

Family Physician Corner

Meigs' Syndrome: A Fibrothecoma Presenting with Torsion

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Meigs' syndrome is rare. It is described as a triad of solid benign ovarian tumor, ascites, and pleural effusion that resolve after tumor resection.

We present a case of a 36-year-old Bahraini female who presented with signs and symptoms of acute pelvic pain and adnexal torsion. She had left ovarian fibrothecoma with elevated CA125, moderate ascites and right-sided pleural effusion.

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Meigs' syndrome is a triad of pleural effusion, ascites, and an ovarian tumor; it is usually a benign condition. However, the signs of Meigs' syndrome indicate a metastatic tumor. According to the British Thoracic Society (BTS), the unilateral pleural effusion with Meigs' syndrome is classified as transudate¹.

Meigs' syndrome was first described by Meigs and Cassa in 1937, as a triad of benign ovarian tumor (fibroma, thecoma and rarely granulosa cells tumor) associated with ascites and pleural effusion which resolves automatically when the tumor is removed. Other tumors of the ovary, such as mucinous cystadenoma, struma ovarii, teratomas, secondary metastatic tumors of ovary, and leiomyoma of the uterus if accompanying with hydrothorax are labeled as "Pseudo-Meigs' Syndrome"².

It is diagnosed only after ruling out ovarian cancer³. Ovarian fibroma or fibrothecoma is a type of sex cord-stromal tumors, consisting almost of 5% to 8% of all types of ovarian tumors, and comprises three histopathologic types of fiber and theca contents including fibroma, thecoma and fibrothecoma⁴.

The majority of cases of ovarian fibroma and fibrothecoma are seen in adolescents and young women with solid pelvic or adnexal mass and benign bio-behavior. Almost 10% to 15% of ovarian fibromas and fibrothecomas are associated with ascites; however, less than 1% are associated with ascites and pleural effusion, identified as "Meigs' syndrome"^{5,6}. There is no particular tumor marker for both these tumors, except sometimes there will be an elevation of CA-125⁷⁻¹⁴.

We report the first documented case of a rare Meigs' syndrome in our facility associating left-sided fibrothecoma, ascites, right-sided pleural effusion and elevation of the CA-125.

THE CASE

A 35-year-old female, para 1, presented with moderate to severe lower abdominal pain for one day. Her menstrual history has been normal for several months other than dysmenorrhea. Pain has not been associated with intestinal or urinary symptoms. Abdominal examination revealed a palpable solid mass arising from the pelvis to umbilicus.

Ultrasound abdomen and pelvis revealed a 14x10 cm abdominopelvic mass with heterogeneous echotexture with no significant internal vascularity seen; the left ovary was not

separately identified, the right ovary was normal and moderate fluid found in the peritoneal cavity.

MRI report revealed 15.5x14x9.5 cm pelvic mass (most likely originating from the left ovary), heterogeneous signal intensity on T1 and T2 with ascites, with contrast enhancement, see figure 1. X-ray chest showed right-sided pleural effusion, see figure 2.

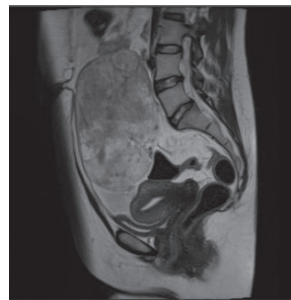


Figure 1 (A)

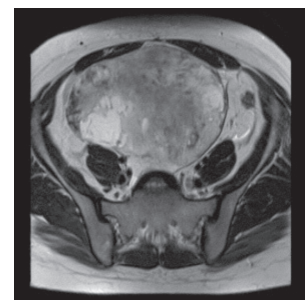


Figure 1 (B)

Figure 1 (A-B): Sagittal and Transverse View of MRI Images Showing Large Pelvic Mass

