

Pseudo-Meigs Syndrome: A Case Report

C Papanikolaou, K Fortounis, S Ainalis, K Biba, A Papanikolaou, G Hatzitheoxaris

Citation

C Papanikolaou, K Fortounis, S Ainalis, K Biba, A Papanikolaou, G Hatzitheoxaris. *Pseudo-Meigs Syndrome: A Case Report*. The Internet Journal of Pathology. 2004 Volume 4 Number 2.

Abstract

Meigs syndrome is defined as the co-existence of benign ovarian fibroma, hydrothorax and ascites. On the contrary, Pseudo-Meigs syndrome is characterized by the co-existence of hydrothorax, ascites and other ovarian- usually malignant-or pelvic tumors.

The case herein presented concerns a 54 year old postmenopausal woman with recurrent, since 1 year, perineal phlegmon and currently established respiratory distress. Her clinical and radiological examination revealed a massive right pleural effusion, mild ascites and a large heterogeneous, with solid and cystic components, pelvic tumor, measured 15x15cm, causing pressure to the bladder. After a preoperative relieving paracentesis and drainage of the pleural effusion-with negative for malignant cells cytologic examination, the patient underwent an exploratory laparotomy with excision of both the tumor and the right ovary. The tumor was diagnosed histologically as an ovarian well differentiated endometrioid carcinoma. The immediate postoperative resolution of hydrothorax and ascites confirmed the diagnosis of pseudo-Meigs syndrome. The patient followed chemotherapy and remains in good condition 18 months after surgery, without any indication of metastatic disease.

INTRODUCTION

The co-existence of pelvic tumor, hydrothorax and ascites has been known since the late 19th century. The features of the disease were described by Meigs and Cass in 1937. In the same year Roads named it "Meigs syndrome". Today, Meigs syndrome is defined as the co-existence of benign ovarian fibroma, hydrothorax and ascites. On the contrary, Pseudo-Meigs syndrome is characterized by the co-existence of hydrothorax, ascites and other ovarian- usually malignant-or pelvic tumors. Both these syndromes should be considered in otherwise healthy postmenopausal women, who present with either new or recurrent hydrothorax and ascites. The preoperative differential diagnosis between them is useless, since the surgical resection of the tumor is the only therapeutic choice, resulting to the resolution of fluid accumulations in both situations.

CASE REPORT

A 54 year old postmenopausal woman admitted in our Clinic because of recurrent, since 1 year, perineal phlegmon and currently established respiratory distress.

The clinical examination revealed sinus tachycardia, absence

of breath sounds at the auscultation of the right hemithorax, perineal redness and a palpable mass of the lower abdomen. A massive right pleural effusion was found at chest x-ray (Fig. 1).

Figure 1

Figure 1 : Massive fluid effusion of the right pleural cavity



Computed tomography of the abdomen demonstrated mild ascites and a large heterogeneous, with solid and cystic components, pelvic tumor, measured 15x15cm, causing

pressure to the bladder (Fig. 2A, B).

Figure 2

Figure 2A: Mild ascites

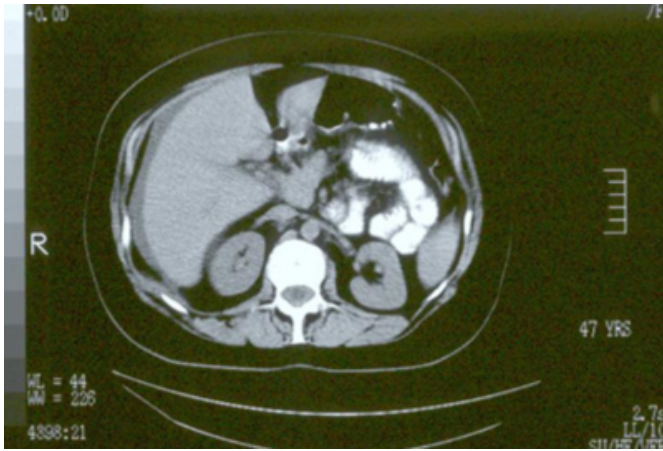
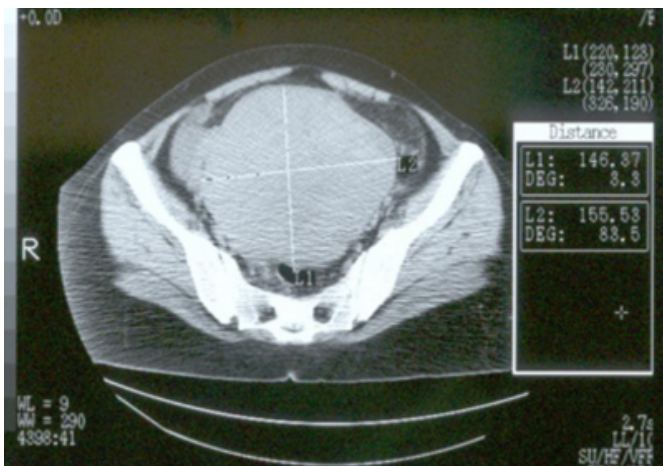


