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Demons-Meigs Syndrome: A Case Report at the Fatick Regional Hospital

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Abstract

Demons-Meigs syndrome combines a benign tumor of the ovary with thoracic and abdominal effusion, it is a rare disease whose pathophysiological mechanism is not yet well understood. This rare syndrome often gives rise to fear of ovarian neoplasia due to the existence of ascites and the significant elevation of CA 125. In this observation, we report the case of Demons-Meigs syndrome diagnosed in a 36-year-old young woman who presented with pleural and peritoneal effusion syndrome with an ovarian mass and a CA 125 level = 406.6 IU/ml. Laparotomy performed revealed very abundant ascites, the uterus and tubes were healthy, no suspicious peritoneal lesions, the left ovary is healthy, a right ovarian mass of 300 × 150 mm. A right annexectomy was performed. The operative suites were simple with drying of the effusions. The control CA 125 returned to normal after 6 months of follow-up. A pathological anatomy result is a fibrothecoma. Demons-Meigs syndrome has a good prognosis; treatment is based on removal of the ovarian tumor.

Keywords

Demons-Meigs Syndrome, Ovarian Tumor, CA 125, Ascites, Hydrothorax, Senegal

1. Introduction

Demons-Meigs syndrome is defined as the presence of ascites with hydrothorax

in association with a benign ovarian [1]. This syndrome was first described in Mali in 1887 by Albert Demons-Meigs *et al*. [2], in 1937 they discovered identical cases and made relevant studies [2] [3]. It is very rare and its physiopathology remains obscure. The following criteria are to be met for the diagnosis of Meigs syndrome: 1) Presence of the benign tumor of the ovary—Fibroma, thecoma, granulosa cell tumor or Brenner tumor; 2) Ascites; 3) Pleural effusion; 4) A resolution of ascites and pleural effusion after removal of the tumor [2] [3] [4] [5] [6]. The most frequently reported tumors are fibroids and thecomas [7] [8]. Before the pathological examination of the surgical specimen, this rare syndrome often gives rise to fear of ovarian neoplasia due to the existence of ascites and the significant elevation of CA 125 [9]. In our context of Regional Hospital Center, we reporta clinical case of Demons-Meigs syndrome, which we took care of and followed for more than 6 months.

2. Patient and Observation

This is Mrs. MC.N, 36 years old 4gestes, 4pares, received for abdomino-pelvic pain, an abdominal mass evolving for 3 months, where the patient thought she was pregnant, when the menstruation appeared that ultrasound was performed objectifying an ascites and an ovarian mass. The medical-surgical history was unremarkable. Clinical examination found an abdomen increased in size (**Figure 1**) with a rounded mass extending from the right iliac fossa up to the umbilicus, mobile with a smooth outline. There is also a syndrome of thoracic effusion (pleurisy) and abdominal (ascites) of great abundance. On vaginal examination: clean vulva, cervix without particularity, normal vaginal wall. On rectal examination: free anal margin with normotonic sphincter, blister rectal containing stools of normal appearance. The ascites exploratory puncture

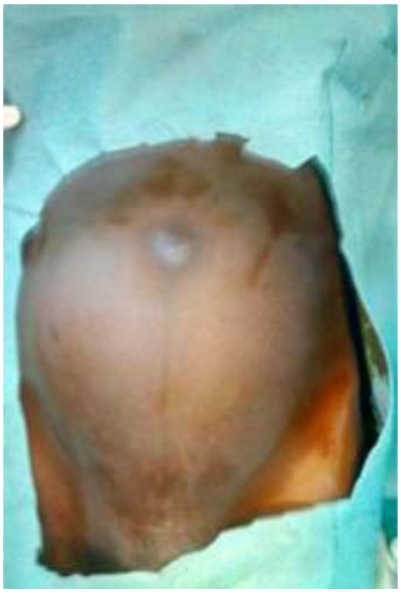

Figure 1. Appearance of the abdomen preoperatively.

bringing back a citrus-yellow liquid. In front of this case, an assessment was carried out, in particular the ultrasound which shows ascites of great abundance, and ahugeright ovarian mass of 304 × 158 mm. The uterus and the left ovary are normal in appearance. The cytology of the ascitic fluid shows an ascitic fluid devoid of malignant cells. An Abdomino-pelvic scanner shows a spontaneously homogeneous pelvic mass of 300 × 15 mm seems to develop in the depends on the right ovary: mucinous evolutionary process associated with ascites of great abundance (**Figure 2**). A frontal chest X-ray shows moderate pleural effusion (**Figure 3**). The CA 125 was 406.6 IU/ml. The hepatic, renal and cardiac tests were normal. Faced with this hugeovarian tumor associated with ascites and a high CA 125, the diagnosis of ovarian cancer is suspected. The patient underwent a preoperative assessment and a preanesthesia visit. Ovarian cancer surgery is planned with an exploratory laparotomy via the supra and subumbilical midline (**Figure 4**). This laparotomy revealed very abundant ascites, a huge solid tumor

