Case #131

NAME Educational Activities Committee

Case provided by:

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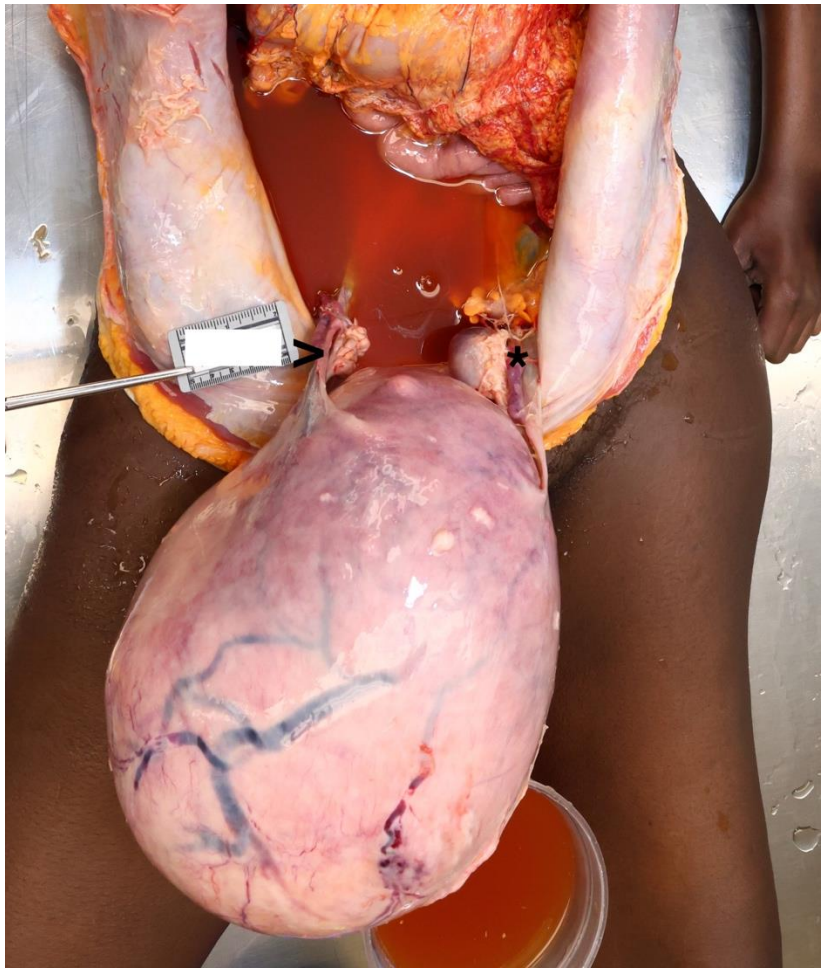


A 40-year-old woman with no known medical history was brought to the emergency room with severe shortness of breath and died shortly after arrival. External examination showed no evidence of trauma. The lower abdomen was distended. Internal examination revealed a large pelvic mass shown in the figures, associated with fibrous adhesions to the abdominal cavity, approximately 1000 mL of ascitic fluid in the abdomen, and 1500 mL of pleural effusion in the bilateral chest cavities. There were no additional relevant findings. Postmortem toxicology, microbiology, and histology of the internal organs were negative or noncontributory.

The autopsy findings described above are most commonly associated with:

A. Impairment of cardiopulmonary function
B. Cystic lesions of the peritoneal lining
C. Histologic evidence of malignancy
D. Chronic complications of autoimmune disorders

Answer...

A. Impairment of cardiopulmonary function (1) (CORRECT ANSWER, 43.24% of responses)

In this case, the sudden death was resulted from acute cardiorespiratory failure due to massive pleural effusions compressing both lungs and impairing cardiac function. This is a rare complication of Pseudo-Meigs syndrome, a variant of the classic Meigs syndrome.

- *Meigs syndrome* is a rare condition defined by the triad of a benign ovarian tumor (most commonly an *ovarian fibroma*), ascites, and pleural effusion.
- *Pseudo-Meigs syndrome* similarly involves ascites and pleural effusion but is typically associated with benign pelvic tumors other than ovarian fibromas, including tumors of the fallopian tube, uterus, mature teratomas, struma ovarii, ovarian leiomyomas, and occasionally metastatic gastrointestinal tumors. Resolution of effusions following tumor resection is the key diagnostic feature in both conditions.

The pathophysiology of Pseudo-Meigs syndrome is not fully understood but involves multiple mechanisms of fluid accumulation without malignancy. Large pelvic tumors may irritate the peritoneum, producing exudative ascites, while compression of lymphatic or venous channels impairs drainage, resulting in transudative ascites. Tumor-secreted factors may also increase vascular permeability, contributing to both ascites and pleural effusions. Pleural fluid typically reaches the thoracic cavity via transdiaphragmatic lymphatics or defects.

