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 bening 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 righ 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 bening 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[1].

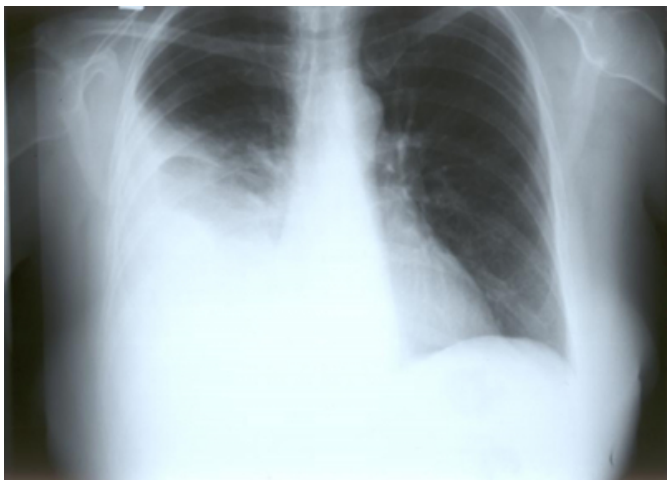
CASE REPORT

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

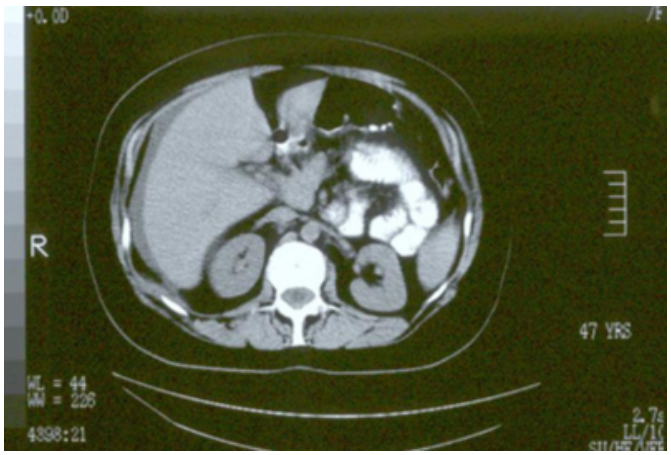
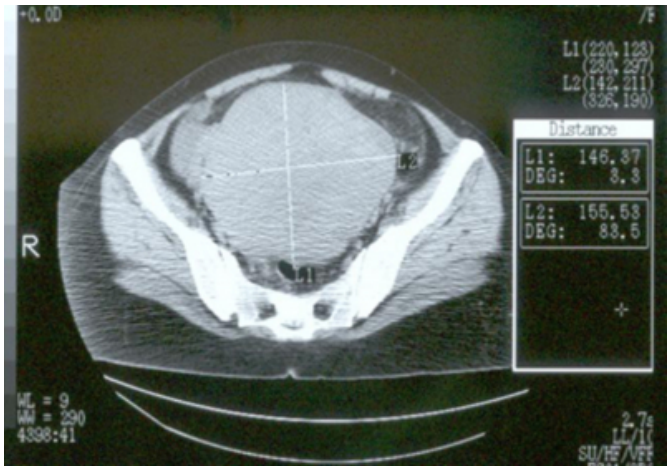


Figure 3

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



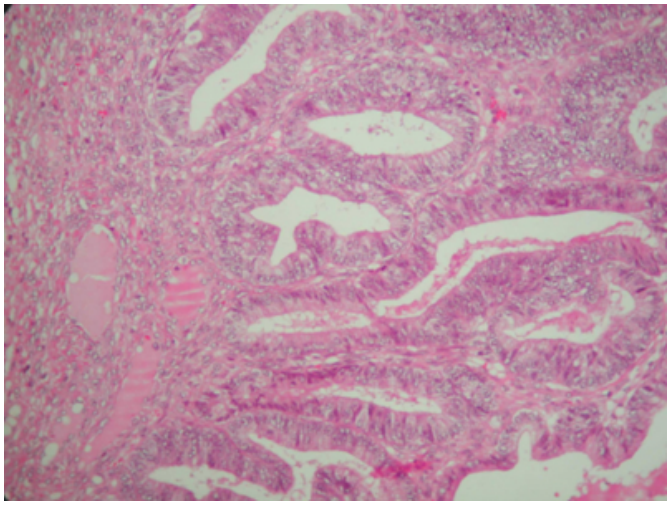
These findings were combined with leycocytosis (WBC: 23100/mm^3, NE: 87,9%, LY: 7,7%, MO: 3,6%, BA: 0,8%), hyperkalemia (K: 5,9 meq/L) and elevation of tumour markers CEA (5,7ng/ml, NR: <3), CA 19-9 (500U/ml, NR: 0-37) and CA 125 (386,8 U/ml, NR: 0-35).

