International Journal of Research in Medical Sciences
Pinzon CLA et al. Int J Res Med Sci. 2020 Oct;8(10):3706-3709
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20204256

Case Report

Meigs syndrome: literature review of a case report

Camilo Levi Acuña Pinzón[1]*, Diego Hinojosa Ugarte[1], Aldo Edyair Jimenez Herevia[1], Miguel Angel Guerrero[2]

[1]Department of Surgery, Hospital Regional de Alta Especialidad del Bajío, Leon-Guanajuato, México
[2]Department of Surgery, Surgical Oncology, Hospital Regional de Alta Especialidad del Bajío, Guanajuato, México

Received: 24 July 2020
Revised: 10 August 2020
Accepted: 03 September 2020

***Correspondence:**
Dr. Camilo Levi Acuna Pinzon,
E-mail: camilolevi@hotmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Ovarian cancer is the presence of one or multiple tumors, which appears in one or both ovaries. These tumors are usually classified as epithelial and non-epithelial. Sex cord-stromal tumors are a group of benign and malignant neoplasms that develop from the sexual cord. Many are functional and therefore have hormonal secretions. Meigs syndrome is defined by the presence of pleural effusion and ascites in association with an ovarian tumor. We present a case of a 55 years old patient who was admitted due to a pelvic tumor, exudative pleural effusion that was difficult to manage, and ascites. A benign ovarian stromal tumor associated with elevated Ca 125 was diagnosed. After the management of the effusions, a 20x20x10 cm ovarian tumor resection was performed by laparotomy, and a transoperative report of a thecoma/fibroma type stromal tumor was received. Meigs syndrome occurs in 1% of ovarian tumors, being very rare before the third decade of life, the pathogenesis of ascites and pleural effusion could be related to the imbalance of hydrostatic forces between arterial flow and lymphovenous drainage culminating in a stromal transudate. Treatment of this syndrome should be focused on tumor debulking surgery and symptomatic treatments such as chest tubes and pleurodesis.

Keywords: Meigs syndrome, Ovary cancer, Effusion pleural, Ascites

INTRODUCTION

Ovarian cancer is the appearance of one or multiple tumors, which appears in one or both ovaries.[1] These tumors are usually classified as epithelial and non-epithelial (germinal, stromal-sexual cords). Ovarian sex cord-stromal tumors (SCST) are a group of benign and malignant neoplasms that develop from the sex cord (sertoli cell tumor, granulosa cell tumor) or stromal cells (fibroma, thecoma, leydig cell tumor) or both (sertoli-leydig cell tumor). Many of the SCST are functional; therefore, they have hormonal secretions, particularly androgens and estrogens. Ovarian SCSTs are less common than epithelial and germ cell tumors.[2]

With the advent of new diagnostic and therapeutic techniques in the '90s, its incidence, prevalence, and mortality have decreased. However, it continues to rank as the ninth leading cause of cancer in women and the fifth leading cause of cancer-related death worldwide.[3] In 2018 in Mexico, GLOBOCAN registered 4,759 new cases of ovarian cancer, obtaining the sixth place of incidence nationwide (Table 1). Having a higher incidence in the sixth decade of life; the estimated number of deaths, as well as the prevalence (in the last 5 years) of ovarian cancer, was 2,765 and 12,942 respectively. It is estimated that for this 2020 there will be 5031 new cases and 2951 deaths from this entity.[4] Among these, benign tumors represent 0.5 to 3.7% of all benign tumors.[5]

Meigs syndrome is defined as the presence of pleural effusion and ascites in association with a benign ovarian tumor.[6]

CASE REPORT

55 years old woman, with no significant history, which started with symptoms 7 months previously, consisting of left costal pain without relief after the administration of NSAIDs, she consulted a primary care hospital where there was evidence of chest radiography with bilateral pleural effusion, Ultrasound of the abdomen with a heterogeneous tumor of cystic components 16.9x9.9x 15.6 cm and ascites. Marker Ca 125 of 1849 ug/ml, for what they decide to refer to our third level institution.

Thoracoabdominal CT was performed where bilateral pleural effusion (100% of the left chest), a dependent tumor of the left ovary, and ascites were observed. A left chest tube and a fluid sample were done, with a high protein and low cellular reported. Due to the persistent flow of chest tube greater than 10 days, pleurodesis was performed. One week later, a chest X-ray was performed with a 70% contralateral pleural effusion reported, for which thoracentesis was decided. Two weeks after tube placement in the left chest, the left pleural effusion persisted, so the pleurodesis was repeated. A percutaneous biopsy was done, without conclusive result, so a laparotomy was performed, finding a 20x20x10cm ovarian tumor which was resected, and a trans-operative report of a tecoma/fibroma like stromal tumor was reported.

