Journal Homepage: - www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/20520
DOI URL: http://dx.doi.org/10.21474/IJAR01/20520

RESEARCH ARTICLE

A CHALLENGING CASE OF VOLUMINOUS OVARIAN FIBROTHECOMA MIMICKING MALIGNANCY: DEMONS-MEIGS SYNDROME

K. Laaouini, W. Garrouani, M. Bendahou Idrissi, N. Mamouni, S. Errarhay, C. Bouchikhi and A. Banani
Hassan II University Hospital of Fez, Department of Gynecology and Obstetrics 1.

Manuscript Info

Manuscript History
Received: 28 December 2024
Final Accepted: 31 January 2025
Published: February 2025

Abstract

Demons-Meigs syndrome is a rare clinical entity characterized by a benign ovarian tumor associated with ascites and pleural effusion, which resolve after tumor removal. We report the case of a 68-year-old postmenopausal woman diagnosed with a large ovarian fibrothecoma, initially suspected of malignancy due to massive ascites and an elevated CA-125 level. The final histopathological diagnosis confirmed a benign fibrothecoma. Surgical treatment led to a complete resolution of symptoms, supporting the diagnosis of Demons-Meigs syndrome. This case highlights the challenges in distinguishing fibrothecoma from malignant ovarian tumors and underscores the importance of histopathological confirmation.

Introduction:-

Ovarian fibrothecomas are rare sex cord-stromal tumors that account for approximately 4% of all ovarian neoplasms (Young & Scully, 2002). These tumors can occur at any age but are most commonly seen in postmenopausal women. While most fibrothecomas are asymptomatic, they can be associated with Demons-Meigs syndrome, a condition characterized by a benign ovarian tumor, ascites, and pleural effusion (Meigs & Cass, 1937).

The presence of ascites and an elevated CA-125 level often raises suspicion for ovarian carcinoma, making differentiation between benign and malignant ovarian tumors challenging (Brown et al., 2021). Imaging modalities such as ultrasound, CT, and MRI play a crucial role in tumor characterization, but final diagnosis relies on histopathological analysis (Jung et al., 2005). Here, we present a case of voluminous ovarian fibrothecoma associated with Demons-Meigs syndrome, emphasizing the importance of accurate diagnosis and appropriate surgical management.

Case Report

A 68-year-old postmenopausal woman, a grand multipara, was referred for the management of endometrial thickening detected during an etiological workup for ascites. Her medical history included a daughter who had died from gastric cancer.

On examination, the patient had a distended abdomen with diffuse dullness, suggesting significant ascites. No palpable mass was identified, and gynecological examination was unremarkable.

Corresponding Author:- K. Laaouini
Address:- Hassan II University Hospital of Fez, Department of Gynecology and Obstetrics 1.

Pelvic ultrasound revealed a multilobulated, heterogeneous, solid ovarian mass associated with massive ascites. The endometrial thickness was measured at 2 cm, with no Doppler signal.

A CT scan of the abdomen and pelvis showed a large amount of nodular ascites and a heterogeneous, solid ovarian mass measuring 13 × 7 cm, freely mobile in the left flank. The uterus was enlarged, with an endometrial hypertrophy of 23 mm. The patient did not attend her scheduled thoracic CT scan.

Serum CA-125 was markedly elevated at 934.4 U/mL, raising concerns about a possible malignant ovarian neoplasm.

An emergency hysteroscopy revealed intrauterine adhesions with a vascularized endometrial polyp, necessitating hysterosurgical intervention. Given the suspicion of an ovarian tumor, an exploratory laparotomy was performed.

Intraoperatively, a firm, smooth, left ovarian tumor was identified, without exophytic growths or peritoneal implants. The right ovary appeared normal, and no evidence of peritoneal carcinomatosis or liver metastasis was found. About 1500 mL of serous ascitic fluid was evacuated and sent for cytological analysis. A left oophorectomy was performed.

Frozen section analysis suggested a benign ovarian fibrothecoma. Multiple peritoneal biopsies and cytological analysis of the ascitic fluid were performed.

