

Meigs Syndrome: A Comprehensive Review of Clinical Presentation, Diagnosis, and Management

Marco Antonio Hernández Mendoza¹, Zaida Fernanda Zarazúa Sánchez², Juan Eduardo Galván Carrasco¹, Luis Enrique Morales Montoya³, Onassis Armando Prado Hernández⁴, Marlo Ramón Narváez Paredes⁵

¹Universidad Autónoma de Guadalajara, Guadalajara, Jalisco. México.

²Universidad del Valle de México, Campus Queretaro, Queretaro. México

³Universidad Autónoma de Sinaloa, Culiacán, Sinaloa. México

⁴Universidad de Guadalajara, Centro Universitario de Ciencias de la Salud, Guadalajara, Jalisco. México

⁵Universidad Autónoma de Tlaxcala, Tlaxcala. México

ABSTRACT

Meigs Syndrome is a rare clinical entity characterized by the triad of benign ovarian tumor, ascites, and pleural effusion. This syndrome, often masquerading as malignant disease, presents a diagnostic challenge for healthcare professionals. In this comprehensive review, we aim to provide a detailed analysis of the clinical manifestations, diagnostic modalities, and management strategies of Meigs Syndrome. We explore the epidemiology of the syndrome, emphasizing its rarity, and discuss the various etiological theories that underlie its pathophysiology. Differential diagnoses, including malignant conditions that mimic Meigs Syndrome, are examined to guide accurate and timely diagnosis. An in-depth analysis of imaging techniques, such as ultrasound, computed tomography, and magnetic resonance imaging, is included, along with the significance of tumor markers and cytology in diagnosis. The cornerstone of treatment primarily involves surgical intervention, and we delve into the various surgical approaches and outcomes. Additionally, we shed light on the postoperative course and long-term prognosis of patients with Meigs Syndrome. This review aims to equip clinicians, radiologists, and surgeons with a comprehensive understanding of this uncommon condition to facilitate prompt recognition and effective management.

KEYWORDS: Meigs, syndrome, ovarian, tumor.

ARTICLE DETAILS

Published On:
25 October 2023

Available on:
<https://ijmscr.org/>

INTRODUCTION

Meigs Syndrome, a clinical syndrome first described by Joe Vincent Meigs in 1937, stands as a rare but intriguing condition in the realm of gynecological and surgical pathology. This syndrome is characterized by the classic triad of a benign ovarian tumor, ascites, and pleural effusion, and its clinical presentation often closely mimics malignancy, posing a significant diagnostic challenge. Over the decades, our understanding of Meigs Syndrome has evolved, and this enigmatic syndrome has continued to captivate the medical community. As such, there remains a need for a comprehensive review of this syndrome to encompass the latest developments in diagnosis and management.^{1,2}

The epidemiology of Meigs Syndrome is characterized by its rarity, with only a small fraction of ovarian tumors manifesting in this distinct manner. Although the precise

etiology of Meigs Syndrome remains a subject of debate, it is essential to explore the various theories that have been proposed, including hormonal, inflammatory, and vascular factors. This article seeks to delve into the available evidence surrounding the pathophysiological basis of Meigs Syndrome.^{1,2}

The diagnostic workup of Meigs Syndrome is multifaceted, involving a combination of clinical evaluation, imaging studies, and laboratory investigations. This review will comprehensively assess the differential diagnosis of Meigs Syndrome, taking into account other malignant and benign conditions that may present with similar clinical features. We will emphasize the importance of advanced imaging modalities, such as ultrasound, computed tomography, and magnetic resonance imaging, in achieving a precise

Meigs Syndrome: A Comprehensive Review of Clinical Presentation, Diagnosis, and Management

diagnosis. Furthermore, the role of tumor markers and cytology in confirming the diagnosis will be explored.^{3,4} Surgical intervention is the mainstay of treatment for Meigs Syndrome, and the choice of surgical approach may vary depending on the clinical context. This article will provide a detailed examination of the surgical techniques employed in the management of Meigs Syndrome and discuss the postoperative course and long-term outcomes. Ultimately, the objective of this comprehensive review is to equip healthcare professionals with the knowledge and tools necessary to recognize and effectively manage this rare but diagnostically challenging syndrome.^{3,4}

EPIDEMIOLOGY

Meigs Syndrome, an exceptionally uncommon clinical entity, is characterized by its distinctive triad of benign ovarian tumor, ascites, and pleural effusion. To comprehensively understand the epidemiological aspects of Meigs Syndrome, it is vital to explore its rarity, demographic distribution, and associated risk factors.⁵

Incidence and Prevalence:

Meigs Syndrome is a rare clinical condition, accounting for a minute fraction of ovarian tumors. Its precise incidence and prevalence remain challenging to ascertain due to its infrequency. Available literature suggests that Meigs Syndrome is an exceedingly rare entity, with sporadic cases reported worldwide. Most instances are documented in the context of case reports or small case series. This scarcity underscores the need for heightened awareness among healthcare providers to facilitate early recognition.⁵

Age and Gender Distribution:

Meigs Syndrome predominantly affects women, as it is intrinsically linked to ovarian pathology. It is most commonly observed in women between the ages of 30 and 60, aligning with the age range during which ovarian tumors are more prevalent. Nevertheless, Meigs Syndrome has been reported in both premenopausal and postmenopausal individuals, emphasizing its potential occurrence across a broad age spectrum.⁵

Demographic Variation:

Epidemiological data on Meigs Syndrome reveal no specific geographic or racial predilection. Cases have been reported in various regions worldwide, without significant variations based on geography or ethnicity. This suggests that the syndrome may have a universal distribution and is not confined to specific populations.⁵

Risk Factors:

The etiological factors contributing to the development of Meigs Syndrome remain elusive. However, there are some associations between the syndrome and specific risk factors. Notably, Meigs Syndrome is most commonly associated with benign ovarian tumors, with fibromas and thecomas being the primary culprits. The hormonal milieu and genetic

predispositions may play a role in the development of these tumors, but further research is needed to elucidate these potential risk factors.⁵

Differential Diagnosis Challenges:

A significant epidemiological aspect of Meigs Syndrome lies in the diagnostic challenge it poses. Due to its clinical presentation mimicking malignant disease, patients often undergo extensive evaluations, including biopsies and invasive procedures. This underscores the importance of prompt and accurate diagnosis to avoid unnecessary medical interventions and psychological distress for patients.⁵

Evolving Understanding:

As our knowledge of ovarian tumors and their clinical manifestations continues to evolve, the epidemiological landscape of Meigs Syndrome may undergo changes. Advancements in imaging techniques and diagnostic modalities, coupled with increased awareness among healthcare professionals, may lead to more accurate and timely diagnoses in the future.⁵

In conclusion, Meigs Syndrome is an exceptionally rare clinical entity with a global presence but no specific geographic or racial predilection. It primarily affects women of a wide age range and is most commonly associated with benign ovarian tumors. The rarity of this syndrome underscores the need for healthcare providers to maintain a high index of suspicion, especially when encountering cases of ascites and pleural effusion in the context of an ovarian mass. Further research is warranted to explore potential risk factors and refine the epidemiological understanding of Meigs Syndrome.⁵

Clinical Manifestations of Meigs Syndrome

Meigs Syndrome, a rare gynecological condition, is defined by a characteristic triad of clinical manifestations: a benign ovarian tumor, ascites, and pleural effusion. These clinical features can closely mimic malignant diseases, necessitating a thorough understanding of the syndrome's clinical presentation for timely diagnosis and management.⁶

Benign Ovarian Tumor:

Tumor Types: The ovarian tumor associated with Meigs Syndrome is most frequently a fibroma or a thecoma. These tumors are typically slow-growing and can attain a considerable size.

Pelvic Pain: Patients may present with pelvic discomfort, which can range from mild to severe, due to the expansive nature of the ovarian mass.⁶

Pelvic Mass: On physical examination, a palpable, firm, and often irregular pelvic mass may be identified. Imaging studies further confirm the presence of the tumor.

Ovarian Torsion: In some cases, the tumor's size may lead to ovarian torsion, causing acute pelvic pain and requiring urgent surgical intervention.⁶

Ascites:

Meigs Syndrome: A Comprehensive Review of Clinical Presentation, Diagnosis, and Management

Accumulation of Fluid: Ascites, characterized by the accumulation of serous fluid within the peritoneal cavity, is a hallmark feature of Meigs Syndrome.

Abdominal Distension: The presence of ascites results in progressive abdominal distension, which can become marked as the volume of fluid increases.⁶

Dyspnea: Severe ascites can compress the diaphragm, leading to dyspnea (shortness of breath) and difficulty in breathing.⁶

Shift in Body Weight: Patients may experience a significant shift in body weight due to fluid retention, which can lead to edema in the lower extremities.⁶

Pleural Effusion:

Fluid Accumulation in the Pleural Cavity: Pleural effusion, characterized by the accumulation of serous fluid in the pleural cavity surrounding the lungs, is another distinctive feature.⁶

Respiratory Symptoms: Patients may report respiratory symptoms, including dyspnea, cough, and chest discomfort, as a result of the pleural effusion's compressive effect on the lungs.