Massive pleural effusions can impair cardiac function by compressing the heart and raising intrathoracic pressure. This reduces venous return and diastolic filling, lowering stroke volume and cardiac output, and may cause hypotension with tamponade-like physiology despite the absence of pericardial fluid. Limited lung expansion leads to hypoxia and pulmonary vasoconstriction, which further increases right ventricular afterload. Together, these changes compromise cardiorespiratory function.

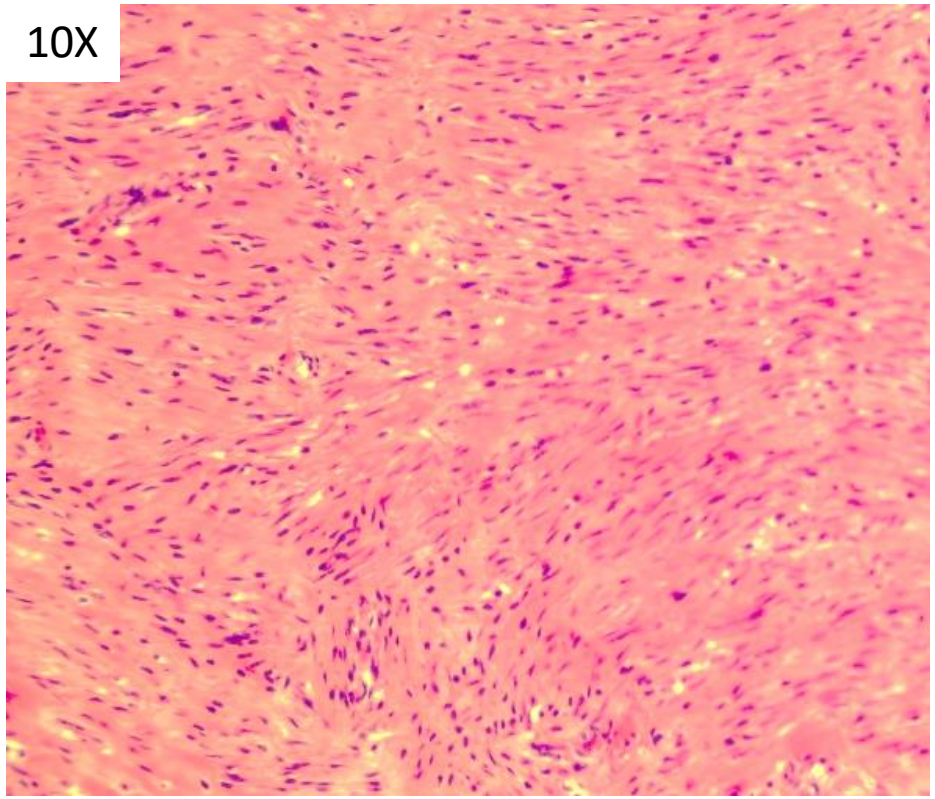
A. Impairment of cardiopulmonary function (2)

