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Case Report

Case report: Sclerosing stromal tumor in a young female: Clinical presentation and management

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ABSTRACT

Background: Sclerosing stromal tumors (SSTs) are rare ovarian neoplasms predominantly affecting young females during their second or third decade of life. This case report presents a comprehensive analysis of a young female patient diagnosed with an ovarian SST, highlighting its clinical presentation, histopathological characteristics, and management.

Clinical Presentation: The emergence of SSTs poses diagnostic challenges due to their rarity and varied clinical manifestations, often mimicking other ovarian neoplasms. Patients typically present with nonspecific symptoms such as abdominal pain, bloating, and irregular menstrual cycles. Imaging studies and histopathological examination are essential for accurate diagnosis.

Histopathological Characteristics: Histologically, SSTs are characterized by a proliferation of fibrous stromal cells interspersed with varying degrees of collagen deposition and hyalinization, posing a diagnostic challenge. Immunohistochemical analysis plays a pivotal role in confirming the diagnosis.

Management: Surgical resection remains the cornerstone of management, with the aim of complete excision while preserving ovarian function whenever possible. Frozen section analysis during surgery aids in intraoperative decision-making. Adjuvant therapy is generally not indicated due to the benign nature of SSTs.

Conclusion: A multidisciplinary approach involving gynecologists, pathologists, and radiologists is crucial for accurate diagnosis and optimal management of SSTs, ensuring favorable outcomes for affected patients. Continued research efforts are warranted to further elucidate the underlying pathogenesis of SSTs and refine treatment algorithms for these rare entities.

1. Introduction

A rare ovarian tumor known as sclerosing stromal tumor typically manifests in young adults during the second and third decades of life. In this report, we present the clinical and histopathological characteristics of a case involving a sclerosing stromal tumor of the ovary, along with a comprehensive review of existing literature on the subject. These tumors exhibit distinct histological features and can be readily identified when there is a heightened level of suspicion in young patients presenting with an ovarian mass. Despite their rarity, sclerosing stromal tumors are generally benign and amenable to successful treatment through enucleation or unilateral ovariotomy.[1]

Ovarian sex cord tumors, including sclerosing stromal tumors, are relatively uncommon neoplasms, comprising approximately 8% of all primary ovarian neoplasms. Our report focuses on the clinical findings and histopathological characteristics of a case involving a sclerosing stromal tumor of the ovary encountered at Saveetha Medical College and Hospital in Chennai.

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The emergence of a sclerosing stromal tumor in the ovary poses a diagnostic challenge due to its rarity and varied clinical presentations. These tumors often masquerade as other ovarian neoplasms, such as granulosa cell tumors or fibromas, necessitating a keen clinical acumen and a thorough understanding of their distinctive features for accurate diagnosis and appropriate management. Typically, patients with sclerosing stromal tumors present with nonspecific symptoms, including abdominal pain, bloating, and irregular menstrual cycles. However, in some instances, they may be asymptomatic and incidentally discovered during routine pelvic examinations or imaging studies.[2]

Histologically, sclerosing stromal tumors are characterized by a proliferation of fibrous stromal cells interspersed with varying degrees of collagen deposition and hyalinization. These tumors often exhibit a biphasic pattern, with alternating areas of cellular and fibrotic components, further complicating their diagnosis. Immunohistochemical analysis plays a crucial role in distinguishing sclerosing stromal tumors from other ovarian neoplasms, with positive staining for vimentin and CD99 and negative staining for inhibin and calretinin being characteristic of these tumors.[3]

The management of sclerosing stromal tumors primarily involves surgical resection, aiming for complete excision while preserving ovarian function whenever feasible. In cases where fertility preservation is not a concern, unilateral salpingo-oophorectomy may suffice, whereas conservative approaches such as enucleation or cystectomy are preferred in young patients desiring future fertility. Given the benign nature of these tumors, adjuvant therapy is generally not indicated, and long-term prognosis following surgical resection is excellent, with low rates of recurrence reported in the literature.[4,5]

Our case report adds to the limited body of literature on sclerosing stromal tumors of the ovary, providing valuable insights into their clinical presentation, histopathological features, and management strategies. Through a multidisciplinary approach involving gynecologists, pathologists, and radiologists, accurate diagnosis and optimal management of these rare ovarian tumors can be achieved, thereby ensuring favorable outcomes for affected patients.