Decreased Breath Sounds: Physical examination may reveal decreased breath sounds on the affected side due to reduced lung expansion.⁶

Clinical Mimicry of Malignancy:

Diagnostic Challenge: The clinical manifestations of Meigs Syndrome closely resemble those of ovarian malignancies, such as ovarian carcinoma or metastatic disease. Therefore, this syndrome poses a diagnostic challenge, necessitating a rigorous evaluation to differentiate it from malignant conditions.⁶

Serum Tumor Markers: Tumor markers like CA-125 may be elevated, further complicating the diagnostic process by mimicking the tumor marker profiles seen in ovarian cancer.⁶

Resolution Post-Tumor Removal:

Distinctive Feature: One of the distinctive clinical features of Meigs Syndrome is the rapid resolution of ascites and pleural effusion following the removal of the benign ovarian tumor.

Postoperative Improvement: Patients experience a notable improvement in their clinical condition after surgical resection of the tumor, highlighting the unique pathophysiological mechanisms at play.⁶

Meigs Syndrome presents with a characteristic triad of clinical manifestations, including a benign ovarian tumor, ascites, and pleural effusion. Accurate recognition of these features, alongside differentiation from malignant conditions, is crucial for effective diagnosis and the initiation of appropriate management strategies. Understanding the clinical presentation of Meigs Syndrome is pivotal in ensuring timely intervention and improved outcomes for affected individuals.⁶

DIAGNOSIS

The diagnosis of Meigs Syndrome is a complex process that requires careful evaluation to differentiate it from malignant

conditions presenting with similar clinical features. This section provides an in-depth exploration of the diagnostic modalities, clinical findings, and laboratory tests that play a pivotal role in establishing a precise diagnosis of Meigs Syndrome.⁷

Clinical Evaluation:

Medical History: A detailed medical history is paramount in the diagnostic process. Inquiry into the onset and progression of symptoms, including abdominal discomfort, dyspnea, and pleuritic chest pain, is essential. The duration of these symptoms and any changes in body weight or gynecological history must be ascertained.⁷

Physical Examination: Physical examination can reveal crucial clinical signs, such as the presence of a palpable pelvic mass, abdominal distension, shifting dullness indicative of ascites, and decreased breath sounds on the affected side in the case of pleural effusion.⁷

Imaging Modalities:

Ultrasonography: Transvaginal or abdominal ultrasonography is a primary imaging modality for the assessment of the pelvic mass, ascites, and pleural effusion. It provides valuable information regarding the size, location, and characteristics of the ovarian tumor.⁷

Computed Tomography (CT) Scan: CT imaging offers a more comprehensive visualization of the pelvic mass and the extent of ascites and pleural effusion. It can aid in assessing any lymph node involvement and is particularly helpful in cases where ultrasound findings are inconclusive.

Magnetic Resonance Imaging (MRI): MRI provides detailed images of the pelvic mass, helping to differentiate between benign and malignant tumors. It can also identify potential torsion of the ovarian mass.⁷

Tumor Markers and Cytology:

CA-125: Serum CA-125 levels may be elevated in Meigs Syndrome, which can add to the diagnostic complexity, as this marker is commonly associated with ovarian malignancies. However, CA-125 is not specific to cancer, and its elevation in Meigs Syndrome should be interpreted cautiously.⁷

Cytological Analysis: In cases with pleural effusion, thoracentesis can be performed to obtain pleural fluid for cytological analysis. Cytology may reveal no malignant cells, further supporting the benign nature of the effusion.⁷

Biopsy:

Tumor Biopsy: While not typically recommended, in some challenging cases, a biopsy of the ovarian mass may be considered. However, it should be approached with caution, as it can lead to complications such as tumor rupture and spillage.⁷

Differential Diagnosis:

Malignant Conditions: It is imperative to rule out ovarian malignancies, particularly ovarian carcinoma, through the utilization of imaging, tumor markers, and, when necessary, biopsy.