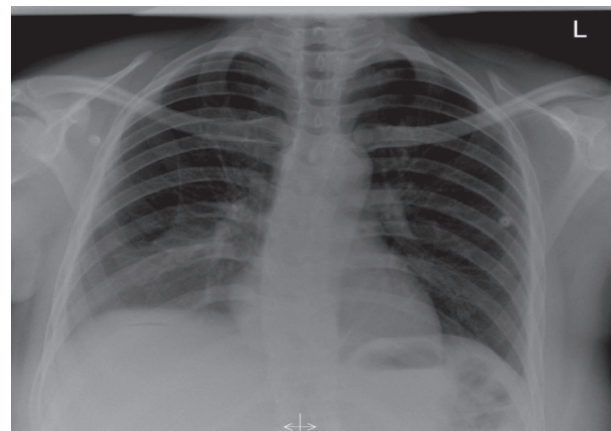


Figure 2: X-ray Chest Showing Right-Sided Pleural Effusion

Her laboratory results were unremarkable except that serum CA125 was high, up to 280 u/ml. Hemoglobin (Hb) 13gm./dl,

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total leukocytes count 9.16, platelets 435 and CRP 17.7 mg/l.

The patient's operative findings were moderate ascites and left-sided solid ovarian mass measuring approximately 15 cm with torsion of adnexa up to 3 times. Uterus, fallopian tubes and right ovary were normal. Omentum, peritoneal surfaces, and undersurface of diaphragm were unremarkable and no visceromegaly was seen. Peritoneal fluid was sent for cytology, the mass was resected, omental, and right ovarian biopsies were taken for metastatic identification.

Histopathology revealed fibrothecoma of left ovary while right ovarian and omental biopsies were normal and peritoneal fluid was inflammatory, see figure 3.

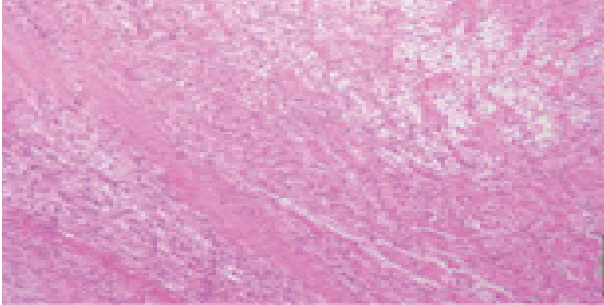


Figure 3: Spindly and Clear Cells with Collagen, H&E, 10X

Postoperative was uneventful and the patient was discharged from the hospital on the 3rd postoperative day. Follow-up showed good recovery and CA-125 level was normal.

DISCUSSION

Meigs' syndrome is rare. The hallmarks of Meigs' syndrome include fibroma or a fibroma-like tumor, ascites, hydrothorax; ascites and hydrothorax completely settle after removal of tumor¹⁵.

The cause of ascitic fluid in Meigs' syndrome is ill-defined. The fluid could be due to transudate from the surface of the tumor, direct pressure of the tumor on adjacent lymphatics or blood vessels, hormonal provocation and torsion of the tumor. The etiology of hydrothorax is also not yet clear. A recent theory suggests that the ascitic fluid is transudate through transdiaphragmatic channels¹⁶.

Raised level of CA-125 and ovarian epithelial carcinoma has been documented. Elevation of up to 26% of CA-125 had been seen in some other non-gynecological cancers and few benign gynecological conditions. The source of this antigen remains unclear; according to some studies, the source could be non-tumor cells and some biochemical factors, such as mechanical irritation of peritoneum or raised intraperitoneal pressure due to a large volume of tumor and ascites¹⁷.

The removal of the tumor in our case was necessary due to the size of the ovarian mass and ensuing pain. It is recognized that giant ovarian mass in Meigs' syndrome is associated with intra-abdominal hypertension and the onset of compartment syndrome¹⁸.

CONCLUSION

Patient with pelvic mass needs to be evaluated systematically and the CA-125 level needs to be tested. The exact diagnosis must be confirmed with histopathology of the mass.

Our patient had left ovarian fibrothecoma with elevated CA125, moderate ascites, and right-sided pleural effusion, which resolved after surgery.

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