2. Case Report

23 year old, unmarried, came with complaints of intermenstrual bleeding since 4-5 months, history of abdominal pain on and off for 1 week usg showed 8.1*6.7cm large heterogenous lesion in right adnexa with multiple cystic areas and increased vascularity, no evidence of any calcification, both ovaries not seen seperately, tumour markers are done, negative patient was taken up for laparoscopic cystectomy 8*7 cm left side cyst seen adherent to left ovary intra operatively, HPE showed sclerosing stromal tumor of ovary –sex cord stromal tumor. Post operative period uneventful. Ovarian neoplasms present a complex and varied spectrum of tumors, with unique histological subtypes and clinical manifestations. In adolescents, ovarian tumors are relatively rare occurrences, predominantly consisting of tumors originating from germ cells. However, among the diverse array of ovarian neoplasms, sex cord-stromal tumors represent a distinct category, encompassing granulosa cell tumors, fibrothecomas, Sertoli and Leydig cell tumors, steroid cell tumors, and the less common sclerosing stromal tumors. While germ cell tumors predominate in this age group, the presence of sex cord-stromal tumors, particularly sclerosing stromal tumors, highlights the importance of considering a broad differential diagnosis in young patients presenting with gynecological symptoms.

Sclerosing stromal tumors, though constituting only a minority (6%) of sex cord-stromal tumors, exhibit unique clinical and histopathological features that warrant specific attention. These tumors have a predilection for affecting younger women, with the average age of diagnosis typically around 30 years. However, as evidenced by the presented case, individuals below this age threshold may also be affected, underscoring the importance of maintaining a high index of suspicion for ovarian neoplasms in young patients presenting with pertinent symptoms.

Clinical manifestations commonly associated with sclerosing stromal tumors are diverse, often presenting with a constellation of symptoms that may include menstrual irregularities, pelvic pain, and nonspecific symptoms attributable to the presence of a pelvic mass. Menstrual irregularities, such as intermenstrual bleeding or irregular cycles, may raise suspicion for an underlying ovarian pathology and prompt further investigation. Additionally, pelvic pain, ranging from mild discomfort to severe abdominal cramping, may be reported by affected individuals and may be attributed to the mass effect exerted by the tumor on surrounding structures or secondary to torsion or hemorrhage within the tumor itself. Furthermore, nonspecific symptoms such as abdominal distension, urinary urgency, or bowel habit changes may result from the presence of a pelvic mass and its impact on neighboring organs. The variability in clinical presentation underscores the importance of a thorough history and physical examination in the evaluation of patients with suspected ovarian neoplasms, with particular attention to gynecological and menstrual history.

Diagnostic evaluation of suspected sclerosing stromal tumors typically begins with imaging studies aimed at characterizing the morphology and vascularity of the ovarian mass. Ultrasonography remains the initial modality of choice due to its widespread availability, non-invasiveness, and ability to provide real-time imaging. On ultrasonography, sclerosing stromal tumors may present as

solid-cystic lesions with varying degrees of vascularity, often demonstrating a heterogeneous appearance due to the presence of fibrous and cystic components. Magnetic resonance imaging (MRI) may provide additional characterization of the tumor, particularly in cases where ultrasonographic findings are inconclusive or when there is a need for further delineation of tumor extent and its relationship to adjacent structures. However, it is essential to recognize that imaging findings of sclerosing stromal tumors may lack specific features to definitively distinguish them from other ovarian neoplasms, necessitating histopathological examination for accurate diagnosis.

Histologically, sclerosing stromal tumors are characterized by a proliferation of fibrous stromal cells interspersed with varying degrees of collagen deposition and hyalinization. This histological pattern imparts a biphasic appearance to the tumor, with alternating areas of cellular and fibrotic components, posing a diagnostic challenge. Immunohistochemical analysis plays a pivotal role in confirming the diagnosis, with sclerosing stromal tumors typically exhibiting positive staining for vimentin and CD99, while staining negatively for inhibin and calretinin. These immunohistochemical markers aid in differentiating sclerosing stromal tumors from other ovarian neoplasms and are instrumental in guiding further management decisions.

