

CLINICAL CASE

Ovarian fibrothecoma associated with Meigs' syndrome and elevated serum CA 125

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Summary

Meigs' syndrome consists of a benign ovarian tumor accompanied by ascites and hydrothorax. Elevated serum CA 125 in postmenopausal women with a solid adnexal mass, ascites and pleural effusion is highly suggestive of a malignant ovarian tumor. We report on an unusual case of a benign fibrothecoma of the ovary associated with Meigs' syndrome and elevated CA 125 level in an 62-year-old white female.

Although rarely, a benign ovarian tumor should be considered in the differential diagnosis of an adnexal mass associated with Meigs' syndrome and elevated serum CA 125 levels.

Key words: CA 125 antigen, fibrothecoma, Meigs' syndrome, ovarian tumor

Introduction

Meigs' syndrome consists of a benign ovarian tumor associated with ascites and hydrothorax [1]. Elevated serum carbohydrate antigen 125 (CA 125) levels in postmenopausal women with a solid adnexal mass, ascites and pleural effusion is highly suggestive of an ovarian malignancy. Patients who present with such a clinical picture due to a benign tumor of the ovary, however, may also have elevated serum CA 125 levels [2,3]. An unusual case of Meigs' syndrome due to a left ovarian fibrothecoma with elevated CA 125 is presented.

Case presentation

A 62-year-old white female presented with a history of progressive dyspnea, abdominal girth, weight loss and decline in physical condition over the last 5 months. Approximately one month before her admission she had undergone thoracic drainage of a right pleural effusion in another hospital. From what was reported, the aspirated fluid was an exudate, negative for malignancy. With recurrence of the pleural effusion, increase in abdominal volume due to ascites and a diagnosed pelvic mass, the patient was admitted to our hospital for further evaluation and treatment.

The patient was submitted to closed drainage of a large right pleural effusion which gave almost 4 liters of a clear fluid in a couple of days. Chemical pleurodesis with bleomycin followed the complete evacuation of the effusion. Cytology of the exudate was negative for malignancy. A computerized tomography (CT) scan of the abdomen revealed a solid left adnexal mass, 14X10 cm in size, without clearly defined borders (Figure 1). The presence of free ascitic fluid was also disclosed. No metastatic disease in the liver, omentum or lymphatic basins was found. Serum CA 125 level was 344 IU/ml (normal range 5-35 IU/ml).

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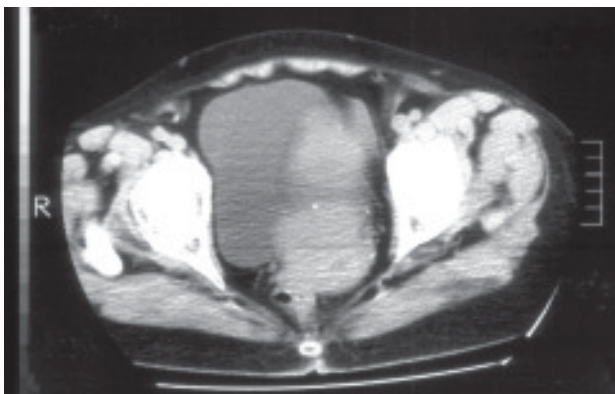


Figure 1. CT scan of the abdomen revealing the presence of a solid left adnexal mass, measuring 14X10 cm and compressing the bladder.

With the clinical preoperative diagnosis of an ovarian malignancy, the patient underwent exploratory laparotomy that brought out evidence of abundant serosanguinous ascites and a left ovarian tumor measuring 8X6.5X3.5 cm with solid lobular appearance. Total abdominal hysterectomy, bilateral salpingo-oophorectomy with complete removal of the large mass, omentectomy and appendicectomy were performed. The patient had an uneventful postoperative course and she was discharged home on the 7th postoperative day. The pathology report ascertained the presence of a benign lesion formed of longitudinal cells with clear cytoplasm and fibrous morphology, arranged in a fascicular fashion. The diagnosis was consistent with a benign fibrothecoma of the ovary (Figure 2). Cytology of the ascitic fluid was also negative for malignancy.