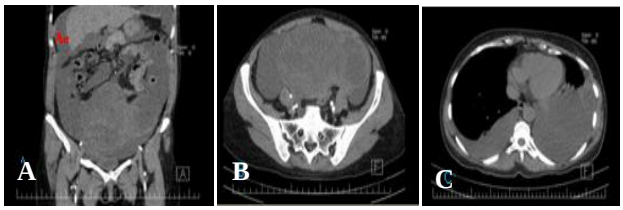

Figure 1: TAC, A) and B) cystic appearance of ovarian tumor as ascites, C) bilateral pleural effusion.

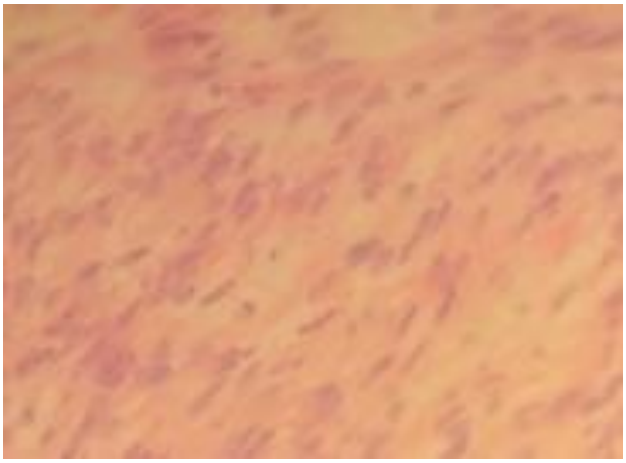

Figure 2: Trans-operative study.

Table 1: Incidence of appearance of the different types of cancers nationwide in Mexico.

Cancer	Number	Interval of uncertainty	Gross rate	ASR (world)
All cancers	105 051	(103781.0-106336.0)	160,0	149,6
Breast	27 283	(25975.8-28656.0)	41,6	39,5
Thyroid	10 188	(8729.8-11889.8)	15,5	14,6
Cervix	7 869	(7469.1-8290.3)	12,0	11,0
Uterine body	7 266	(6311.5-8364.9)	11,1	10,6
Colorectal	7 105	No disponible	10,8	10,0
Ovary	4 759	(4375.8-5175.8)	7.2	6.8

DISCUSSION

The clinical presentation of ovarian neoplasms can be acute or subacute. Women who present acutely will generally have advanced disease with a condition that requires urgent attention and evaluation (pleural effusion, intestinal occlusion). The most frequent form of clinical presentation is subacute (pelvic or abdominal pain, gastrointestinal symptoms) in women with early or advanced disease.[7]

Meigs syndrome occurs in 1% of ovarian tumors, being very infrequent before the third decade of life but increasing progressively until reaching a peak in the seventh decade of life.[8]

The pathogenesis of ascites and pleural effusion could be related to hydrostatic forces, which result in the formation of transudates at the tumor level due to a mismatch between arterial flow and lymphatic-venous drainage. However, its mechanism is still unknown, so different theories are proposed. One of them proposes the filtration of interstitial fluid to the peritoneum through the ovarian capsule of the tumor, later this fluid moves from the peritoneal cavity to the pleural due to diaphragmatic defects or via lymphatic channels causing exudative pleural effusion. According to another hypothesis, direct pressure on the lymphatic vessels within the tumor can cause leakage from the superficial lymphatic system located just below the lining of the neoplasm. Other authors postulate that ascites is caused by irritation on the surface of the peritoneum that is in contact with the tumor. Most of the hypotheses seem to converge, however, in the pathogenesis of hydrothorax, it is considered secondary to ascites, which is also related to pelvic mass, despite the fact that in some cases hydrothorax is the first manifestation of the syndrome. An imbalance between blood supply, tumor size, and

venous and lymphatic drainage may be responsible for edema and stromal transudation.[8,9]

Some studies have suggested that fluid accumulation could be related to proteins such as endothelial growth factor that increases capillary permeability.[8]

Approximately 70% of pleural effusions are on the right side, 15% are on the left side and, as in our patient, 15% are bilateral.[8] Interleukins 1B, 6, 8, and tumor necrosis factor-alpha have been implicated in the pathogenesis of ascites and hydrothorax, probably due to the hyperpermeability of the ovarian and peritoneal vasculature with consequent fluid transudation.[6] Classically, the pleural effusion of Meigs syndrome has been described as transudative; However, a meta-analysis concluded that most reports describe it as exudates.[10]

The Ca 125 antigen is a glycoprotein with a high molecular weight recognized by the OC 125 antibody, being used as a tumor marker associated with ovarian carcinoma. There are some case reports of Meigs syndrome, such as the one described in our patient, where elevated Ca 125 is found; The mechanism by which this elevation occurs is not yet known, however immunohistochemical staining for Ca 125 suggests that serum elevation of Ca 125 antigen is secondary to mesothelial expression.[8] A Ca 125 level greater than 1000 U/mL is quite unusual.[6]

