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Demons Meigs Syndrome: About Two Cases.

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ABSTRACT

Demons Meigs syndrome is the association of a benign ovarian tumor, a peritoneal and pleural effusion. Its physiopathology is still obscure till today. Through a rare case, and a review of the literature, we will discuss the diagnostic, etio-pathogenic, therapeutic, and evolutionary criteria of this syndrome.

Keywords

Demons Meigs syndrome, Fibroma, Fibrothecoma, Pleurisy, Ascites.

Introduction

Demons Meigs syndrome is a rare pathology, first described in 1937. It is a combination of three elements: an ovarian tumor, a pleural and peritoneal effusion. The ovarian tumor is most often a fibroid, a fibrothecoma, a granular cell tumor, or less often a Brenner tumor. The prevalence and incidence of this syndrome is not fully known. The treatment of Demons Meigs syndrome is surgical, with a good clinical course. Recurrences are extremely exceptional. We present two reports of patients with typical Demons Meigs syndrome, with disappearance of effusions after surgical removal of the ovarian tumor.

Observation 1

Mrs K.S, 28 years old, 2G 2P, who presented in consultation for chronic pelvic pain evolving for 8 months, with a notion of weight loss of 10 kg. The clinical examination had found an abdominal-pelvic mass reaching the umbilicus, independent of the uterus, and associated with significant ascites. Pelvic ultrasound revealed a well-limited cystic left latero uterine mass, with regular contours and without partitions or endocystic vegetations, measuring 7.3 x 6.3 cm (Figure 1). Pelvic CT showed a pelvic mass of probable left ovarian origin, heterogeneous, measuring 10 x 10.3 mm, extending over 9.8 mm and an associated pelvic effusion (Figure 2). The chest X-ray performed as part of the pre-anesthetic work-up revealed a left pleural fluid effusion of moderate size (Figure 3). The CA125 marker was elevated to 177U/ml for a normal of less than 35U/ml. A laparotomy was indicated, with exploration of a 16 cm solid cystic left latero uterine mass, at the expense of the left ovary, and a seroserosal effusion of moderate abundance. On the other hand, the contralateral ovary and the uterus did not present any macroscopic abnormalities. A lumpectomy and aspiration of the ascites fluid were performed. The postoperative course was simple. The patient was declared discharged at D3 postoperatively. The final histological result was in favor of an ovarian fibrothecoma and an inflammatory ascites fluid. At the time of the monitoring consultations, a significant regression was noted, then disappearance of the pelvic and peritoneal effusion over a total duration of one month and a half.

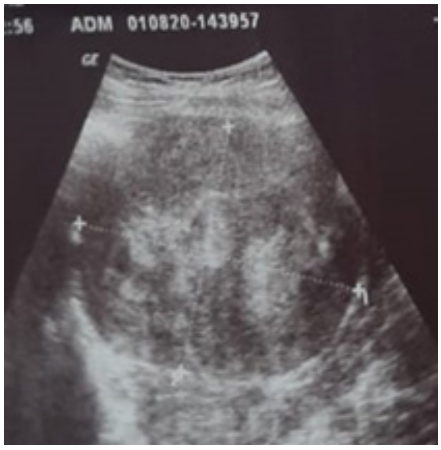

Figure 1: Left latero-uterine mass, measuring 7.3 x 6.3 cm.

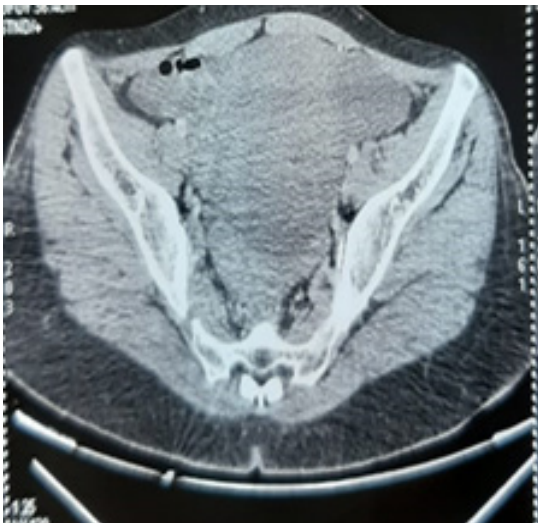

Figure 2 : Left latero-uterine ovarian mass measuring 10 x 10.3x9.8 mm

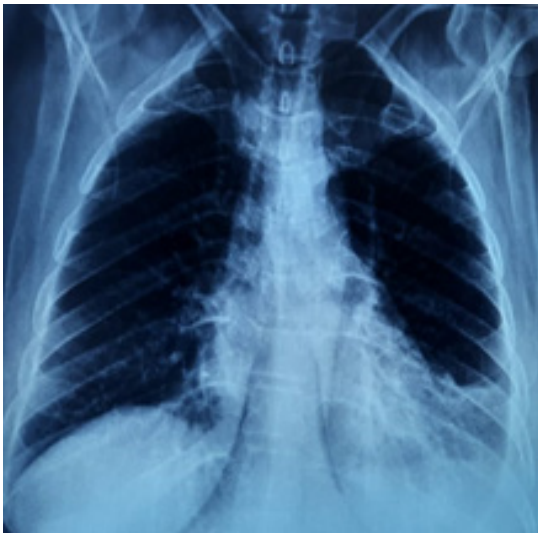

Figure 3: Left pleural effusion of moderate size.