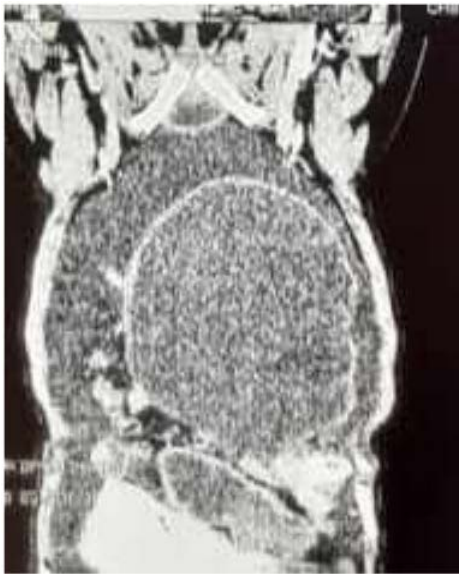

Figure 2. Abdominal computed tomography (CT).

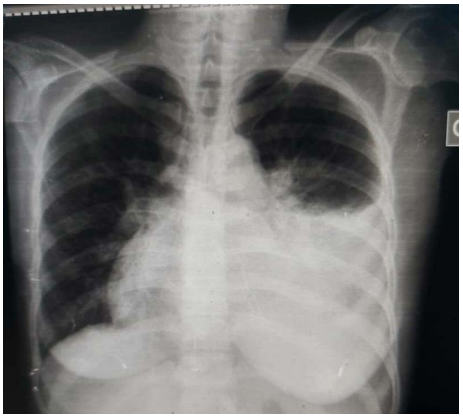

Figure 3. A chest X-ray showed hydrothorax.

developed at the expense of the right ovary, mobile whose limit is clear. The uterus and left adnexa were healthy. Procedures performed: Aspiration of 1200 ml of ascites fluid with a lemon yellow appearance, then right adnexectomy with preservation of the uterus left adnexa (**Figure 5** and **Figure 6**). Pathological analysis of the surgical specimen (**Figure 7** and **Figure 8**) was in favor of right ovarian fibrothecoma, no sign of malignancy (**Figure 9**). The patient was discharged on the eighth postoperative day. The postoperative period was simple, marked by the drying up of the effusions and the drop in CA 125 after 6 months of follow-upon an outpatient.

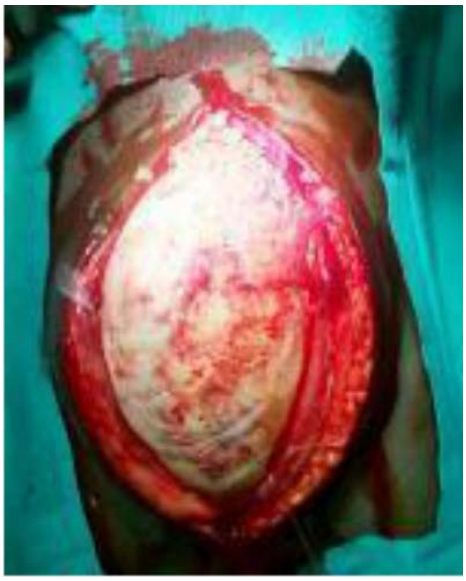

Figure 4. Appearance of the abdomen when opened.

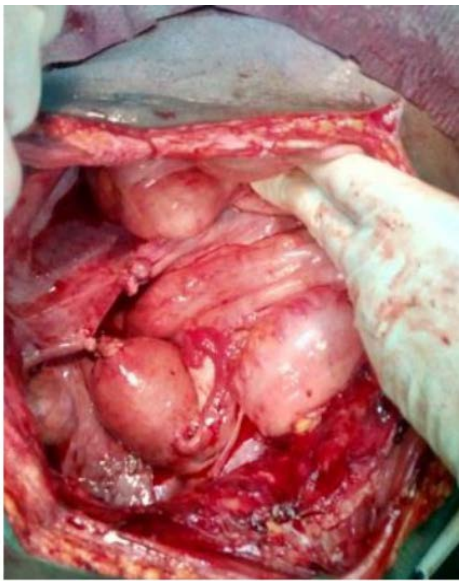

Figure 5. Right adnexectomy with preservation of the uterus and the left annex.

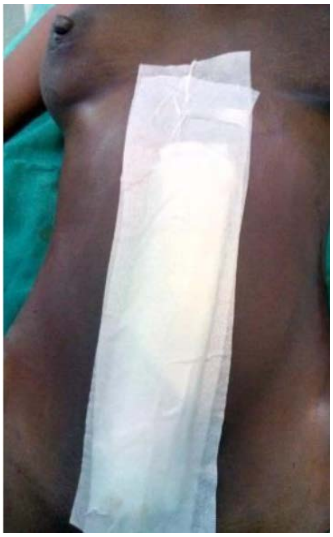

Figure 6. Postoperative appearance of the abdomen.

