Scientific Research Publishing

Open Journal of Pathology, 2023, 13, 73-78
https://www.scirp.org/journal/ojpathology
ISSN Online: 2164-6783
ISSN Print: 2164-6775

Demons-Meigs Syndrome with a High CA 125 Level: A Case Report at Soavinandriana Center Hospital Antananarivo

Razafindrafara Herilalao Elisabeth[1], Razafimahefa Vahatra Joëlle[2], Rakotondrainibe Fenohery Nalisoa[3], Randimbinirina Zakarimanana Lucas[4], Rabarison Manoahasina Ranaliarinosy[5], Randrianjafisamindrakotroka Nantenaina Soa[5]

[1]Department of Pathology, Soavinandriana Center Hospital (CENHOSOA), Antananarivo, Madagascar
[2]Department of Pathology, University Hospital Andrainjato, Fianarantsoa, Madagascar
[3]Department of Pathology, Anosiala University Hospital, Antananarivo, Madagascar
[4]Department of Surgery, Soavinandriana Center Hospital (CENHOSOA), Antananarivo, Madagascar
[5]Department of Pathology, Joseph Ravoahangy Andrianavalona University Hospital, Antananarivo, Madagascar
Email: rherilalaoelisabeth@yahoo.fr

How to cite this paper: Elisabeth, R.H., Joëlle, R.V., Nalisoa, R.F., Lucas, R.Z., Ranaliarinosy, R.M. and Soa, R.N. (2023) Demons-Meigs Syndrome with a High CA 125 Level: A Case Report at Soavinandriana Center Hospital Antananarivo. *Open Journal of Pathology*, **13**, 73-78.
https://doi.org/10.4236/ojpathology.2023.132007

Received: December 28, 2022
Accepted: February 5, 2023
Published: February 8, 2023

Abstract

Introduction: Demons-Meigs syndrome combines a benign tumor of the ovary with ascites and pleural effusion. It is a rare disease and the pathophysiological mechanism is not yet well understood. The aim of our study is to report a case of Demons-Meigs syndrome, to determine its different characteristics and to make clinicians aware of the malignant predictive value of CA 125 in front of an ovarian tumor. **Observation:** The patient was 42 years old and had no previous history. She presented with intense abdominal pain of sudden onset, prompting a medical consultation. On clinical examination, the abdomen was distended with signs of peritoneal effusion and right pleural effusion. Ultrasound revealed a well-circumscribed, ovarian mass with an internal fluid component and hypervascularization on Doppler. The CA 125 level was elevated (293.9 U/ml). Exploratory laparotomy revealed two bilateral, firm, solid ovarian tumors without vegetation or peritoneal lesions. Macroscopically, one of the tumors was well circumscribed, lobulated with a smooth outer surface. It measured 20 × 17 × 8 cm and weighed 1400 g. The other tumor measured 19 × 11 × 5 cm, weighed 1090 g and had the same characteristics as the other tumor. Histologically, both tumors were a proliferation of fibroblastic spindle cells organized in short intersecting or storiform bundles, without cyto-nuclear atypia or excess mitoses. The diagnosis retained was bilateral ovarian fibroma in the context of a Demons-Meigs syndrome. **Conclusion:** Demons-Meigs syndrome is a rare entity. The concomitant elevation of the CA 125 level is not always an indicator of ovarian cancer.

The curative treatment is surgical based on tumor removal ensuring the disappearance of peritoneal and pleural effusions.

Keywords

Ovarian Tumor, Demons-Meigs Syndrome, Ascite, Pleural Effusion, CA 125, Madagascar

1. Introduction

Demons-Meigs syndrome combines a benign ovarian tumor, ascites and pleural effusion. It is a rare pathology and its pathophysiology remains obscure [1]. The ovarian tumor frequently corresponds to a fibroma and a thecoma [2]. Fibroma accounts for 1% to 4% of ovarian tumors and is observed in 1% of cases in the context of Demons-Meigs syndrome [3]. The aim of our study is to report a case of Demons-Meigs syndrome, to determine its different characteristics and to make clinicians aware of the malignant predictive value of CA 125 in front of an ovarian tumor.