Observation 2

Mrs N.E, 68 years old, 4G 4P, with a pathological history of arterial hypertension for 10 years under amlodipine 5mg, type 2 diabetes under insulin, who presented to our training for progressive abdominal distension associated with vomiting and chronic constipation. The clinical examination revealed a patient in good general condition, with a slight respiratory discomfort accentuated in the supine position, a slightly distended abdomen with a dullness of the flanks (Figure 4). Pelvic ultrasound revealed a well-limited cystic left latero uterine mass with thick walls in places, with echogenic content measuring 2.6x2.5 cm, associated with a pelvic effusion (Figure 5). Pelvic MRI showed a left latero uterine formation with a long axis of 21 cm, and a large peritoneal effusion (Figure 6). A chest X-ray showed a right pleural effusion (Figure 7). CA125 was elevated to 476 IU/ml for a normal of less than 35U/ml, CA19-9 was 5.5 and ACE was less than 1.7. Cervical and uterine smear revealed no abnormality.

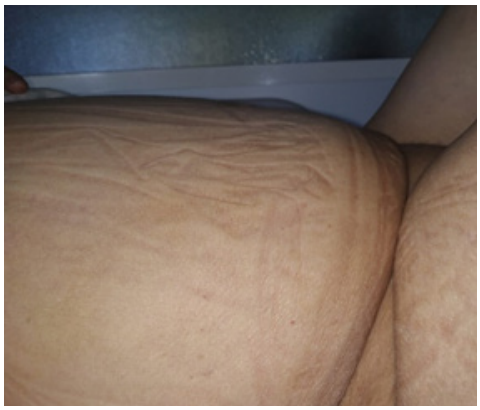

Figure 4 : Abdominal distension.

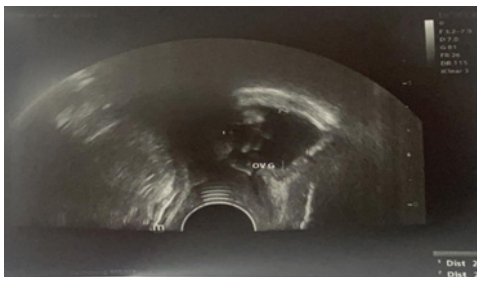

Figure 5 : Latero uterine mass mesuring 2,6x2,5 cm.

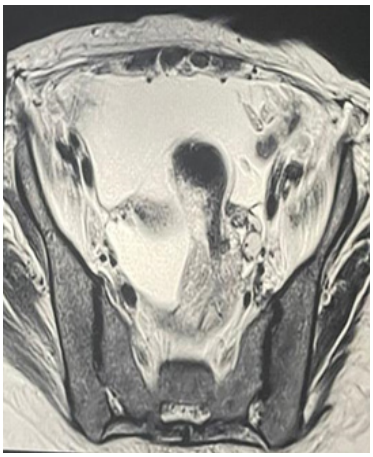

Figure 6: Left latero-uterine mass, measuring 21 mm, with abundant effusion.

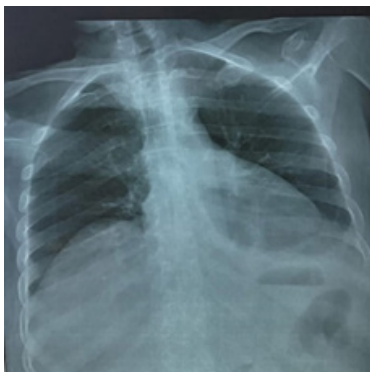

Figure 7: Right pleural effusion of moderate size.

Surgical exploration revealed a 3 cm solid cystic left latero uterine mass, at the expense of the left ovary, and a seroserosal effusion of medium abundance. On the other hand, the contralateral ovary and uterus were without abnormalities. A total hysterectomy with bilateral adnexectomy and aspiration of the ascites fluid were performed. The postoperative course was simple. The final histology was in favor of an ovarian fibrothecoma. The uterus, the contralateral adnexa, and the ascites fluid collection were free of malignant cells. At postoperative follow-up, there was a significant regression of pelvic and peritoneal effusion over a total duration of three months and disappearance of pleurisy.

Discussion

Demons Meigs syndrome is a rare disease first described in 1937 by gynecologist Joe Vincent Meigs and pulmonologist John Class in the Journal of Obstetrics and Gynecology. The name Demons associated with the syndrome goes back to Albert Jean Octave Demons in 1887, when he reported nine cases of ovarian tumor associated with ascites and pleural effusion.