Meigs Syndrome: A Comprehensive Review of Clinical Presentation, Diagnosis, and Management

Other Benign Conditions: The diagnostic process should also consider other benign conditions, such as uterine fibroids or benign ovarian cysts, which can mimic Meigs Syndrome to a certain extent.⁷

Resolution Post-Surgery:

Diagnostic Criterion: A fundamental diagnostic criterion for Meigs Syndrome is the rapid resolution of ascites and pleural effusion following the surgical removal of the benign ovarian tumor. This clinical observation, also known as "pseudo-Meigs Syndrome," serves as a confirmatory diagnostic feature, distinguishing it from true Meigs Syndrome.⁷

In summary, diagnosing Meigs Syndrome involves a multidisciplinary approach, incorporating clinical evaluation, imaging modalities, laboratory tests, and careful consideration of differential diagnoses. The diagnostic journey is further complicated by the potential elevation of serum tumor markers, necessitating a cautious interpretation of these results. Ultimately, the resolution of ascites and pleural effusion after tumor resection serves as a definitive diagnostic criterion, providing essential guidance for healthcare professionals in confirming the presence of Meigs Syndrome and initiating the appropriate course of treatment.⁷

TREATMENT

The management of Meigs Syndrome is primarily surgical, with the goal of addressing the benign ovarian tumor and its associated manifestations, including ascites and pleural effusion. This section provides an in-depth exploration of the treatment modalities, surgical techniques, and postoperative care involved in managing Meigs Syndrome.⁸

Surgical Intervention:

Oophorectomy: The cornerstone of treatment for Meigs Syndrome is the surgical removal of the affected ovary containing the benign tumor. This procedure is typically accomplished through oophorectomy, which can be performed via laparoscopy or laparotomy, depending on the patient's clinical condition and the surgeon's preference.⁸

Laparoscopy vs. Laparotomy:

Laparoscopy: Minimally invasive laparoscopic surgery offers advantages such as smaller incisions, reduced postoperative pain, shorter hospital stays, and faster recovery. Laparoscopy is preferred when the tumor can be safely excised using this approach.⁸

Laparotomy: In cases of larger or complex tumors, or when ovarian torsion is present, an open laparotomy may be necessary to ensure complete tumor removal and assess the extent of the disease.⁸

Intraoperative Evaluation:

Peritoneal Washing: During surgery, peritoneal washing is often performed to obtain peritoneal fluid for cytological analysis. This can help rule out malignant cells and provide further confirmation of the benign nature of the ovarian tumor.⁸

Biopsy Consideration: In rare and challenging cases, intraoperative biopsy of the tumor may be considered. However, this should be approached with caution to avoid potential complications.

Postoperative Care:

Immediate Relief: Following surgery, patients experience rapid relief from ascites and pleural effusion. This clinical observation serves as a confirmatory diagnostic feature and is a reassuring aspect of postoperative care.⁸

Hospital Stay: The length of hospitalization depends on the surgical approach, overall patient condition, and any complications that may arise. Most patients can expect a relatively short hospital stay, especially when a laparoscopic approach is employed.⁸

Pain Management: Postoperative pain management is essential, and patients are typically prescribed pain medications to alleviate discomfort.⁸

Follow-Up: Regular follow-up appointments are crucial to monitor the patient's progress and ensure there is no recurrence of Meigs Syndrome or related complications.⁸

Long-Term Prognosis:

Favorable Prognosis: Meigs Syndrome carries a favorable long-term prognosis, with most patients experiencing a complete resolution of symptoms and no recurrence of ascites or pleural effusion following tumor removal.⁹

Ovarian Function: In premenopausal patients, the preservation of ovarian function is a significant consideration. Surgeons aim to spare the unaffected ovary when feasible to maintain hormonal balance and fertility.⁹

Reproductive Considerations: Meigs Syndrome does not typically impact future fertility, and patients can conceive naturally after recovery.⁹

Consideration of Pseudo-Meigs Syndrome:

Pseudo-Meigs Syndrome: In cases where ascites and pleural effusion persist after ovarian tumor removal, a subset of patients may develop "pseudo-Meigs Syndrome." This condition necessitates further evaluation to identify potential underlying causes, such as adhesions or peritoneal irritation, which may require additional treatment.^{9,10}

The treatment of Meigs Syndrome revolves around the surgical resection of the benign ovarian tumor, addressing ascites, and resolving pleural effusion. The choice of surgical approach, whether laparoscopic or open, is determined by the clinical presentation and tumor characteristics. Postoperative care focuses on pain management and monitoring for any complications. The favorable long-term prognosis of Meigs Syndrome, along with its minimal impact on ovarian function and fertility, underscores the importance of timely diagnosis and surgical intervention to ensure optimal patient outcomes.^{11,12}

CONCLUSION

Meigs Syndrome, a rare and intriguing gynecological condition, characterized by the triad of a benign ovarian

Meigs Syndrome: A Comprehensive Review of Clinical Presentation, Diagnosis, and Management

tumor, ascites, and pleural effusion, has long captivated the medical community. In this comprehensive exploration of Meigs Syndrome, we have delved into its epidemiology, clinical manifestations, diagnostic challenges, and treatment modalities. The intricate interplay of these elements provides a fascinating tapestry of understanding surrounding this unique syndrome.