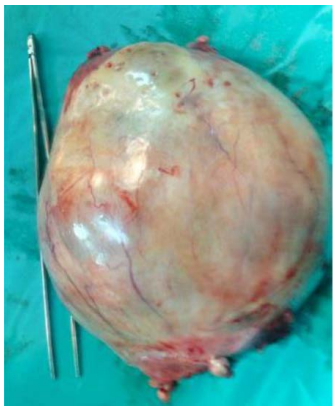

Figure 7. Macroscopic aspect of the operative part.

Figure 8. Macroscopic appearance after a longitudinal section of the operative part.

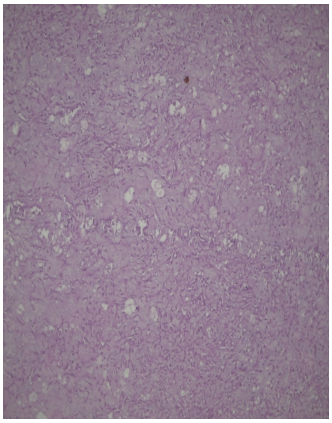

Figure 9. H & E staining, 100× magnification.

3. Discussion

Demons-Meigs syndrome is a rare anatomo-clinical entity that is seen in 0.25% of ovarian tumors; and which most often affects women in the pre- and post-menopausal period, between 40 and 50 years old [10]. A significant increase in CA 125 associated, peritoneal and pleural effusion and the presence of an ovarian tumor are more suspicious of ovarian neoplasia [7]. It groups together in its typical form the Funck-Brentano conditions [11] which are: Anatomo-clinical conditions with association of pleurisy, ascites and ovarian fibroma; progressive conditions with recurrence of the pleural effusion after puncture and drying of the latter after excision of the ovarian tumor; physicochemical conditions with identical pleural and ascitic fluid, in the form of a sero-fibrinous transudate without germs or neoplastic cells. Taking these conditions into account, our patient meets this definition. In fact, in our observation, the cytology of the ascitic fluid carried out in the diagnostic process revealed a fluid with the absence of malignant cells and no isolated germ. The resorption of the effusions after the tumor has been removed. The pathogenesis of Demons-Meigs syndrome remains unclear [5] [6] [7]. Several hypotheses have been put forward to explain the clinical triad of this syndrome, namely association of ascites, effusion thoraciqueand benign ovarian tumor. Among the hypotheses explaining the triad, we can cite the vascular theory (venous and lymphatic) developed in 1944 by Dockerty and Masson [12] which explains the appearance of ascites by a partial obstruction of the venous return, linked to a torsion of the pelvic tumor. There is then transudation of serous fluid through the capsule. For Meigs *et al.* [13], the genesis of ascites is explained by an increase in pressure in the intra-tumor lymphatics causing fluid to leak through the peritoneum. The passage of peritoneal fluid to the pleural cavity would follow transdiaphragmatic lymphatic pathways. Indeed in the association of an ovarian mass and a peritoneal effusion, always raises fears of a malignant process [14] [15], which led us in our therapeutic approach to carry out a cytology of the ascites fluid and of performing a wide midline laparotomy given the size of the tumor. In our observation, the CA 125 was

elevated preoperatively. The elevation of this tumor marker once again raises fears of a malignant origin. Indeed, CA 125 is high in more than 10% of benign tumors as has been proven in a series by Shen *et al*. [16]. The drop in the level of CA 125 postoperatively in our observation shows that this rate is linked to tumor size. our result on the evolution of the CA 125 level corroborates with the results of other observations on Demons-Meigs syndrome [17] [18] [19].

The age of our patient is 36 years old. This age is lower than most cases described in the literature, Boufettal *et al*. [20] in Tunisia, Mssrouri *et al*. In Morocco [21] and Mwansa *et al*. [17] in Congo who described cases occurring in patients aged 65 and 66, respectively, but is similar to that which was described in our country Senegal by Cissé *et al*. [22] who is 25 years old. A genetic predisposition could explain the appearance of the syndrome in young women in the Senegalese population. The course of our patient is marked by spontaneous drying after removal of the ovarian tumor. This spontaneous resorption is one of the criteria for this pathology although cases of recurrence have been described after a very long remission of 30 years [23]; which could lead to a bilateral adnexectomy in peri-menopausal patients.

4. Conclusion

Although rare, Demons-Meigs syndrome should be known to surgeons because it can mimic advanced ovarian neoplasia. The existence of ascites and/or pleural effusion does not necessarily mean malignancy in the presence of an ovarian tumor. Preoperative recognition of this syndrome is possible. It makes it possible to avoid carrying out heavy surgical procedures in the treatment of ovarian cancer and unnecessary in this benign pathology which has a good prognosis.

Contributions from the Authors

AIT, AM, patient care and follow-up, AM, AIT, AK, MB, ERP, IDD, MSD, BT, SK, AD are the major contributors to the writing and correction of the manuscript.

Conflicts of Interest

The authors declare no conflict of interest.

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