The cornerstone of management for sclerosing stromal tumors is surgical resection, with the primary objective being complete excision while preserving ovarian function whenever feasible. The surgical approach may vary depending on factors such as tumor size, location, and patient preferences regarding fertility preservation. In cases where fertility preservation is a consideration, conservative surgical approaches, such as enucleation or cystectomy, may be employed to spare the affected ovary. Alternatively, unilateral salpingo-oophorectomy may be performed in cases where there is a concern for malignancy or when the tumor extensively involves the ovary. Importantly, given the benign nature of sclerosing stromal tumors, adjuvant therapy such as chemotherapy or radiation is generally not indicated, and long-term prognosis following surgical resection is favorable, with low rates of recurrence reported in the literature.

In conclusion, the unique clinical and histopathological characteristics of sclerosing stromal tumors highlight the importance of considering this rare ovarian neoplasm in the differential diagnosis of young women presenting with gynecological symptoms. Through a comprehensive diagnostic approach involving imaging studies, histopathological examination, and immunohistochemical analysis, accurate diagnosis and optimal management of sclerosing stromal tumors can be achieved, ultimately ensuring favorable outcomes for affected patients. Continued research efforts are warranted to further elucidate the underlying pathogenesis of sclerosing stromal tumors and to refine treatment algorithms for these rare entities.

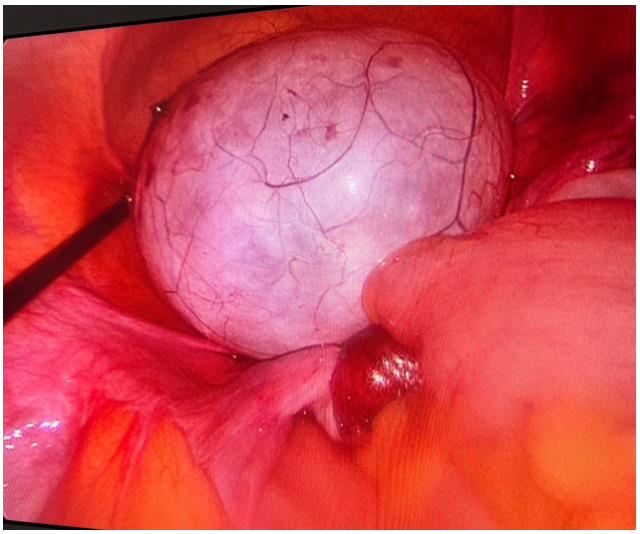

Figure 1: Intra op image of left ovarian cyst

3. Discussion

Sclerosing stromal tumors (SSTs) offers valuable insights into the clinical presentation, differential diagnosis, and diagnostic challenges associated with this rare ovarian neoplasm. Chalvardjian and Scully's seminal description in 1973 laid the foundation for understanding this entity, which comprises a small proportion (2% to 6%) of ovarian tumors originating from the stroma. Despite being recognized for nearly five decades, the literature documents less than 100 cases, highlighting its rarity and the limited understanding of its pathogenesis and clinical behavior.[5] The predilection for young adults, particularly during the second and third decades of life, underscores its occurrence in reproductive-age individuals, with over 80% of cases reported in this demographic Chalvardjian A et al.[6]

The hormonal activity of SSTs varies, with most being hormonally inactive. However, when hormonally active, these tumors often manifest androgenic characteristics and are frequently encountered in the setting of pregnancy Tameish S.[7] Clinical manifestations typically include menstrual irregularities and pelvic pain, though these symptoms may vary depending on the tumor's size and hormonal activity Mehra P et al.[8]

Given the diverse histological spectrum of ovarian tumors, distinguishing SSTs from other entities is crucial for accurate diagnosis and appropriate management. Thecoma/fibroma, metastases, and malignant epithelial ovarian tumors constitute important differentials that must be considered Shi J et al.[9] Particularly challenging is

the discrimination between SSTs and juvenile granulosa cell tumors (JGCTs) with prominent stromal sclerosis. While both entities may exhibit overlapping features, certain histopathological characteristics aid in differentiation. SSTs often demonstrate a characteristic vascular pattern and mitotic activity, whereas JGCTs typically present with follicular structures, increased mitotic activity, and typical granulosa cell morphology Anantharaju A et al.[10] However, this demarcation may not always be straightforward, necessitating ancillary techniques such as immunohistochemistry for accurate classification.