Epidemiologically, Meigs Syndrome stands as a rarity in the realm of gynecological disorders, with a limited number of reported cases worldwide. It exhibits a broad demographic distribution, affecting women across various age groups and ethnic backgrounds. Though not confined to specific populations, its predilection for benign ovarian tumors, such as fibromas and thecomas, is a consistent theme.

The clinical presentation of Meigs Syndrome is characterized by a perplexing mimicry of malignant disease. The triad of a benign ovarian tumor, ascites, and pleural effusion can lead to diagnostic challenges, as the syndrome closely resembles ovarian malignancies. This diagnostic complexity is compounded by the potential elevation of serum tumor markers, particularly CA-125, which is commonly associated with ovarian cancer. Differentiating Meigs Syndrome from malignancy is a critical task, requiring a careful evaluation of clinical findings and diagnostic test results.

The cornerstone of Meigs Syndrome management is surgical intervention. Oophorectomy, achieved through laparoscopy or laparotomy, forms the crux of the treatment strategy, allowing for the complete removal of the affected ovarian mass. In most cases, the resolution of ascites and pleural effusion post-surgery serves as a diagnostic confirmation and provides immediate relief to patients. Long-term prognosis is generally favorable, with minimal impact on ovarian function and fertility, especially in premenopausal women.

It is important to recognize that some cases may present the challenging phenomenon of "pseudo-Meigs Syndrome," wherein the resolution of ascites and pleural effusion is not achieved post-tumor removal. In such instances, further evaluation and intervention may be necessary to identify and address underlying causes.

In conclusion, Meigs Syndrome, with its triad of benign ovarian tumor, ascites, and pleural effusion, continues to enthrall clinicians, radiologists, and surgeons. Its rarity, mimicry of malignancy, and the diagnostic complexities it presents make it a diagnostic puzzle to be unraveled. Early recognition and accurate diagnosis are pivotal for the successful management of this condition. Surgical removal of the ovarian tumor, whether by laparoscopy or laparotomy, remains the definitive treatment modality, with the promise of a favorable long-term prognosis. As our understanding of Meigs Syndrome evolves, ongoing research and clinical experience will further illuminate this intriguing clinical entity, ensuring improved outcomes and a brighter future for those affected by this rare syndrome.

REFERENCES

- I. Meigs JV. Fibroma of the ovary with ascites and hydrotorax.. *Am J Obstet Gynecol*, 33 (1937), pp. 249-67
- II. Meigs JV. Fibroma of the ovary with ascites and hydrotorax. Meigs' syndrome.. *Am J Obstet Gynecol*, 67 (1954), pp. 962-87
- III. Lurie S. Meigs' syndrome: the history of the eponym. *Eur J Obstet Gynecol Reprod Biol*, 92 (2000), pp. 199-204
- IV. Latta RJ, Lee PD. Meigs' syndrome in a young woman. *J Adolesc Health Care*, 1 (1981), pp. 313-5
- V. Liu TH, Yan SS, Lee YN. Meigs' syndrome: a case report.. *Zhonghua Yi Xue Za Zhi*, 44 (1989), pp. 322-4
- VI. Bierman SM, Reuter KL, Hunter RE. Meigs' syndrome and ovarian fibroma: CT findings.. *J Comput Assist Tomogr*, 14 (1990), pp. 833-4
- VII. Walker JL, Manetta A, Mannel RS, Liao SY. Cellular fibroma masquerading as ovarian carcinoma. *Obstet Gynecol*, 76 (1990), pp. 530-1
- VIII. Le Bouedec G, Glowaczower E, De Latour M, Fondrinier E, Dauplat J. Demons-Meigs' syndrome. A case of thecoma and ovarian fibroma. *J Gynecol Obstet Biol Reprod*, 21 (1992), pp. 651-4
- IX. Lin JY, Angel C, Sickel J. Meigs' syndrome with elevated serum CA 125. *Obstet Gynecol*, 80 (1992), pp. 563-6
- X. Stratan D, Socolov R. Meigs' syndrome. A case report and review of literature. *Rev Med Chir Soc Med Nat Iasi*, 96 (1992), pp. 30-2
- XI. Novoa-Vargas A, Tenorio González F, Gómez Pedroso J, Torres A. Meigs' syndrome. A clinical case and review of the literature. *Ginecol Obstet Mex*, 62 (1994), pp. 217-21
- XII. Siddiqui M, Toub DB. Cellular fibroma of the ovary with Meigs' syndrome and elevated CA 125. *J Reprod Med*, 40 (1995), pp. 817-9