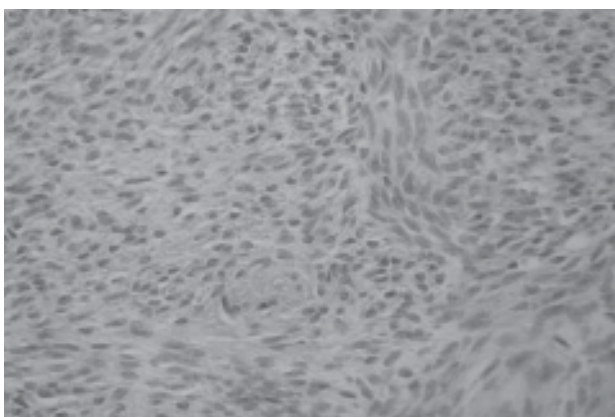


Figure 2. Histopathological examination revealing a stromal lesion with tumor cells arranged in a fascicular fashion. The diagnosis is consistent with a benign fibrothecoma of the ovary (H&E X40).

The patient was asymptomatic thereafter with a normal serum CA 125 level (26.6 IU/ml) on the 30th postoperative day.

Discussion

Elevated serum CA 125 in postmenopausal women with a solid adnexal mass, ascites and pleural effusion are highly suggestive of a malignant ovarian tumor. Patients with Meigs' syndrome, however, can also have elevated tumor markers, thus, obscuring the correct preoperative diagnosis. The presented case is an extremely unusual example of an ovarian tumor associated with abnormal CA 125 levels together with cytologically negative pleural and ascitic fluid, which proved to originate from a benign fibrothecoma of the ovary. Only 15 cases of Meigs' syndrome with elevated CA 125 levels have been described in the literature [4]. The most common histological types of the presented benign ovarian tumors were cellular fibroma (4 cases), thecoma (4 cases), fibroma-thecoma (3 cases), fibroma (3 cases), and granulosa cell tumor (1 case). Serum CA 125 levels ranged from 42.3 to 2120 IU/ml. One patient presented with levels higher than 5000 IU/ml [4].

Fibrothecoma of the ovary is a rare ovarian tumor associated with Meigs' syndrome in less than 2% of the cases [5,6]. Several hypotheses might explain the accumulation of ascitic fluid in Meigs' syndrome. The main theory is that the transudation mechanism through the tumor mass exceeds the capacity for reabsorption from the peritoneal surface. Another suggested mechanism implicates the congestion of the peritoneal lymphatic vessels and regional veins caused by the neoplastic lesion itself or by vasoactive substances released by the tumor cells [3].

It is presumed that the occurrence of pleural effusion is secondary to the passage of ascitic fluid to the pleural space through the diaphragm or diaphragmatic vessels. Alternatively, congenital defects of the diaphragm, which are more common on the right side, may contribute to the development of a clinically significant right hydrothorax [7].

The CA 125 tumor marker is generally elevated in patients with malignant ovarian tumors. It can, however, be markedly elevated in benign disorders, such as endometriosis, pelvic inflammatory disease and uterine leiomyoma. Serum CA 125 levels can also increase in pericardial, pleural and peritoneal irritation or inflammation [4,8]. As a result, exploratory laparotomy or laparoscopy and precise histopathological examination are frequently required for the cor-

rect diagnosis and treatment since high serum CA 125 by itself cannot predict the presence of an ovarian malignancy [4].

Postmenopausal women with clinical palpable pelvic mass, pleural effusion, ascites and elevated serum CA 125 levels probably suffer from malignant ovarian tumors. Benign fibrothecoma of the ovary, however, as part of Meigs' syndrome, should be also considered as an unusual but existing diagnostic possibility.

References

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