Rarely, the presence of vacuolated cells and signet ring cells within an edematous stroma may mimic Krukenberg tumors, posing a diagnostic challenge. Immunohistochemical profiling plays a pivotal role in distinguishing SSTs from metastatic adenocarcinomas, ensuring appropriate clinical management Fischer AK et al.[11]

SSTs represent a rare subset of ovarian tumors characterized by distinctive clinical and histological features. Despite their infrequent occurrence, clinicians must be vigilant in considering SSTs in the differential diagnosis of ovarian neoplasms, employing a comprehensive approach that integrates clinical, radiological, and histopathological findings for accurate diagnosis and optimal patient care.

4. Conclusion

Diagnosing ovarian sclerosing stromal tumors (SST) relies on a comprehensive understanding of the key characteristics associated with this rare neoplasm. One of the hallmark features is the age of the patient, with SSTs typically occurring during the second or third decade of life. This demographic trend underscores the importance of considering SSTs in the differential diagnosis of ovarian tumors in young adults, especially in reproductive-age females presenting with pelvic masses or related symptoms. Furthermore, the unilateral presentation of the tumor is often observed, which can aid in distinguishing SSTs from other ovarian pathologies, although bilateral cases have been reported in the literature, albeit less frequently.

In addition to demographic factors, the macroscopic and histopathological features of SSTs provide essential diagnostic clues. Macroscopically, these tumors typically present as solid masses with variable degrees of fibrosis and calcifications. The firm consistency and well-defined borders of SSTs contrast with the cystic nature commonly observed in other ovarian neoplasms. Histopathologically, SSTs are characterized by a proliferation of spindle cells arranged in a storiform or fascicular pattern within a collagenous or fibrotic stroma. The presence of characteristic vascular patterns, such as prominent hyalinized vessels, further supports the diagnosis of SSTs. Moreover, immunohistochemical analysis can be instrumental in confirming the diagnosis by demonstrating positive staining for vimentin, CD10, and smooth muscle actin, while typically being negative for inhibin and calretinin, which are commonly expressed in granulosa cell tumors.

Given the rarity of SSTs and the potential for diagnostic ambiguity, a comprehensive diagnostic approach is warranted. This includes conducting tumor marker assessments, hormone tests, ultrasonography, and magnetic resonance imaging (MRI) studies. Tumor markers such as CA-125 may be elevated in some cases, although they lack specificity for SSTs and are often within normal limits. Hormone tests are particularly relevant in cases where the tumor exhibits hormonal activity, such as androgenic manifestations, which may necessitate further endocrine evaluation and management. Imaging studies, including ultrasonography and MRI, are essential for characterizing the extent of the tumor, assessing its vascularity, and delineating its relationship with surrounding structures. Specifically, ultrasonography can provide valuable information regarding the tumor's size, echogenicity, and presence of calcifications, while MRI offers superior soft tissue resolution and can aid in surgical planning.

In the surgical setting, obtaining frozen biopsies of the ovarian mass is recommended to facilitate intraoperative decision-making regarding the extent of surgical resection and to guide subsequent management strategies. Frozen section analysis allows for rapid histopathological evaluation of the tissue, enabling the surgeon to assess the nature of the lesion and determine the need for further surgical intervention, such as oophorectomy or fertility-sparing procedures. Moreover, frozen section analysis can help differentiate SSTs from other ovarian tumors with overlapping histological features, thereby preventing diagnostic pitfalls and ensuring appropriate management.

Diagnosing ovarian sclerosing stromal tumors (SSTs) requires a multifaceted approach that integrates clinical, radiological, and histopathological findings. Key characteristics essential for diagnosis include the patient's age, tumor presentation, macroscopic and histopathological features, as well as ancillary diagnostic modalities such as tumor markers, hormone tests, and imaging studies. Additionally, frozen section analysis during surgery plays a pivotal role in facilitating intraoperative decision-making and ensuring optimal patient management. By employing a comprehensive diagnostic strategy, clinicians can accurately identify and appropriately manage SSTs, thereby optimizing patient outcomes and prognosis.

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None.

6. Conflict of Interest

None.

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