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Title: Pseudo-Meigs’ syndrome: A case report and review of the literature

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Short Running Title: Case report on Pseudo-Meigs’ syndrome

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SUMMARY

Pseudo-Meigs’ syndrome is a rare syndrome, which should be included in differential diagnosis for pleural and ascitic effusions. Patients with Pseudo-Meigs’ syndrome may present a diagnostic problem and should always undergo exploratory laparotomy. Surgical therapy can result in complete remission of the disease in cases of benign tumors.
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ABSTRACT

Introduction
Meigs' syndrome and Pseudo Meigs' syndrome both presents with hydrothorax and ascites. Meigs' syndrome is characteristically associated with ovarian fibroma whereas Pseudo-Meigs' syndrome is associated with any ovarian or pelvic tumors, other than ovarian fibroma.

Case report
The case presented here concerns a 48 year old perimenopausal woman with a long history of 8-10yrs of abdominal distension. Her examination revealed a right pleural effusion, massive ascites and large heterogeneous pelvic tumor, measuring 42x31cm. After a preoperative ascitic tapping, the patient underwent an exploratory laparotomy with excision of the tumor, uterus and the right ovary. The tumor was diagnosed histologically as an ovarian mucinous cystadenoma. The postoperative resolution of hydrothorax and ascites confirmed the diagnosis of Pseudo-Meigs' syndrome. The patient remains in good condition 12 months after surgery.

Conclusion
Pseudo-Meigs' syndrome being a rare syndrome, with a good prognosis should be included in differential diagnosis in women presenting with unexplained hydrothorax and ascites.

Key words: Pseudo- Meigs’ syndrome, Ascites, Hydrothorax, Ovarian tumor
INTRODUCTION

Meigs’ syndrome is a rare condition, defined as the co-existence of benign ovarian fibroma, pleural effusion and ascites. While, Pseudo-Meigs’ syndrome is characterized by the co-existence of pleural effusion, ascites and other ovarian or pelvic tumors. It was Meigs and Cass who brought out the significance of pleural effusion and ascites in ovarian fibroma. These syndromes should be considered in otherwise healthy postmenopausal women, who present with either hydrothorax or ascites. For both these syndromes, surgical resection of the tumor is the only therapeutic choice, resulting in resolution of fluid accumulations [1].

CASE REPORT

A 48 year-old perimenopausal woman came with history of abdominal distension since last 8-10 years, difficulty in breathing with increasing intensity over the past few months. She became very uncomfortable in supine position. She had no medical or surgical history of note. She is para 3 with uneventful vaginal deliveries. She took no regular medication and had no family medical history of note (Figure 1). Auscultation revealed absence of breath sounds at the right lower hemithorax and normal heart sounds. On abdominal examination massive ascites was noted. The mass was not palpable because of the tense ascites. Chest x-ray revealed mild right sided pleural effusion (Figure 2). ECG was within normal limits. On ultrasound abdomen, a massive multi septate cystic mass with suspected ovarian origin, with massive ascites was noted. CT-scan revealed a huge multiseptate mass with solid and cystic components measuring 42*31 cm arising from the pelvis. Left side ovary was not visualised and uterus was normal sized. Massive ascites was noted. No obvious lymphadenopathy was seen. Her serum CA 125 was 49 U/ml (normal <35U/ml). AFP was within normal limits while beta hcG was not detectable. Her serum proteins were slightly below normal. Routine blood investigations, including LFT’S and RFT’S were within normal limits. Ascitic tap fluid cytology revealed low cellular fluid comprising of lymphocytes and mesothelial cells. No evidence of malignancy.
A preoperative diagnosis of left ovarian tumor was made and nearly 6 L of ascitic fluid was drained in two settings in ward three days before surgery and the day before surgery. In theatre, under epidural anaesthesia, a wide bore silicone catheter was inserted and nearly 18 L of ascitic fluid was tapped slowly over a period of 60 minutes in lateral position. Then in supine position, through a midline incision from pubic symphysis to 2cm above umbilicus, a mass measuring 42x31cm, weighing 9 kg was removed, originating from left ovary.