2. Observation

The patient was 42 years old and had no previous history. Several weeks before her admission, she presented with intense abdominal pain of sudden onset, triggered by prolonged sitting and calmed by non-steroidal anti-inflammatory drugs, prompting a medical consultation. On clinical examination, the abdomen was distended with signs of peritoneal effusion and right pleural effusion. Ultrasound revealed a well-circumscribed, irregularly contoured ovarian mass with an internal fluid component (**Figure 1**) and hypervascularization on Doppler. There was another right lateropelvic mass, tissue, hypoechoic, heterogeneous, measuring 119 × 89 mm, associated with a cluster of right iliac lymph nodes. These lesions were accompanied by a large, finely echogenic intraperitoneal fluid effusion and a medium-sized, nonpartitioned right pleural fluid effusion. The CA 125 level was elevated (293.9 U/ml). Exploratory laparotomy revealed two bilateral, firm, smooth ovarian tumors without vegetation or peritoneal lesions, which were removed (**Figure 2**). Macroscopically, one of the tumors was well circumscribed, lobulated with a smooth outer surface. It measured 20 × 17 × 8 cm and weighed 1400 g. The section slices were white, solid, firm, heterogeneous with cystic areas of 0.5 cm to 4 cm with mucoid content. The other tumor measured 19 × 11 × 5 cm, weighed 1090 g and had the same characteristics as the other tumor. Histologically, both tumors were a proliferation of fibroblastic spindle cells organized in short intersecting or storiform bundles, without cyto-nuclear atypia or excess mitoses (**Figure 3** and **Figure 4**). In some places, tubular or tubulocordal structures were identified. The diagnosis retained was bilateral ovarian fibroma in the context of a Demons-Meigs syndrome. Regarding

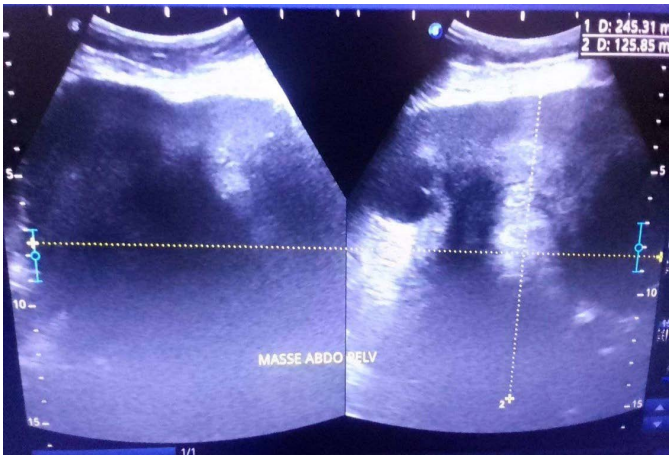

Figure 1. Ultrasound: Well circumscribed ovarian mass with an internal fluid component. Source: Department of Surgery, Soavinandriana Center Hospital (CENHOSOA), Antananarivo Madagascar.

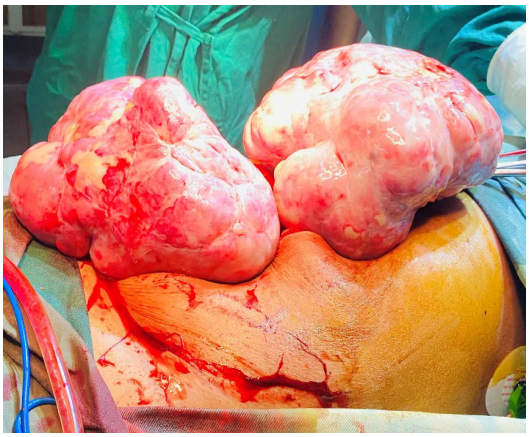

Figure 2. Bilateral ovarian tumors. Well circumscribed mass with, smooth, lobulated surface. Source: Department of Surgery, Soavinandriana Center Hospital (CENHOSOA), Antananarivo Madagascar.

