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Enormous ovarian fibroma with elevated Ca-125 associated with Meigs' syndrome. Presentation of a rare case

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Summary

In medicine, Meigs' syndrome is the triad of ascites, pleural effusion, and benign ovarian tumor (fibroma, Brenner tumour, and occasionally granulosa cell tumour). It resolves after the resection of the tumor. Because the transdiaphragmatic lymphatic channels are larger in diameter on the right, the pleural effusion is classically on the right side. The etiologies of the ascites and pleural effusion are poorly understood. Atypical Meigs' syndrome,characterized by a benign pelvic mass with right-sided pleural effusion but without ascites, can also occur. As in Meigs syndrome, pleural effusion resolves after removal of the pelvic mass. The authors would like to share their own experience of a case of Meigs' syndrome associated with an enormous ovarian fibroma and elevated Ca-125.

Key words: Meigs' syndrome; Ascites; Ovarian fibroma.

Introduction

The ovarian cancer represents the sixth most common cancer in women worldwide with the highest morbidity. It is predominantly a disease among postmenopausal women with an average age at diagnosis of 60 years [1]. The combination of solid adnexal mass, elevated Ca-125 antigen, ascites, and pleural effusion suggests preoperatively the existence of a malignant ovarian tumor [2]. The final diagnosis is defined after the excision of the tumor and the histological confirmation. On the contrary, there are many cases with similar clinical symptoms, among postmenopausal women, in which the histological confirmation reveals a benign condition, known as Meigs' syndrome. There are few cases reported in the current literature [3] (Table 1).

The authors want to share their experience of a case of a 61-year-old female patient with an enormous right ovarian fibroma associated with elevated Ca-125 antigen and Meigs' syndrome.

Case Report

A 61-year-old patient (grava 6, para 3), with a history of hyperlipidaemia and hypertension was admitted to the Department of Obstetrics and Gynecology, in Konstantopouleio-Patision General Hospital, complaining of severe abdominal pain and fever. The physical examination revealed an enormous palpable pelvic mass that occupied the entire pelvic cavity up to the height of the navel. The uterus identification was very difficult due to the pelvic mass, which was ponderous and located at the region of the right adnexa. The patient received the appropriated prophylactic antibiotic treatment.

An ultrasound revealed the presence of solid mass 109 x 109 x 96 mm, probably located at the region of the right adnexa and ascitic effusion diffuse into the peritoneal cavity. The Pap-smear was free of malignancy. The Ca-125 tumor marker count was 210.10 U/ml (normal range 0-35). All others revealed no signs of malignancy.

A CT examination showed the presence of solid lesion maximal diameter 110 x 100 x 90 mm with clear and smooth boundaries, which began from the height of the navel and reached the minor pelvis and there was the presence of ascetic and pleural effusion. There were no signs of enlarged or infiltrated para-aortic, iliac or inguinal lymph nodes. The MRI examination confirmed the presence of solid mass maximal diameter 11.5 cm in contact with the uterus. The mass reached the promontorium, containing a cystic section with extended proliferations, mainly in the posterior part. These findings were consistent with primary ovarian malignancy.

The patient underwent a exploratory laparotomy. It revealed a solid red-yellow necrotic tumor maximal diameter 15 cm located at the right ovary. The omentum, the bladder, and loops of small bowel were solid and adhered to the tumor. There was an image of diffuse inflammation, an obtuse detachment of the intestine, and other tissues from the tumor. The frozen section of the right adnexa revealed the presence of ovarian fibroma. The patient underwent total hysterectomy and left salpingo-oophorectomy.

The histological report revealed the presence of ovarian tumor with a maximal diameter of 13 cm with attributes compatible with ovarian fibroma. The tumor consisted of plenty of thrombosed vessels, extensive hemorrhage and sections of cystic degeneration findings due to torsion of the tumor. The patient had an uneventful course and was discharged at the seventh postoperative day.

Discussion

Meigs' syndrome is defined as the triad of benign ovarian tumor with ascites and pleural effusion that resolves

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Table 1. — *Summary of reported cases of Meigs' syndrome with elevated serum Ca-125 level [3].*

Author	Year	No. of cases	Age	Histopathology U/ml	Ca125
Jones and Surwit	1989	1	70	Fibroma/thecoma	226
Hoffman	1989	1	39	Thecoma	498
Martin *et al.*	1990	1	NR	Granulosa cell tumor	307
Walker *et al.*	1990	2	52, 67	Cellular fibroma	>5,000
Le Bouedec *et al.*	1992	2	66, 76	Fibroma /thecoma	645
Williams *et al.*	1992	1	74	Luteinized Thecoma	329
Lin *et al.*	1992	2	74, 72	Fibroma	2,120 7,000
Turan *et al.*	1993	1	63	Thecoma	744
Sidiqui et Toub	1995	1	73	Celular fibroma	1,780
Tmmerman *et al.*	1995	2	71,73	Fibroma	484.5 42,3
Abad *et al.*	1997	1	51	Cellular fibroma	577
Chan *et al.*	2000	1	13	Fibroma	970
Patsner	2000	6	62, 57, 52, 60, 72, 58	Fibroma	185, 850 850, 520 64, 1,200 80
Buttin *et al.*	2001	1	67	Brenner tumor	759
Lopez *et al.*	2002	2	78,68	Fibroma	498, 265
Vireira *et al.*	2003	1	65	Thecoma	319
Moran-Mendosa *et al.*	2004	1	46	Fibroma	1,808
Mongkol Benjapibal *et al.*	2009	1	56	Fibroma	1,064

after the resection of the tumor [3]. Ovarian fibroma is reported in 2-5 % of surgically removed ovarian tumors and Meigs' syndrome is observed in about 1% [4].

In many cases, not only the physical examination, but also the imaging findings reveal an image compatible with malignancy. After the resection of the tumor, the final histological examination confirms the presence of an ovarian fibroma and secondary the existence of Meigs' syndrome. If the operative management is appropriate, the prognosis seems to be very good. Life expectancy after surgical removal of the tumor equivalences that of the general population [5].

Rarely ovarian malignancy and less frequently metastatic tumors of the ovaries can reveal the same syndrome, characterizing with benign hydrothorax and ascites. In these cases, the condition is called Pseudomeigs' syndrome. [6] In both syndromes, the tumor excision represents the optimal management and indicates the increase of hydrothorax and ascites [7].

One of the most still not answered mechanisms remains the pathophysiology of Meigs' syndrome. [8] According to current literature, it is considered unclear and represents a controversial issue. The presence of the tumor can trigger mediators and other inflammatory substances, which can increase the capillary permeability and lead to production of peritoneal fluid [9]. We must never forget the role of hormonal stimulation and the lymphatic or vessel effect. Pleural effusion is correlated with the peritoneal fluid. The outspread consists in lymph vessels spreading through the diaphragm [10].

The Ca-125 tumor marker indicates the possibility of an ovarian malignancy but it is found elevated during some physiologic conditions such as menstruation or pregnancy and in some benign conditions such as endometriosis, peritonitis, uterine leiomyoma, pleuritis, pericarditis, and peritonitis [11].

Exploratory laparotomy and histological confirmation are required in order to establish the correct diagnosis, since elevated serum Ca-125 levels can be falsely positive for ovarian malignancy [12].

Conclusion

In many cases the combination of pelvic mass, peritoneal fluid, and pleural effusion with elevated serum Ca-125 level indicates an ovarian malignancy. On the contrary, Meigs' syndrome mimics the same clinical course changing the prognosis and the survival rate of the patients. The final diagnosis can be established through the resection of the tumor and the histological confirmation.

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