Among the differential diagnoses of Meigs syndrome are malignant ovarian tumors, lung or intestinal cancer, nephrotic syndrome, congestive heart failure, liver cirrhosis, and tuberculosis.[8]

Using ultrasound, ascites and pleural effusion can be detected with the presence of a well-defined adnexal mass without increased vascularity. Other studies such as magnetic resonance imaging and tomography can be considered to exclude metastatic disease before treatment. Despite the great contribution of imaging techniques, the diagnostic suspicion of Meigs syndrome is made through the clinic and the final diagnosis is usually post-surgical with the resolution of ascites, pleural effusion, and histological confirmation of the tumor.[8]

The treatment of choice for the tumor is exploratory laparotomy with unilateral oophorectomy in premenopausal women or bilateral oophorectomy in cases of menopausal or perimenopausal women.[11] In the literature, most ovarian fibroids were removed by laparotomy; a laparoscopic approach has rarely been reported. This may be due to the large size of the tumors at presentation and the suspicion of being a malignant ovarian disease.

With a better preoperative diagnosis, the laparoscopic approach can be successfully performed, especially when the tumor is small. Sample extraction is difficult, time-consuming, and requires the use of electrical morcellation. As in other cystic ovarian tumors, care must be taken to avoid contamination and to leave any tumor fragment in the peritoneal cavity during morcellation, as unexpected malignancy occasionally occurs.[12] The management of pleural effusion and ascites includes the performance of thoracentesis and paracentesis, respectively.[8]

With the resection of the ovarian tumor, ascites and pleural effusion resolve; furthermore, the life expectancy after surgical resection of the tumor is the same as in the general population.[5,8] In our case; the pleural effusion required thoracostomy tube placement and two pleurodeses prior to resection of the adnexal lesion, after which she had a good evolution and was discharged.

In conclusion, Meigs syndrome is a rare entity that requires a high level of clinical suspicion in patients with exudative pleural effusion and elevated Ca 125 levels.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Matulonis UA, Sood AK, Fallowfield L, Howitt BE, Sehouli J, Karlan BY. Ovarian cancer. Nature Reviews Disease Primers. 2016;2(1):16061.
2. Gershenson D. Sex cord-stromal tumors of the ovary: Epidemiology, clinical features, and diagnosis in adults.Available at: https://www.uptodate.com. Accessed on 1 May 2020.
3. Hoffman BL, Schorge JO, Bradshaw KD, Halvorson LM, Schaffer JI, Corton MM. Cáncer ovárico epitelial. In: Williams Ginecología. 3rd ed. New York, NY: McGraw-Hill; 2017.
4. Global cancer observatory. Available at: https://gco.iarc.fr/. Accessed on 28 March 2020.
5. Hanley KZ, Mosunjac MB. Practical Review of Ovarian Sex Cord-Stromal Tumors. Surgical pathology clinics. 2019;12(2):587-620.
6. Riker D, Goba D. Ovarian mass, pleural effusion, and ascites: revisiting Meigs syndrome. Journal of bronchology & interventional pulmonology. 2013; 20(1):48-51.
7. Hen L, Berek JS. Epithelial carcinoma of the ovary, fallopian tube, and peritoneum: Clinical features and diagnosis. Available at: https://www.uptodate.com. Accessed on 1 May 2020.
8. Saha S, Robertson M. Meigs' and Pseudo-Meigs' syndrome. Australasian journal of ultrasound in medicine. 2012;15:29-31.
9. Barranco R, Molinelli A, Gentile R, Ventura F. Sudden, unexpected death due to pseudo-meigs syndrome: a case report and review of the literature. The American journal of forensic medicine and pathology. 2019;40(1):89-93.
10. Krenke R, Maskey-Warzechowska M, Korczynski P, Zielinska-Krawczyk M, Klimiuk J, Chazan R, et al.

Pleural effusion in meigs' syndrome-transudate or exudate?: systematic review of the literature. Medicine. 2015;94(49):e2114.
11. Fremed MA, Levin TL, Sun KY, Weiser DA. Ovary preservation in the treatment of childhood Meigs syndrome. Pediatric blood & cancer. 2015;62(11): 2011-4.
12. Leung SW, Yuen PM. Ovarian fibroma: a review on the clinical characteristics, diagnostic difficulties, and management options of 23 cases. Gynecologic and obstetric investigation. 2006;62(1):1-6.

Cite this article as: Pinzon CLA, Ugarte DH, Herevia AEJ, Guerrero MA. Meigs syndrome: literature review of a case report. Int J Res Med Sci 2020;8:3706-9.