

Atypical Meigs syndrome: A rare case with Review of literature

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Abstract

Meigs Syndrome is a rare presentation which includes a benign ovarian tumor that is fibroma along with ascites and pleural effusion. One percent of ovarian tumors present as Meigs syndrome. Some patients present with either ascites or pleural effusion with benign tumor of ovary which is classified as ‘Atypical or Incomplete Meigs syndrome’. The syndrome is common in postmenopausal women around fifty years of age and its peak incidence is seen in the seventh decade. Various theories have been proposed for pathophysiology of ascites and pleural effusion in Meigs syndrome. Similar clinical presentation can also be seen in metastatic malignancies hence it is important to diagnose benign nature of ovarian tumor in case of Meigs syndrome for proper treatment and management of patient. We present a case of Atypical Meigs syndrome in a seventy-year-old female patient to highlight its clinical presentation and histopathological features.

Keywords: Atypical Meigs Syndrome, Benign Ovarian Tumor, Fibroma, Histopathological Examination.

Introduction

Meigs syndrome is a rare presentation where a benign ovarian tumor presents along with ascites and pleural effusion, it comprises one percent of ovarian tumors. Atypical Meigs Syndrome includes a combination of benign ovarian tumor along with either ascites or pleural effusion^[1,2]. JoeVinscent Meigs and John W. Cas had reported seven cases of Fibroma of ovary associated with ascites and pleural effusion which was later named as Meigs syndrome. They reported the resolution of ascites and pleural effusion after removing the tumor and thus Meigs syndrome became a distinct entity.^[1,3]

Case Report

A seventy-year-old female patient, presented with abdominal distension and breathlessness for two months. There was no history of weight loss. Ultrasonography abdomen revealed massive ascites. Chest X Ray was within normal limits. CA125 levels were 6.7 U/ml. Intra operative examination revealed a huge right ovarian mass along with massive ascites. The tumor was removed and sent for histopathological examination.

On gross examination we received a huge ovarian mass measuring 14 x 6.5 x 4.5 cm. External surface was grey white and bosselated, the capsule was intact. Cut section showed homogenous grey white areas with whorled appearance. The tumor was hard in consistency.



Figure 1: A huge grey white bosselated tumor. Cut section showing whorled appearance.

Microscopic examination revealed spindle shaped neoplastic cells arranged in fascicles having benign looking elongated nuclei with tapering ends. Stroma showed fibrocollagenous tissue.

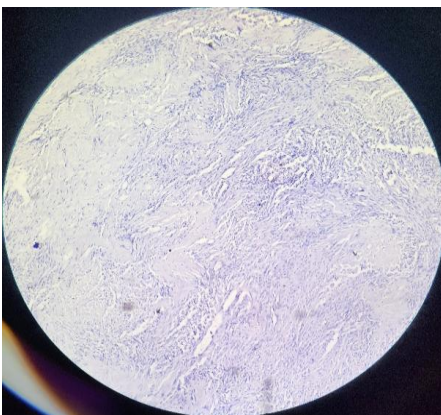


Figure 2

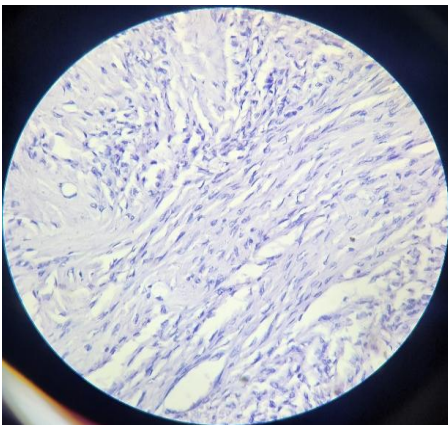


Figure 3

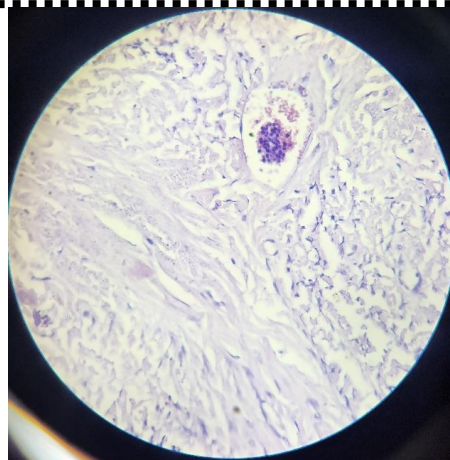


Figure 4

Figure 2, 3, 4: Showing spindle shaped neoplastic cells in fascicles with elongated nuclei without atypia. Stroma shows collagenisation(40 x ,100 x and 400 x H&E)

Diagnosis offered was fibroma arising from right ovary – Atypical Meigs Syndrome.

Discussion

Few patients with Meigs syndrome present with either ascites or pleural effusion along with benign ovarian tumor. These patients present in postmenopausal age group, the peak incidence in their seventh decade^[1,4].

Symptoms related to Meigs syndrome are because of ovarian tumor or effusion. They could remain indolent unless tumor secretes steroid hormones. Androgen excess can present as virilization while excess estrogen present as abnormal uterine bleeding or endometrial neoplasm's. Symptoms related to tumor are abdominal distension due to large tumor mass, urinary incontinence, weight loss or pedal edema.

Symptoms related to pleural effusion are dyspnea, dry cough and pleurisy. Pleural effusion is usually right sided. In our case pleural effusion was not present. Hence the case was classified as Atypical Meigs Syndrome.

Diagnosis of this syndrome is done with imaging studies like USG-Abdomen, CT-scan and X-ray chest^[4]. Fluid analysis reveals either transudate or exudate^[5]. Histopathological examination of ovarian tumors is most

important in diagnosing this syndrome which reveals a benign nature of ovarian tumor.

At present the etiology of pleural effusion and ascites in Meigs syndrome is subject to debate. It probably occurs by means of a transudative mechanism through the surface of the tumor that exceeds the resorptive capacity of peritoneum^[5,6,7]. Other explanations are hormone stimulation, lymphatic obstruction by the tumor, release of inflammatory cytokines and growth factors by the neoplastic cells. Pleural effusions evolve due to translocation of ascitic fluid to the thoracic cavity via diaphragmatic pores. Right sided pleural effusion is commonly seen and maybe because of larger diameter of transdiaphragmatic lymphatic channels on the right side.^[5,8]

Differential Diagnosis of this syndrome includes ovarian carcinoma, liver cirrhosis, other cancers like gastrointestinal cancer, lung cancer and congestive cardiac failure.

Pseudo Meigs syndrome includes ascites and pleural effusion in patients with pelvic and abdominal tumors other than benign tumors included in the definition of Meigs syndrome.^[9]

Meigs syndrome includes Benign ovarian tumor hence it has good prognosis. Pleural effusion and ascites resolve permanently on removal of tumor which is positive definitive treatment.^[5]

Conclusion

Meigs syndrome should be kept in the differential diagnosis of female patients presenting with ascites and pleural effusion. In the absence of either pleural effusion or ascites, the syndrome is classified as Atypical Meigs Syndrome. As the differential diagnosis is metastatic carcinoma, it is important to make correct diagnosis with histopathological examination of ovarian tumor. Early detection with appropriate laboratory investigations and

imaging studies helps in the proper management of patients.

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