Meigs' Syndrome

The three cardinal features of Meigs' syndrome are[1]:

- A benign ovarian tumour (a fibroma, or fibroma-like tumour).
- Ascites.
- Pleural effusion.

Meigs' syndrome is named after Joe Vincent Meigs, an American obstetrician and gynaecologist (1892-1963), who reported a case series in 1937 and subsequently described the syndrome. Meigs proposed limiting the diagnosis of Meigs' syndrome to benign and solid ovarian tumours with ascites and pleural effusion, and with the condition that removal of the tumour cures the patient without recurrence. To conform to the diagnosis of true Meigs' syndrome the ovarian tumour may be a fibroma, thecoma, cystadenoma, or granulosa cell tumour. If the tumour is resected, the fluid resolves.

Epidemiology[2, 3]

Meigs' syndrome is rare. Fibromas account for around 3% of all ovarian tumours, and Meigs' syndrome for 1-2% of those. It tends to occur most often in postmenopausal women, with an average age of presentation of around 50 years.

Presentation

Most features are related to ascites and pleural effusion but before the menopause there may be menstrual symptoms too.

- Fatigue.
- Dyspnoea (initially on exertion).
- Pelvic pain or bloating, constipation.
- Swollen abdomen with associated weight gain or weight loss.
- Non-productive cough.
- Amenorrhoea or irregular menstruation.

Examination

- Reduction in lung capacity may produce tachypnoea and tachycardia.
- Examination of the chest will reveal dullness to percussion over the effusion. There will be decreased breath sounds and decreased tactile vocal fremitus.
- The effusion tends to be right-sided but can be bilateral. There appears to be no adequate explanation for this unilaterality. A large right-sided effusion will displace the mediastinum to the left with deviation of the trachea to the left and displacement of the apex beat.
- Abdominal examination may reveal a tumour arising from the pelvis but this may be obscured by ascites. The features of ascites include a fullness of the flanks and shifting dullness.
- Pelvic examination may reveal an ovarian mass.

Differential diagnosis

The main differential diagnosis is with a malignant ovarian tumour:

- They are much more common than Meigs' syndrome and tend to produce profuse ascites with a high protein content.
- Pleural effusion is less common unless due to pulmonary metastases.
Meigs’ syndrome is a diagnosis of exclusion, once ovarian cancer has been ruled out[1].

Other considerations include:

- Other cancers, particularly colon[4, 5].
- Metastatic breast cancer[6].
- Tuberculosis.
- Nephrotic syndrome.
- Congestive heart failure.
- Cirrhosis.

Investigations

- Check urine for protein.
- Routine blood tests would include FBC, U&E, LFTs, including plasma proteins.
- Cancer antigen 125 (CA 125) is often elevated both in serum and in pleural and peritoneal fluid[1].
- However, CA 125 is not reliable and cases are described with very high CA 125 and with normal levels[7]. It can also be normal in ovarian malignancy.
- CXR (AP and lateral) will show the degree of pleural effusion.
- Abdominal ultrasound will demonstrate the ascites and should outline the ovarian tumour too.
- Imaging also includes CT of the chest, abdomen and pelvis, and MRI of the pelvis.
- Paracentesis and aspiration of pleural fluid:
  - These procedures also help to relieve symptoms. Fluid should be sent for cytology. This is very important in distinguishing malignant ascites from Meigs’ syndrome.
  - The fluid tends to have the features of an exudate, although some studies describe it as a transudate[8]. In ovarian carcinoma the protein content is usually high.
  - Pleural and ascitic fluid should also be examined for protein, glucose, amylase, cell count, organisms and AAFB if indicated.
- If congestive heart failure is suspected, ECG will be required. Echocardiogram is indicated only if the ECG is abnormal.

Associated diseases

- Pseudo-Meigs’ syndrome is characterised by pleural effusion, ascites and benign ovarian tumours other than fibromas as described by Meigs. The tumours may include those of the Fallopian tube, uterus, round ligament, mature teratomas, struma ovarii or ovarian leiomyomas, as well as those originating in the colon, breast and stomach[4].
- Pseudo-pseudo Meigs’ syndrome may occur in patients with systemic lupus erythematosus and describes the presence of pleural effusions, ascites and an elevated CA 125 level[9, 10].
- Atypical Meigs’ syndrome consists of a benign pelvic mass with a right-sided pleural effusion but no ascites present. This is rare. The pleural effusion resolves after resection of the tumour.

Management

The essential management is surgical removal of the tumour. Before operation, aspiration of pleural effusion and ascites may be required to improve pulmonary function[1].

The operation includes full laparotomy to exclude other causes of malignancy, including bowel:

- In women of reproductive age a unilateral salpingo-oophorectomy is usually performed.
- In girls who are before the menarche, wedge resection may be preferred if feasible.
- After the menopause an operation of total abdominal hysterectomy with bilateral salpingo-oophorectomy is usual.
Prognosis

Within weeks to months of operation the ascites and pleural effusion resolve and the CA 125 returns to normal. Postoperative resolution of the fluid is part of the definition of the disease. As it is a benign tumour the prognosis is excellent. If there is functioning ovarian tissue, fertility should be preserved.

Further reading & references

- Joe Vincent Meigs; whonamedit.com


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