Figure 3

Figure 2B: Large heterogeneous, with solid and cystic components, pelvic tumor measured 15x15cm and causing pressure to the bladder



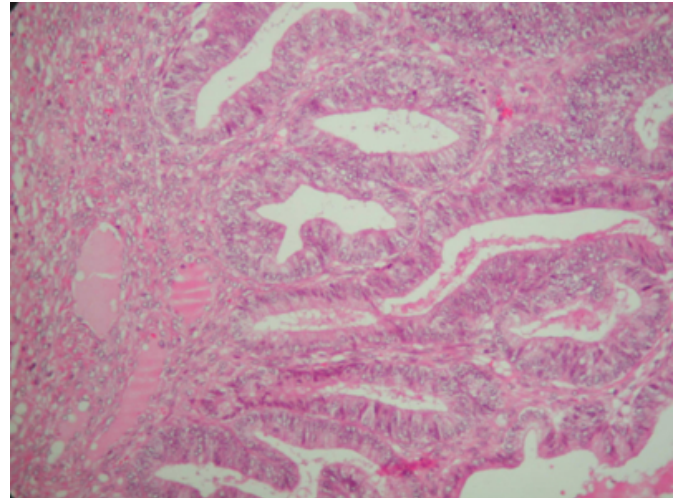
These findings were combined with leucocytosis (WBC: 23100/mm³, NE: 87,9%, LY: 7,7%, MO: 3,6%, BA: 0,8%), hyperkalemia (K: 5,9 meq/L) and elevation of tumour markers CEA (5,7ng/ml, NR: <3), CA 19-9 (500U/ml, NR: 0-37) and CA 125 (386,8 U/ml, NR: 0-35).

A preoperative paracentesis and drainage of pleural effusion was necessary to relieve the patient's dyspneic symptomatology. The cytologic examination of the fluid was negative for malignant cells.

The patient underwent an exploratory laparotomy with excision of both the tumor and the right ovary. The tumor was diagnosed histologically as an ovarian well differentiated endometrioid carcinoma (Fig. 3).

Figure 4

Figure 3 : Papillar and tubular formations of endometrioid carcinoma developed into ovarian substrate (H-E X200)



The immediate postoperative resolution of hydrothorax and ascites confirmed the diagnosis of pseudo-Meigs syndrome (Fig.4).

Figure 5

Figure 4 : Resolution of hydrothorax after tumor removal



The patient followed chemotherapy and remains in good condition 18 months after surgery, without any indication of metastatic disease.

DISCUSSION

The pseudo-Meigs syndrome can be combined with either benign or malignant neoplasms (Tabl.1).

Figure 6

Table 1: Tumors Associated With Pseudo-Meigs Syndrome

BENIGN TUMORS	
■	Ovarian tumors, other than fibromas
□	Stromal tumors
□	Struma ovarii
□	Teratomas
□	Cystadenomas
■	Paraovarian fibromas
■	Uterine leiomyomas
■	Leiomyomas of broad ligament
MALIGNANT TUMORS	
■	Primary ovarian tumors
□	Adenocarcinomas
□	Endometrioid carcinomas
■	Secondary metastatic ovarian tumors, from primary gastrointestinal cancers

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^{2, 3}.

The majority of ovarian tumors, associated with hydrothorax and ascites, have a diameter of more than 6cm. 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.

The pseudo-Meigs syndrome is clinically important because it resembles metastatic pelvic cancer. Especially in patients with malignant ovarian tumors, cytologic examination of the body cavity effusions is essential to differentiate between reactive process and metastatic tumor spread⁴. While detection of malignant cells is a marker of metastatic disease and a sign of bad prognosis, benign effusions of pseudo-Meigs syndrome affect neither disease stage nor the patient's prognosis. Determination of the presence or absence of tumor spread is based primarily on cellular morphology study, but if distinction between reactive mesothelial and cancer cells is difficult, immunocytochemistry may be necessary.