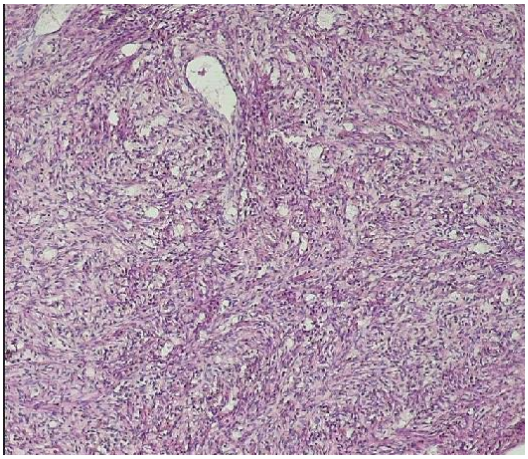

Figure 3. Stromal ovarian tumor with fibroblastic cells. Bland spindled to ovoid, nuclei with pointy ends and eosinophilic cytoplasm, HE ×100. Source: Department of Pathology, Soavinandriana Center Hospital (CENHOSOA), Antananarivo, Madagascar.

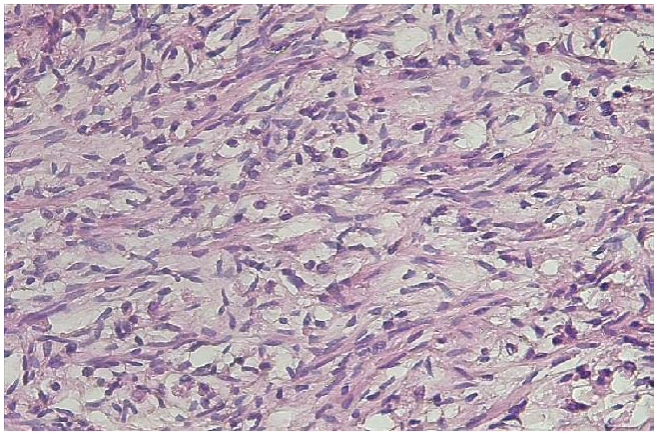

Figure 4. Benign ovarian fibroma composed of fibroblastic cells within a variably collagenous stroma, HE ×400. Source: Department of Pathology, Soavinandriana Center Hospital (CENHOSOA), Antananarivo, Madagascar.

the management, follow up and prognosis, the postoperative course was simple with disappearance of the pleural and peritoneal effusions at the 45th postoperative day.

3. Discussion

Demons-Meigs syndrome is defined by the triad of benign ovarian tumor, ascites and pleural effusion [1]. The ovarian tumor frequently corresponds to a fibroma and a thecoma [2]. Fibroma accounts for 1% to 4% of ovarian tumors and is observed in 1% of cases in the context of Demons-Meigs syndrome [3].

Demons-Meigs syndrome is much more common in postmenopausal women, especially those around 50 years of age, and its peak incidence is in the seventh decade. It is extremely rare in women under 30 years of age [4] [5]. In the case of our patient, she was a 42-year-old woman near perimenopause.

In Demons-Meigs syndrome, ovarian tumors can be fibroma, thecoma, cystadenoma, or granulosa tumor, but the most frequently reported are fibroma and thecoma [2]. Fibroma is often unilateral but is sometimes bilateral. In our patient, it was a bilateral fibroma. Other tumors such as teratoma or struma ovarii may be associated with ascites and pleural effusion but in these cases, according to Peparini N and al, it would be a pseudo Demons-Meigs syndrome [6].

Several theories would explain the appearance of peritoneal and pleural effusions in Demons-Meigs syndrome [7]. First, mechanical theories, by vascular compression or cardiac decompensation were evoked. Then, the secretory theory, by mechanical irritation of the peritoneum due to a tumor (foreign body) or to the existence of a germinative secretory epithelium on the tumor was noted. Then, anaphylactic theories were added by histaminic toxicosis of the peritoneum by vasodilating histamine. Endocrine theories by endocrine dysregulation of the hyperestrogenic type could be due to the presence of a secreting tumor, either by minimal thecoma or by islands of thecal proliferation [7]. According to the experiments of Meigs [2], the persistence of the pleuroperitoneal

duct would explain the pleural effusions. According to Liou JH [8], pleural effusions result from transudation of fluids through the transdiaphragmatic lymphatic trunks classically on the right side. Ascites would be attributed to fluid transudation related to direct pressure on the surrounding lymphatics and vessels, hormonal stimulation, or tumor torsion [8].

