth factor, fibroblast growth factor and interleukin-6 were found in the blood, pleural fluid and ascitic fluid, with a significant decrease after tumor removal, suggesting local production by the tumor[6]. Another theory proposes that the pressure exerted by the tumor itself on the pelvic and abdominal lymphatic vessels hinders lymphatic drainage, resulting in intraperitoneal accumulation of fluid[10]. It has also been proposed that ascites can be attributed to stromal tumor edema (edematous fibroids) and interstitial fluid transudation, resulting in fluid leakage[10,11]. Possibly, the pathogenesis of ascites in Meigs syndrome is related to a combination of these mechanisms and the characteristics of the ascites fluid depend on the relative contribution of each of them6.

In Meigs syndrome, it is believed that pleural effusion may be secondary to the passage of ascitic fluid into the pleural space through the diaphragm, because of congenital defects (diaphragmatic pores) that tend to be more common on the right side, or through the diaphragmatic lymphatic channels[1,6,10]. In a systematic review, Krenke et al.[6] evaluated the characteristics of pleural fluid in patients with Meigs syndrome and analyzed the prevalence of transudative and exudative pleural effusion. In patients with classical Meigs syndrome, pleural effusion was most commonly seen on the right side (70.3%) and the mean total volume of drained pleural fluid was 2,500 ml, ranging between 1,500 and 4,865 ml. A total of 72% of the patients had exudative pleural effusion, and 66% of the ascitic fluid analyzed was classified as exudative. The prevalence of exudative effusion in patients with Meigs syndrome corroborates the most accepted pathophysiological mechanisms involved in the accumulation of pleural fluid. However, transudative pleural effusion does not exclude the diagnosis of Meigs syndrome[6].

Elevated CA-125 is generally associated with epithelial ovarian cancer, however, this marker has low specificity and may be related to many other malignant or benign conditions, such as uterine fibroids, ascites, inflammation of the peritoneum or pleura, endometriosis, liver cirrhosis and pelvic inflammatory disease[5,12]. In Meigs syndrome, CA-125 may be significantly increased, although levels above 1,000 have rarely been reported[1]. This CA-125 serum increase may be related to increased intraperitoneal pressure caused by ascites and mesothelial cellirritation[8,11]. Some studies cited by Nguyen et al.[1] showed that high levels of CA-125 are associated with higher ascites volume, however, tumor size was not proportionally related to CA-125 levels.

Management of Meigs syndrome involves paracentesis and thoracentesis for relief of symptoms caused by ascites and pleural effusion, respectively. The treatment of choice is exploratory laparotomy with surgery and staging. Intraoperatively, freeze biopsy may be performed to confirm ovarian fibroma. The choice of surgical technique will often depend on the patient's age. Women of reproductive age can undergo unilateral salpingo-oophorectomy, whereas postmenopausal women undergo total abdominal hysterectomy with bilateral salpingo-oophorectomy[4]. In pediatric patients, preservation of the affected ovary may be attempted, given the benign nature of the fibroma and the minimal recurrence rate after excision, unless the mass cannot be separated from the ovary. The definitive diagnosis of Meigs syndrome is postoperative with resolution of ascites and pleural effusion, and histological confirmation of the tumor[11,12]. When well conducted, cases of Meigs syndrome have a good prognosis and, after tumor resection, life expectancy is equal to that of the general population[4].

CONCLUSION

Meigs syndrome is a rare clinical entity, but should be considered as a differential diagnosis in patients with adnexal masses, pleural effusion and ascites. Definitive diagnosis is made by histological confirmation of ovarian or fibroma-like fibroma and resolution of symptoms after surgical removal of the tumor. Dyspnea may be the main initial symptom in this type of patient and CA-125 may be elevated, even if it is a benign neoplasm. The prognosis is usually good and the chances of relapse are minimal.

CONTRIBUTIONS

All authors contributed substantially in the design and/or planning of the study; in the collection, analysis and interpretation of data; in the writing and critical review; and approved the final version to be published.

DECLARATION OF CONFLICT OF INTERESTS

There is no conflict of interest to declare.

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