At this point, must be underlined that an ovarian mass combined with pleural and peritoneal effusions not always represents an advanced malignancy, even with elevation of CA 125 value^{5, 6}. There are some benign pelvic lesions causing pseudo-Meigs syndrome, which are associated with elevated levels of this tumor marker, such as struma ovarii,

ovarian cystadenomas, uterine leiomyomas and broad ligament leiomyomas^{3, 7,8,9,10,11,12,13}. CA 125 levels decline to the normal range after tumor resection.

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

Figure 7

Table 2: Rare Clinical Entities Associated With Pseudo-Meigs Syndrome

■	Amylase-producing serous papillary ovarian neoplasm with elevated pleural fluid amylase ¹⁴
■	Broad ligament leiomyoma with bilateral reversible hydronephrosis requiring ureteric stenting ¹³
■	Paraovarian fibroma, an extremely rare neoplasm, probably of paramesonephric origin ¹⁵
■	Leiomyoblastoma of the colon ¹⁶
■	Pedunculated uterine leiomyoma with parasitized blood supply from omentum ¹⁰
■	After a single dose GnRH-Analogue administration for the treatment of uterine leiomyoma ¹⁷
■	Hydropic degenerating uterine leiomyoma ^{3, 12}
■	Uterine leiomyoma in a patient with severe curvature of the spine and dorsolumbar scoliosis ¹⁸
■	Uterine leiomyoma with bladder attachment and double blood supply from uterus and bladder ¹⁹
■	A variant of pseudo-Meigs syndrome in a 8 years old female patient with persistent bronchorrhea and absence of hydrothorax ²⁰

There also reported 6 cases of pseudo-Meigs syndrome caused by secondary ovarian tumors from gastrointestinal cancers²¹. 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.

There is an interesting current study that sets the question whether common uterine leiomyomas and uterine leiomyomas causing pseudo-Meigs syndrome are cytogenetically related or whether functionally differences in tumour phenotype, and supports a model in which accumulation of the independent mutations- a classical structural rearrangement involving HMGA2 and RAD51L1, in combination with a loss of the second RAD51L1 allele- might play a major role in the development of pseudo-Meigs syndrome²².

CONCLUSION

Pseudo-Meigs syndrome should be considered as a rare differential diagnosis for pleural and ascites effusions. Patients with pseudo-Meigs syndrome may present a diagnostic problem as they masquerade as carcinoma with malignant effusions. Thus they should always undergone exploratory laparotomy. Surgical therapy has a very important role for the complete remission of the disease in

cases of benign tumors, and for the remission of pleural and ascites effusions in cases of malignant tumors.

CORRESPONDENCE TO

Papanikolaou Ch. 28 Sokratous street 55134 Thessaloniki, Greece Tel. 0030 2310 433906 0030 2310 892362 e-mail: ascl@ippokratio.gr

