Pseudo-Meigs’ Syndrome Caused by a Uterine Leiomyosarcoma: A New Clinical Condition

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Introduction

Meigs’ syndrome is characterized by an association of a benign ovarian tumor with peritoneal and pleural effusions without malignant cells. Meigs [1] separated his definition into true Meigs’ syndrome, which he preferred to call Demons-Meigs’ syndrome, and pseudo-Meigs’ syndrome distinguished by different tumor types.

No data are available regarding the incidence of pseudo-Meigs’ syndrome; it is considered quite rare, even more so than Meigs’ syndrome.

We describe a rare case of pseudo-Meigs’ syndrome caused by a uterine leiomyosarcoma not reported in the literature so far.

Case Report

A 30-year-old woman arrived at our clinic with pelvic pain and dyspnea. She underwent an exploratory laparotomy for a large pelvic mass complicated by ascites and hydrothorax. Cytological examination of the effusions was negative for malignant cells. Histological analysis of the tumor mass revealed a uterine epithelioid leiomyosarcoma. After surgery, we observed resolution of the effusions. Conclusion: Uterine leiomyosarcoma and pseudo-Meigs’ syndrome are two rare entities. To our knowledge, there are no similar reports in the literature, and therefore we present this new clinical condition due to its high scientific evidence.

massive right pleural effusion was found by chest X-ray (fig. 1). Computed tomography (CT) of the abdomen showed mild ascites and a large heterogeneous pelvic tumor (19 x 17 cm) with solid and cystic components. The lymph nodes of the mesenteric and paraesophageal compartment were enlarged (fig. 2). Blood tests revealed anemia (HGB: 7.4 g/dl, HCT: 25%, GR: 3.09 x 10^6/H9262 l), alkalosis (pH 7.45, PO2 67 mm Hg, PCO2 36 mm Hg, HCOO 3 – 25 mmol/l) and elevation of tumor markers (CA-125: 1,184 U/ml, CA 19.9: 151 U/ml). Drainage of pleural effusion was necessary to relieve the patient’s dyspneic symptomatology. Cytologic and bacteriologic examination of the fluid was negative for malignant cells. The patient underwent an exploratory laparotomy. As the tumor mass appeared partially necrotic and involved the uterus and both ovaries, we performed biopsies. The intraoperative frozen section examination gave evidence for ‘indefinite stromal tumor’. Based on the inconclusive result of the intraoperative examination and the infiltrative appearance of the tumor mass, we decided to perform a total hysterectomy with bilateral salpingooophorectomy. Cytological examination of the peritoneal fluid was negative for malignant cells, and final histological analysis of the tumor mass revealed a ‘uterine epithelioid leiomyosarcoma with marked deep adenomyosis’.

A few days after surgery, the chest radiograph showed resolution of the pleural effusion (fig. 3), and a CT of the abdomen demonstrated a minimum effusion perihepatitis and the disappearance of lymphadenopathy (fig. 4). After about a month, the tumor marker levels returned to normal. After the resolution of ascites, the patient received a cycle of chemotherapy with bleomycin. Since then, she has received clinical examinations every 3 months. Follow-up at 15 months showed no evidence of tumor recurrence.

**Conclusion**

Historically, Meigs’ syndrome is defined by four main features: benign ovarian tumor (fibroma or thecoma, granulosa cell tumor) with fluid in the abdomen and chest without malignant cells; removal of the tumor should cure the patient [1]. The term pseudo-Meigs’ syndrome was first coined in 1954 by Meigs. This represented an attempt to separate cases caused by ovarian fibrous tumors from those ones caused by all other benign or malignant gynecological tumors.

The pleural and abdominal effusions were massive in our case and the fluid cytology was negative for malignant cells although the patient was suffering from a malignant tumor of the uterus.

The origin of the ascitic fluid remains unclear. Some authors [2, 3] postulate different mechanisms, such as active secretion by the tumor or the peritoneum, lymphatic obstruction, venous obstruction, inflammatory reactions, toxins and low serum proteins.