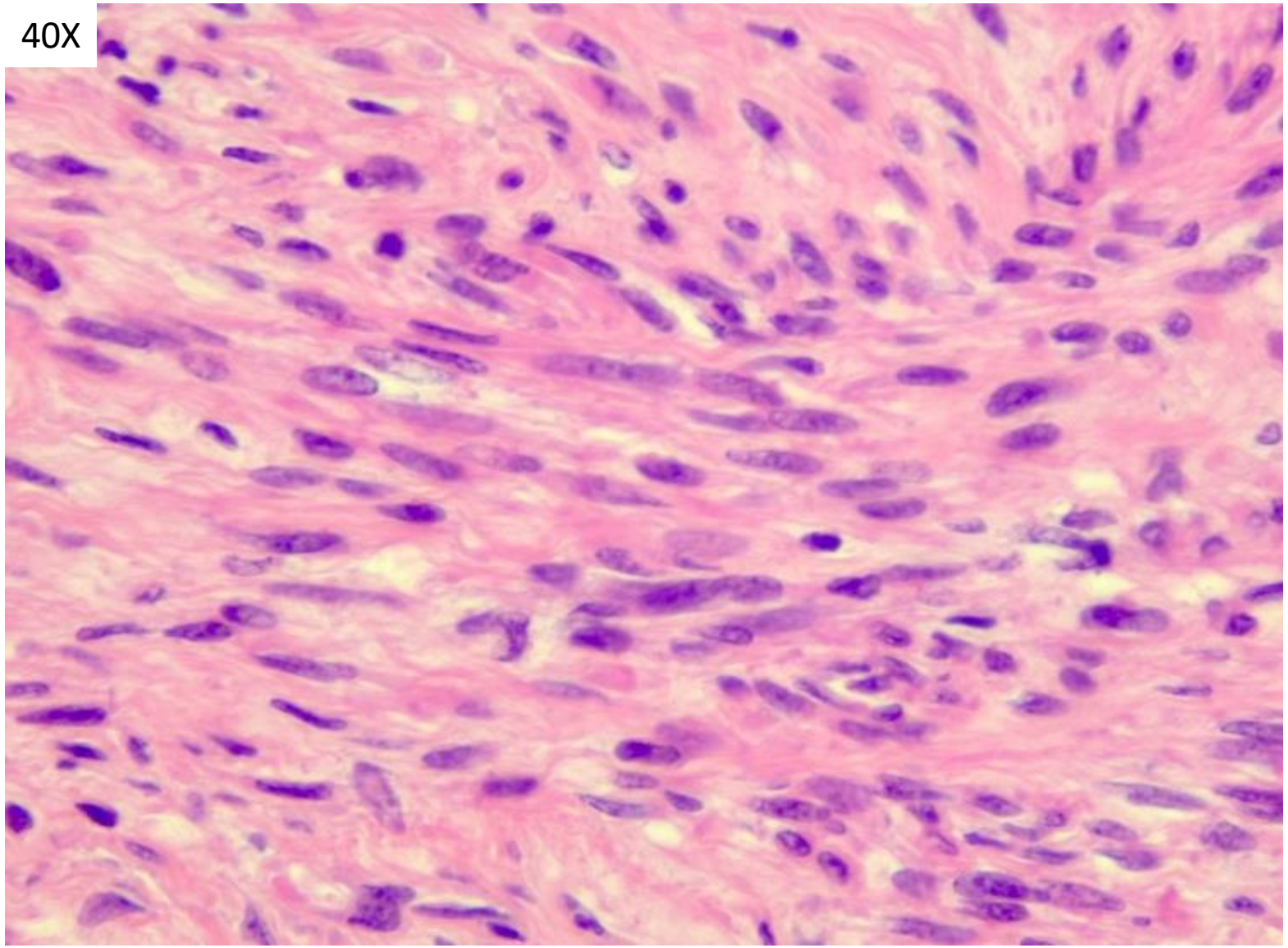
In this case, the large mass was a benign uterine leiomyoma, a tumor that has previously been associated with Pseudo-Meigs syndrome in clinical reports. “Giant” uterine leiomyomas are generally defined as masses measuring >10 cm in diameter or weighing >11.4 kg, and they are relatively rare in clinical practice. Due to their size, ischemia, necrosis, and cystic degeneration may occur, which complicates the differentiation from uterine sarcomas and ovarian tumors. A well-defined central necrosis is common in benign giant leiomyomas because the peripheral areas of the mass cannot receive adequate blood supply, resulting in cell death.

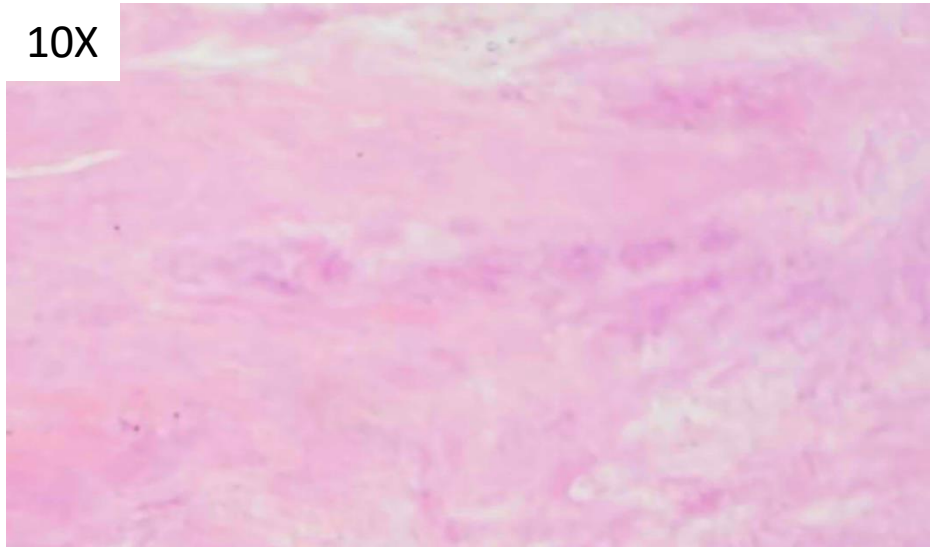
Gross examination of the mass in the reported case revealed a large mass with a smooth to bosselated external surface, and focal loose adhesions to surrounding tissue. The cut surface was solid, whorled, and tan-white, with a well-demarcated central yellow-white, friable, and softened necrotic area.