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 righ ovary. The tumor was diagnosed histologically as an ovarian well differentiated endometrioid carcinoma (Fig. 3).

Figure 4

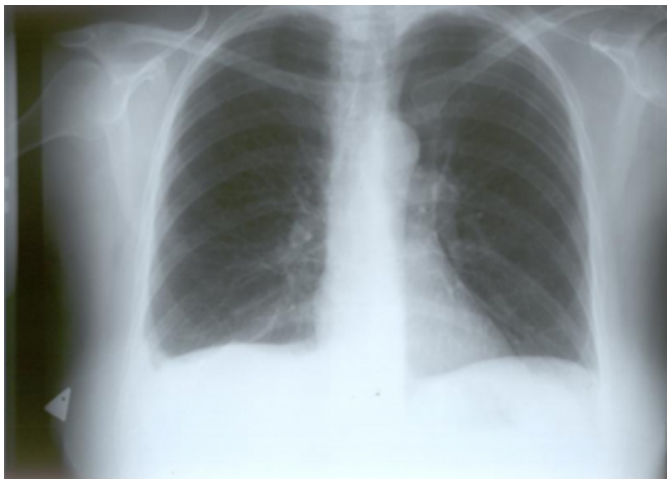
Figure 3 : Papillar and tubular formations of endometrioid carcinoma developed into ovarian substrate (H-E X200)



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



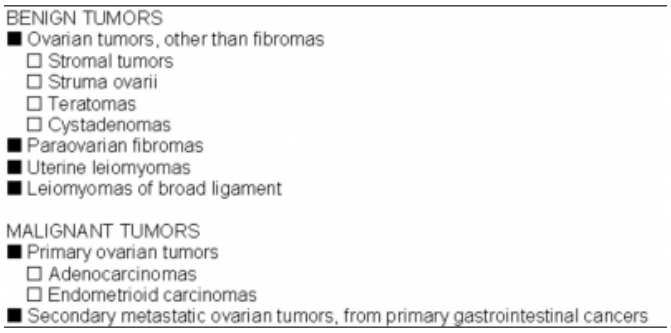
The patient followed chemotherapy and remains in good condition 18 months after surgery, without any indication of metastatic disease.

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.

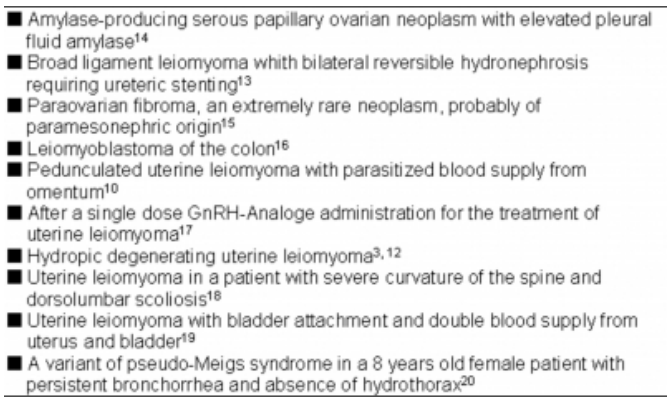
The pseudo-Meigs syndrome is clinically important because it resembles metastatic pelvic cancer. Especially in patients with malignant ovarian tumors, cytologic examination of the body cavity effusions is essential to differentiate between reactive process and metastatic tumor spread[4]. While detection 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 mesothial 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 whith 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-Analoge 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 cancers[21]. 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 syndrome[22].

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 syndome. 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. Kaohsing 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, Hagemeijer-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