References

1. Kazanov L, Ander DS, Enriquez E, Jaggi FM. Pseudo-Meigs syndrome. *Am J Emerg Med* 1998; 16 (4): 404-405
2. Santopaolo D, Rotondo A, Alfe M, Canciello P, Rito Marcone G, Cusati B. [Meigs syndrome with bilateral hydrothorax]. *Minerva Ginecol* 1993; 45 (5): 263-266
3. Amant F, Gabriel C, Timmerman D, Vergote I. Pseudo-Meigs syndrome caused by hydropic degenerating uterine leiomyoma with elevated CA 125. *Gynecol Oncol* 2002; 83 (1): 153-157
4. Wiatrowska B, Krajci P, Berner A. [Pseudo-Meigs syndrome]. *Tideskr Nor Laegeforen* 2000; 120 (3): 364-366
5. Santangelo M, Battaglia M, Vescio G, Sammarco G, Galleli G, Vetere A, Sommela L, Triggiani E. [Meigs syndrome: Its clinical picture and treatment]. *Ann Ital Chir* 2000; 71 (1): 115-119
6. Domingo P, Montiel JA, Monill JM, Prat J. Pseudo-Meigs syndrome with elevated CA 125 levels. *Arch Intern Med* 1998; 158 (12): 1378-1379.
7. Huh JJ, Montz FJ, Bristow RE. Struma ovarii associated with pseudo-Meigs syndrome and elevated serum CA 125. *Gynecol Oncol* 2002; 86 (2): 231-234
8. Bethune M, Quinn M, Rome R. Struma ovarii presenting as acute pseudo-Meigs syndrome with an elevated CA 125 level. *Aust N Z J Obstet Gynecol* 1996; 36 (3): 372-373
9. Long CY, Chen YH, Chen SC, Lee JN, Su TH, Hsu SC. Pseudo-Meigs syndrome and elevated levels of tumor markers associated with benign ovarian tumors: Two cases reports. *Kaohsiung J Med Sci* 2001; 17 (11): 582-585
10. Kebapci M, Aslan O, Kaya T, T Yalsin O, Ozalp S. Redunculated uterine leiomyoma associated with pseudo-Meigs syndrome and elevated CA 125 level: CT features. *Eur Radiol* 2002; 12: 127-129
11. Migishima F, Jobo T, Hata H, Sato R, Ikeda Y, Arai M, Kuramoto H. Uterine leiomyoma causing massive ascites and left pleural effusion with elevated CA 125: A case report. *J Obstet Gynecol Res* 2000; 26 (4): 283-287
12. Dunn JS Jr, Anderson CD, Method MW, Brost BC. Hydropic degenerating leiomyoma presenting as pseudo-Meigs syndrome with elevated CA 125. *Obstet Gynecol* 1998; 92 (4): 648-649
13. Brown RS, Marley JL, Cassoni AM. Pseudo-Meigs syndrome due to broad ligament leiomyoma: A mimic of metastatic ovarian carcinoma. *Clin oncol CR Coll Radiol* 1998; 10(3):198-201
14. Gramer SF, Bruns DE. Amylase-producing ovarian neoplasm with pseudo-Meigs syndrome and elevated pleural fluid amylase: Case report and ultrastructure. *Cancer* 1979; 44 (5): 1715-1721
15. Giannakopoulos K, Giannakopoulou CH, Matalliotakis I, Neonaki M, Papanicolaou N, Koumantakis E. Pseudo-Meigs syndrome caused by paraovarian fibroma. *Eur J Gynecol Oncol* 1998; 19 (4): 389-390
16. Koak YP, Thomas JM. Leiomyoblastoma of the colon presenting as pseudo-Meigs syndrome. *Eur J Surg Oncol* 1999; 25 (4): 446-447
17. Lee MJ, Kazer RR. Massive ascites after leuprolide acetate administration for the treatment of leiomyomata uteri. *Fertil Steril* 1992; 58 (2): 416-418
18. Terada S, Suzuki N, Uchide K, Akasofu K. Uterine leiomyoma associated with ascites and hydrothorax. *Gynecol Obstet Invest* 1992; 33 (1): 54-58
19. Weise M, Westphalen S, Fayyazi A, Emons G, Krauss T. Pseudo-Meigs syndrome: Uterine leiomyoma with bladder attachment associated with ascites and hydrothorax: A rare case of a rare syndrome. *Onkologie* 2002; 25 (5): 443-446
20. Koutras A, Fischer S. Variant of pseudo-Meigs syndrome. *Int J Gynecol Obstet* 1983; 21 (2): 179-182
21. Nagakura S, Shirai Y, Hatakeyama K. Pseudo-Meigs syndrome caused by secondary ovarian tumors from gastrointestinal cancer: A case report and review of the literature. *Dig Surg* 2000; 17 (4): 418-419
22. Amant F, Debiec-Rychter M, Schoenmakers EF, Hagemeyer-Hausman A., Vergote I. Cumulative dosage effect of a RAD51L1/HMGA2 fusion and RAD51L1 loss in a case of pseudo-Meigs syndrome. *Genes Chromosomes Cancer* 2001; 32 (4): 324-329

Author Information

Ch. Papanikolaou

1st Surgical Clinic, Hippokratio General Hospital

K. Fortounis

1st Surgical Clinic, Hippokratio General Hospital

S. Ainalis

1st Surgical Clinic, Hippokratio General Hospital

K. Biba

1st Surgical Clinic, Hippokratio General Hospital

A. Papanikolaou

Department of Pathology, Hippokratio General Hospital

G. Hatzitheoxaris

1st Surgical Clinic, Hippokratio General Hospital