Microscopic examination of the mass showedintersecting bundles of uniform, spindle-shaped smooth muscle cells with elongated, blunt-ended (“cigar-shaped”) nuclei and eosinophilic cytoplasm, with minimal cytologic atypia and no evidence of infiltrative growth. Examination of the necrotic areas showed coagulative necrosis of smooth muscle cells, loss of nuclear detail, hyalinization, and ghost-like outlines of muscle fibers.

The ovaries were not involved, as showed in the autopsy photo. Both lungs were completely collapsed, explaining the shortness of breath the subject experienced before becoming unresponsive.







Microscopic appearance of the uterine mass and necrotic areas.

Other responses…

B. Cystic lesions of the peritoneal lining (Incorrect – 11.07% of responses)

While the autopsy findings of ascites, fibrous adhesions, and abdominal distension may rise concerns for primary peritoneal tumors, cystic lesions of the peritoneal lining are typically seen in benign multicystic peritoneal mesothelioma, a rare tumor of the peritoneum that typically presents with multiple cystic lesions, fibrous adhesions, and varying degrees of ascites, features that overlap with those seen in this patient. While both conditions can produce extensive adhesions and fluid collections, the large pelvic mass is a hallmark for Pseudo-Meigs syndrome.

C. Histologic evidence of malignancy (Incorrect – 40.37% of responses)

Pseudo-Meigs syndrome most often arises in association with benign pelvic tumors other than ovarian fibromas. Reported examples include tumors of the fallopian tube, uterine neoplasms such as leiomyomas, and ovarian lesions including mature teratomas and struma ovarii. The unifying feature is the development of ascites and pleural effusion that resolve following surgical removal of the mass.

Although usually linked to benign pathology, rare cases have described malignant tumors (such as ovarian carcinomas or gastrointestinal metastases) presenting with the same clinical triad.

Intraperitoneal adhesions are uncommon in benign uterine leiomyomas and may raise suspicion for malignancy. Whereas malignant tumors often produce adhesions through infiltration or peritoneal irritation, giant leiomyomas can also generate adhesions due to their size, local compression and inflammation of the peritoneal lining, focal ischemic necrosis, or cystic and pedunculated degeneration.

Importantly, the diagnosis of Pseudo-Meigs syndrome rests on the clinical course or autopsy findings rather than the tumor’s microscopic features. Histologic confirmation of benignity or malignancy is therefore not required.

D. Chronic complications of autoimmune disorders (Incorrect – 5.33% of responses)

While Pseudo-Meigs syndrome typically involves pelvic tumors causing ascites and pleural effusion, there is a rare but distinct condition known as **Pseudo-pseudo Meigs syndrome (PPMS)** that can mimic this presentation without any detectable pelvic mass. PPMS, also referred to as **Tjalma syndrome**, is most commonly associated with autoimmune disorders, particularly **Systemic Lupus Erythematosus (SLE)** and other connective tissue diseases. In these cases, patients generally present with ascites, pleural effusions, and elevated serum CA-125 levels similar to those seen in Meigs and Pseudo-Meigs syndromes. However, the underlying cause is related to systemic inflammation and immune dysregulation rather than tumor-related fluid accumulation.

REFERENCES

1. Barranco R, Molinelli A, Gentile R, Ventura F. Sudden, unexpected death due to Pseudo-Meigs syndrome: a case report and review of the literature. Am J Forensic Med Pathol. 2019;40(1):89-93. doi:10.1097/PAF.0000000000000438

2. Hlaise KK, Shingange SM. Sudden death associated with Meigs syndrome: an autopsy case report. Am J Forensic Med Pathol. 2012;33(1):58-60. doi:10.1097/PAF.0b013e3181edf318

3. Miyoshi A, Miyatake T, Hara T, Tanaka A, Komura N, Komiya S, et al. Etiology of ascites and pleural effusion associated with ovarian tumors: literature review and case reports of three ovarian tumors presenting with massive ascites, but without peritoneal dissemination. Case Rep Obstet Gynecol. 2015;2015:414019. doi:10.1155/2015/414019

4. Yaguchi A, Ban K, Koshida Y, Fujikami Y, Ogura E, Terada A, et al. Pseudo-Meigs syndrome caused by a giant uterine leiomyoma with cystic degeneration: a case report. J Nippon Med Sch. 2020;87(2):80-6. doi:10.1272/jnms.JNMS.2020_87-205

5. Dang V, Rofail A, Bowman JM, Peloquin J. A case of pseudo-pseudo Meigs' syndrome despite optimized immunosuppressive therapy for systemic lupus erythematosus. Cureus. 2023;15(10):e46753. doi:10.7759/cureus.46753

6. Ye Z, Jiang Y, Yan K, Yu C. Uterine torsion with degeneration and infarction of giant leiomyoma in a postmenopausal woman: a case report. Medicine (Baltimore). 2023;102(45):e35964. doi:10.1097/MD.0000000000035964