Definitive histopathological examination confirmed fibrothecoma, characterized by spindle-shaped fibroblastic cells interspersed with theca-like cells, with no signs of malignancy. The peritoneal fluid cytology was negative for malignant cells.

The postoperative period was uneventful, and the patient was discharged on postoperative day 3. Remarkably, the ascites completely regressed within one month. With a two-year follow-up, no recurrence of ascites or tumor was observed, confirming the diagnosis of Demons-Meigs syndrome.

Discussion:-

Ovarian fibrothecomas are benign sex cord-stromal tumors composed of fibrous and theca-like cells. They are often found in postmenopausal women and can sometimes produce estrogen, leading to endometrial hyperplasia or abnormal uterine bleeding (Young & Scully, 2002).

Demons-Meigs syndrome, first described by Meigs and Cass (1937), is characterized by the triad of a benign ovarian tumor, ascites, and pleural effusion. This condition resolves spontaneously after tumor removal, distinguishing it from malignant ovarian tumors (Meigs & Cass, 1937; Alessi et al., 2018).

Mechanism of Ascites Formation:

The exact pathophysiology of ascites formation in fibrothecoma remains unclear but is thought to involve:

1. Increased peritoneal permeability, leading to fluid accumulation.
2. Lymphatic obstruction due to the tumor's mechanical effect.
3. Irritation of the peritoneum, triggering fluid exudation (Jung et al., 2005).

Differential Diagnosis and Diagnostic Challenges:

This case presented diagnostic challenges due to:

1. Elevated CA-125 levels: Typically associated with ovarian malignancy, but also elevated in benign conditions like Meigs syndrome, endometriosis, and peritoneal irritation (Brown et al., 2021).
2. Endometrial thickening: The enlarged uterus (23 mm endometrial thickness) raised suspicion for estrogen-producing tumors or endometrial carcinoma, necessitating further evaluation.
3. Radiological findings: The heterogeneous ovarian mass with ascites initially suggested a malignant etiology, but final diagnosis relied on histopathology (Outwater & Dunton, 2012).

Management Approach:

1. Surgical removal of the tumor is the primary treatment. In Demons-Meigs syndrome, ascites and pleural effusion resolve spontaneously after tumor excision.

2. Histopathological confirmation is essential to rule out malignancy.
3. Long-term follow-up is crucial to monitor recurrence and ensure complete resolution of symptoms.

Figure 1:- Abdominopelvic CT scan showing the presence of a large abdominopelvic tumor with thickened walls and multiple septations, associated with ascites.

Conclusion:-

This case highlights the diagnostic complexity of ovarian fibrothecomas when associated with ascites and an elevated CA-125 level. Demons-Meigs syndrome, though rare, should be considered in postmenopausal women presenting with a solid ovarian mass and ascites. Surgical removal provides both diagnostic and curative benefits, with excellent long-term outcomes. Awareness of this entity can help avoid unnecessary radical interventions.

Références:-

1. Meigs JV, Cass JW. Fibroma of the ovary with ascites and hydrothorax. Am J Obstet Gynecol. 1937;33:249-267.
2. Young RH, Scully RE. Ovarian sex cord-stromal tumors. In: Kurman RJ, ed. Blaustein's Pathology of the Female Genital Tract. Springer, 2002.
3. Brown J, Peres LC, Schildkraut JM. Epidemiology and risk factors for ovarian tumors in postmenopausal women. J Clin Oncol. 2021;39(4):347-355.
4. Jung SE, Lee JM, Rha SE, et al. CT and MRI findings of sex cord-stromal tumor of the ovary. AJR Am J Roentgenol. 2005;185:207-215.
5. Outwater EK, Dunton C. Imaging of the ovary and adnexa: Clinical issues and applications of MR imaging. Radiology. 2012;264(3):647-664.
6. Alessi JV, Templeman C, Kornblith A. Ovarian sex cord-stromal tumors: Current treatment strategies. Gynecol Oncol. 2018;149:3-10.