In our case, it is possible that tumor necrosis caused the transudation of interstitial edema fluid determining ascites. Another plausible explanation is that pressure of the tumor itself caused the escape of fluid through the surface lymphatics. A discrepancy between arterial supply to a large tumor tissue and its venous and lymphatic drainage could lead to stromal edema and transudation. In fact, CT revealed a diffuse lymphadenopathy in our case that resolved spontaneously after removal of the pelvic mass. Unfortunately, it was not possible to perform a lymph node sampling due to technical difficulties.

The mechanism for the appearance of the hydrothorax is different. Most authors agree that the pleural fluid is transferred from the abdomen. The fluid travels to the pleural spaces from the communications across the diaphragm and the partially patent foramen of Bochdalek. Other potential communications are the diaphragmatic, aortic and caval apertures; these potential communications may become patent following increases in intra-abdominal pressure and the negative intrathoracic pressure [4].

Another aspect to stress is the increased value of CA-125 (1,184 U/ml) in our case. The literature has reported several cases of pseudo-Meigs’ syndrome with elevated CA-125 [5]. Although the precise mechanism remains unclear, a biochemical factor, mechanical irritation from a large tumor or an increase in intraperitoneal pressure from a large volume of ascites might be the primary factor in this process.
Since 1954, case histories have been published regularly. They have dealt essentially with cases of tumors other than fibromas, hence the name pseudo-Meigs’ syndrome. Similarly, malignant tumors of the ovary have sometimes been included, the removal of which brings about the disappearance of the effusion with no malignant cells [6].

Several authors [7, 8] have published cases of uterine leiomyomas associated with hydrothorax and ascites. Other authors [9, 10] have reported cases of pseudo-Meigs syndrome secondary to ovarian metastases from gastrointestinal and mammary cancer.

In recent years, several pathological entities were considered as pseudo-Meigs’ syndrome. Therefore, for clarity it has become necessary (table 1) to include in Meigs’ syndrome all benign genital tumors, and bring together all other entities (malignant tumors with no neoplastic cells in effusions, pseudotumors and other rarities) under the name of pseudo-Meigs’ syndrome [11].

In light of this new classification, our case report appears to be a rarity. The pseudo-Meigs’ syndrome is an uncommon condition, such as uterine leiomyosarcoma, that accounts for approximately 1% of female genital tract malignancies [12]. Furthermore, our patient was only 30 years old, while both uterine leiomyosarcoma and Meigs’ syndrome usually occur in women over 40 years of age [11]. The concomitant presence of a uterine leiomyosarcoma with ascites and right hydrothorax without malignant cells is not reported in the literature. Huang et al. [13] reported the only case that can be compared to ours. He described pseudo-Meigs’ syndrome caused by a uterine smooth muscle tumor of uncertain malignant potential.

**Fig. 2.** Coronal and axial CT images: large pelvic tumor (19 × 17 cm), mild ascites and diffuse lymphadenopathy.

**Fig. 3.** Chest X-ray: resolution of pleural effusion after surgery.
In our case, the absence of malignant cells in peritoneal and pleural fluid and the resolution of the clinical condition after the removal of the tumor allow us to classify this case as pseudo-Meigs’ syndrome, which thus far is unique.

**Disclosure Statement**

The authors have no conflicts of interest to declare.

**Table 1. New classification of pseudo-Meigs’ syndrome**

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<th>Common criteria</th>
<th>Meigs’ syndrome</th>
<th>Pseudo-Meigs’ syndrome</th>
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<td>Hydrothorax</td>
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<td>Cytology negative for malignant cells</td>
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<td>Resolution of the effusions after the removal of the tumor</td>
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<td>Pelvic mass</td>
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<th>Criteria of differentiation</th>
<th>Meigs’ syndrome</th>
<th>Pseudo-Meigs’ syndrome</th>
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<td>Examples</td>
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<td>All benign genital tumors</td>
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<td>– Ovarian fibroma</td>
<td>– Ovarian cancer</td>
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<td>– Uterine leiomyoma</td>
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<td>– Granulosa cell tumors</td>
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**References**