A significant elevation of the CA 125 level to a normal of less than 35 IU/l raises the suspicion of an ovarian malignancy. In our observation, the CA 125 level was 293.9 IU/l. During Demons-Meigs syndrome, it is classical to demonstrate an elevation of CA 125 which is an antigenic marker of the coelomic epithelium and its derivatives [9]. Several authors such as Plaisant N and al [9] or Santangelo M and al [1] have demonstrated that an elevated CA 125 level is not a good indicator of ovarian malignancy.

Treatment is based on surgical removal of the ovarian tumor which ensures that the effusions are dried up [10]. In our patient, she underwent bilateral oophorectomy. The postoperative course was simple with disappearance of the pleural and peritoneal effusions at the 45th postoperative day.

Demons-Meigs syndrome is a benign condition with a good prognosis with early management. Pleural effusion and ascites disappear permanently after tumor removal. The postoperative life expectancy of patients with this syndrome is equivalent to that of the general population after surgery [11].

4. Conclusion

Demons-Meigs syndrome is a rare entity. The concomitant elevation of the CA 125 level is not always an indicator of ovarian cancer. The curative treatment is surgical based on tumor removal ensuring the disappearance of peritoneal and pleural effusions.

Consent from the Patient

We obtained the consent from the patient for the case study.

Conflicts of Interest

The authors declare no conflict of interest.

References

[1] Santangelo, M., Battaglia, M., *et al.* (2000) Meigs' Syndrome: Its Clinical Picture and Treatment. *Annali Italiani di Chirurgia*, **71**, 115-159.

[2] Bretelle, F., Portier, M.P., Boubli, L. and Houvenaeghel, G. (2000) Recurrence of Demons-Meigs Syndrome. A Case Report. *Annales de Chirurgie*, **125**, 269-272. https://doi.org/10.1016/S0001-4001(00)00128-8

[3] Gargano, G., De Lina, M., Zito, F., *et al.* (2004) Ovarian Fibroma: Our Experience of 34 Cases. *European Radiology*, **14**, 798-804.

[4] Saha, S. and Robertson, M. (2012) Meigs's and Pseudo-Meigs' Syndrome. *Australasian Journal of Ultrasound in Medicine*, **15**, 29-31. https://doi.org/10.1002/j.2205-0140.2012.tb00140.x

[5] Shiau, C.S., Chang, M.Y., Hsieh, C.C., Hsieh, T.T. and Chiang, C.H. (2005) Meigs's Syndrome in a Young Woman with a Normal Serum CA-125 Level. *Chang Gung Medical Journal*, **28**, 587-591.

[6] Peparini, N. and Chirletti, P. (2009) Ovarian Malignancies with Cytologically Negative Pleural and Peritoneal Effusions: Demons' or Meigs' Pseudo-Syndromes? *International Journal of Surgical Pathology*, **17**, 396-397. https://doi.org/10.1177/1066896909336441

[7] Nemeth, A.J. and Patel, S.K. (2003) Meigs Syndrome Revisited. *Journal of Thoracic Imaging*, **18**, 100-103. https://doi.org/10.1097/00005382-200304000-00009

[8] Liou, J.H., Su, T.C. and Hsu, J.C. (2011) Meigs' Syndrome with Elevated Serum Cancer Antigen 125 Levels in a Case of Ovarian Sclerosing Stromal Tumor. *Taiwanese Journal of Obstetrics and Gynecology*, **50**, 196-200. https://doi.org/10.1016/j.tjog.2011.01.011

[9] Plaisant, N., Courtieu, C., *et al.* (1999) Demons-Meigs' Syndrome and Elevated CA125: Diagnosis of Ovarian Cancer and Therapeutic Implications. *Contracept Fertil Sex*, **27**, 47-50.

[10] Le Bouëdec, G., Glowaczower, E., *et al.* (1992) Demons-Meigs' Syndrome. A Case of Thecoma and Ovarian Fibroma. *Journal de Gynécologie Obstétrique et Biologie de la Reproduction*, **21**, 651-654.

[11] Kortekaas, K.E., and Pelikan, H.M. (2018) Hydrothorax, Ascites and an Abdominal Mass: Not Always Signs of a Malignancy—Three Cases of Meigs's Syndrome. *Journal of Radiology Case Reports*, **12**, 17-26. https://doi.org/10.3941/jrcr.v12i1.3209