Demons Meigs syndrome is characterized by the presence of pleural and peritoneal effusion in patients with ovarian fibroid or fibrothecoma. Ascites has been found in 10-15% of patients with fibromatous formations, while pleural effusion has been identified in 1%. The latter is frequently located on the right side in about 70%, while in 15% it is on the left side or bilateral [1]. Demons Meigs syndrome is more than a simple clinico-pathological triad. It is an anatomical, evolutionary and prognostic condition, making this syndrome a particular pathological entity. Our observation is an example of the typical Demons Meigs syndrome by the presence of a fibrothecoma, ascites and right pleural effusion.

The mean age of onset is approximately 50 years, with age limits between 40 and 60 years. A proportional increase with age has been observed [2]. Our two patients were respectively 28 and 68 years old.

Other benign or malignant tumors of the ovary (mature teratomas, mucinous cystadenoma, and serous cystadenoma) can also be associated with pleural and peritoneal effusion in the context of pseudo-Meigs syndrome [3]. On the other hand, cases of pseudo Meigs demons have been reported in prepuberty in which the benign tumor was represented by other tumor types (teratoma or cystadenoma) [1].

An elevation of CA 125 is classical in this syndrome. The literature suggests that this elevation is not an indicator of malignancy, but only in relation to the importance of the effusion and the size of the ovarian tumor [4-6].

The pathophysiology of pleural effusion can be explained as follows: The pleura is composed of two visceral and parietal layers leaving a virtual space: the pleural cavity. There is a process of filtration of liquid; which depends on the hydrostatic balance and osmotic pressures; from the capillary vessels in the subpleural space, and given to the pleural cavity. And therefore the presence of a minimal amount of fluid in the virtual cavity is physiological. The pleural effusion in cases of ovarian tumors, corresponds to an exudate. The fluid moves from the peritoneal cavity to the pleural cavity through diaphragmatic defects or lymphatic channels. The effusion is usually located on the right side and may be massive on occasion, with characteristic biochemical or cellular effects not specific to the fluid [7].

The etiology of ascites has been explained by several mechanisms:

- Partial torsion of the ovarian vascular pedicle leading to venous engorgement and transudation, which enters the pleural cavity through the diaphragmatic lymphatics or by defects of the diaphragm which are more common on the right.
- The exudation of the peritoneum as a result of tumor degeneration or changes in its capsular veins or by its secretion [7].
- Vascular endothelial growth factor (VEGF), which increases capillary permeability, would also be associated with pleural and peritoneal fluid formation. Ishiko et al. [9] reported a significant difference between VEGF levels in pleura and peritoneal fluid before and after tumor removal in patients with Meigs or pseudo-Meigs syndrome. The size of the pleural effusion is largely independent of the amount of ascites. The association between the pelvic tumor and ascites is confirmed by the rapid resolution of fluid after tumor removal.

The treatment of Demons-Meigs syndrome is surgical, by exploratory laparotomy. It corresponds to the removal of the ovarian tumor. If the patient is in the prepubertal period, a lumpectomy, a partial oophorectomy or a unilateral salpingectomy will be proposed. In women of childbearing age, surgical treatment will be a unilateral adnexectomy. For postmenopausal patients, treatment may include total hysterectomy with bilateral adnexectomy [10]. The contralateral ovary is healthy in one third of cases [11].

Demons-Meigs syndrome involves remission of ascites and pleural fluid after surgical removal of the tumor. The rapid reconstitution of effusions is always observed after iterative punctures in the absence of surgery. This fact was noted and established by Albert Jean Octave Demons in 1903. The case of our patients is similar to those reported in the literature, with a complete disappearance of ascites and hydrothorax postoperatively. The cure rate for Demons Meigs syndrome is high, and the life expectancy after surgery is the same as that of the general population [12]. Recurrence is extremely rare. Only one case of recurrence of Demons Meigs syndrome after initial treatment has been described to date by Bretelle et al. in a 30-year-old woman. Surgical removal of a benign fibrothecoma led to a second time to a disappearance of the associated effusions [2].

Genital tuberculosis poses a diagnostic problem in the presence of peritoneal and pleural effusion, in the absence of an ovarian tumor. It is worth looking for in our context.

Conclusion

Demons-Meigs syndrome is defined by the association of an ovarian tumor, pleural effusion and ascites. The ovarian tumor is benign, and is usually an ovarian fibroid or fibrothecoma. The CA 125 tumor marker has elevated values initially similar to ovarian cancers, but the values return to normal after surgical treatment. After surgical removal of the ovarian tumor, remission of ascites and pleural effusion is the rule. Although this syndrome has been described for more than 100 years, it poses diagnostic problems, with its unusual character and the involvement of several factors, including: molecular, hormonal, genetic and mechanical in its etiopathogenic mechanism. The only available treatment is surgical removal. The remission of ascites and pleural effusion is the rule.

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