There was no any evidence of metastasis or lymphadenopathy. Omental and peritoneal biopsy were taken. Hysterectomy with bilateral salpingo oophorectomy was done. Grossly uterus with cervix measured 9x6 cms, right ovary measured 2.5x1.3 cms, both unremarkable (Figure 3).

On histopathology report, the mass was diagnosed as an ovarian mucinous cystadenoma. The pleural effusion resolved by post-operative day 10. The patient remains in good condition 12 months after surgery.

DISCUSSION

Meigs’ syndrome is defined as a triad of benign ovarian tumor (ovarian fibroma), ascites and pleural effusion. Though the association of pleural effusion with benign pelvic tumor was described by Salmon in 1934, it was Meigs and Cass whom brought out the significance of pleural effusion and ascites in ovarian fibroma. It is to be noted that the ascites and effusion resolves completely after resection of tumor. Meigs’ syndrome is a benign disease with a good prognosis.

Psedo-Meigs’ syndrome on the other hand consists of ascites and pleural effusion associated with any pelvic tumor other than fibroma. It is clinically important as it may resemble metastatic pelvic cancer. Cytological examination of the body cavity effusions is essential to differentiate between reactive process and metastatic tumor spread.

The etiology of the fluid accumulation in Meigs’ and Pseudo Meigs’ syndrome remains unclear, although it appears to be related to lymphatic obstructions. Most likely it is due to filtration of interstitial fluid in the peritonium through tumor capsule and diffusion to the pleural space through diaphragmatic lymphatic vessels and apertures. The effusion can be moderate or massive and is grossly transudative, but
occasionally contains blood cells[2,3]. It is usually observed with ovarian tumor larger than 6 cm and they completely regress after neoplasm removal.

Malignant tumor with ascites should be differentiated by detection of malignant cells on cellular morphology study and immunochemistry whenever necessary. It is important to understand that an ovarian mass combined with pleural and peritoneal effusion not always represents an advanced stage of malignancy[4]. CA125 values are also not reliable in distinguishing, as some benign pelvic tumors causing Pseudo-Meigs’ syndrome are associated with elevated levels of the tumor marker, like ovarian cystadenomas, struma ovarii, uterine and broad ligament leiomyomas[5-10].

The case presented here is unique with respect to the large amount of ascitic fluid drained, nearly 25 litres. It is to be noted that Meigs’ or Pseudo-Meigs’ syndrome can present with such large ascites. Sudden release of this fluid during surgical entry into the abdomen can lead to sudden drop in blood pressure of the patient and cardio-respiratory collapse. Keeping this thing in mind, we drained some fluid preoperatively. During the surgery, before entering abdomen, we drained the remaining ascitic fluid slowly with a large bore catheter over 60 mins.

In the literature, very few reports have been published on Pseudo-Meigs’ syndrome with such a big mass developing over a period of 8-10 years. It differs in some important respects. Firstly, the age of patient is below fifty, with a history of abdominal distension since 7-8 years back, much younger than when the majority of these tumors present. Secondly, we have not seen a case report with such a big ovarian tumor removed i.e. 42x31cm size.

**CONCLUSION**

Pseudo-Meigs’ syndrome is a rare syndrome, but with a good prognosis. It should be included in differential diagnosis in women presenting with unexplained hydrothorax and ascites. Though a benign disease, prompt diagnosis should be made by ultrasound and subsequent tissue diagnosis. Suspicion of this syndrome will prevent undue delay in diagnosis and management of patients.
CONFLICT OF INTEREST
The authors declare no conflict of interests

AUTHOR’S CONTRIBUTIONS
Divya Dayanandan Kallarackal
Group 1- Conception and design, Acquisition of data, Analysis and interpretation of data
Group 2- Drafting the article, Critical revision of the article
Group 3- Final approval of the version to be published

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Group 2- Drafting the article, Critical revision of the article
Group 3- Final approval of the version to be published

REFERENCES


FIGURE LEGENDS

Figure 1: Pre-operative picture of patient (written informed consent taken for this picture).

Figure 2: Pre-operative chest x ray of patient.

Figure 3: Mass after dissection
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Figure 2: Pre-operative chest x ray of patient.

Figure 3: Mass after dissection