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Case Study

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SIGNET RING CELL STROMAL TUMOR OF THE OVARY: A RARE CASE REPORT

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ABSTRACT

Signet ring cell stromal tumor of the ovary is an exceptionally rare subtype of sex cord-stromal tumors characterized by signet ring-like morphology without mucin production. These tumors can pose diagnostic challenges due to their unusual histological appearance, which may mimic metastatic signet ring cell carcinomas, particularly from the gastrointestinal tract. We present a case of a 52-year-old postmenopausal woman with an incidental finding of an ovarian mass discovered during evaluation for nonspecific abdominal discomfort. Histopathological and immunohistochemical analysis confirmed the diagnosis of a signet ring cell stromal tumor. This case underscores the importance of accurate morphological evaluation and immunoprofiling in the diagnosis of this rare entity to avoid misclassification and inappropriate management. A comprehensive review of the literature is also included to contextualize this rare diagnosis.

INTRODUCTION

Sex cord-stromal tumors of the ovary account for approximately 8% of all ovarian neoplasms and are typically composed of granulosa, theca, Sertoli, or Leydig cells, or a combination thereof [Young RH, 2005]. Within this category, the signet ring cell stromal tumor is exceedingly rare and was first described as a distinct subtype in recent years [McCluggage WG, 2016]. These tumors are histologically characterized by the presence of cells with intracytoplasmic vacuoles pushing the nucleus to the periphery, mimicking the morphology of signet ring cells seen in mucin-producing carcinomas [Zhang X et al., 2018].

The key diagnostic challenge lies in differentiating these tumors from metastatic signet ring cell carcinomas, especially those originating from the gastrointestinal tract, which are far more common and have significantly different clinical implications [Ulbright TM, 2000]. Immunohistochemical analysis is crucial for the correct diagnosis, as these stromal tumors typically express markers such as inhibin, calretinin, and FOXL2, and are negative for epithelial markers like cytokeratin 20 and CDX2 [Irving JA et al., 2006].

Given their rarity, very few cases have been reported in the literature, and long-term behavior remains uncertain. Most documented cases suggest a benign clinical course [Zamecnik M, 2005]. Herein, we report a rare case of a signet ring cell stromal tumor in a postmenopausal woman, emphasizing the histopathological and immunohistochemical features that led to the diagnosis.

CASE DETAILS

A 52-year-old postmenopausal woman presented to the outpatient gynecology clinic with complaints of intermittent lower abdominal discomfort for three months. The pain was dull, non-radiating, and not associated with gastrointestinal or urinary symptoms. She denied weight loss, altered bowel habits, or vaginal bleeding. Her medical history was unremarkable, and she had no prior surgeries. Family history was negative for any gynecologic or gastrointestinal malignancies.

On physical examination, the abdomen was soft, and no palpable mass was noted. Speculum and bimanual pelvic examinations were unremarkable. Routine blood investigations, including complete blood count and liver and renal function tests, were within normal limits. Tumor markers including CA-125, CEA, and CA 19-9 were also within normal ranges.

Transvaginal ultrasound revealed a well-defined, hypoechoic, solid lesion measuring approximately 4.8 × 4.5 × 3.6 cm in the left adnexal region. MRI pelvis showed a well-circumscribed, solid ovarian mass with intermediate T1 and T2 signal intensity without evidence of ascites or lymphadenopathy. The radiologic impression favoured a benign ovarian neoplasm.

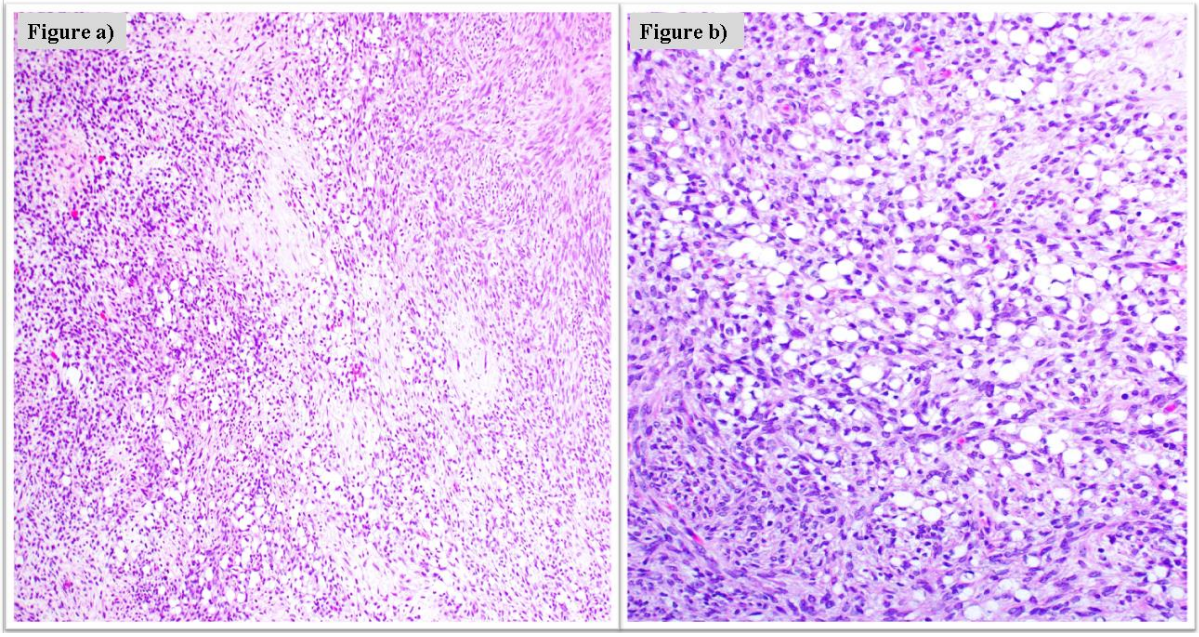
The patient underwent laparoscopic left salpingo-oophorectomy. Intraoperatively, the left ovary was enlarged with a firm, smooth, encapsulated mass. The uterus and contralateral adnexa appeared grossly normal.

No peritoneal implants or ascites were observed. The specimen was sent for histopathological examination.

Pathological Findings: Grossly, the ovarian mass measured 5.0 × 4.8 × 4.0 cm, was firm in consistency, and had a gray-white cut surface with focal yellowish areas. No hemorrhage or necrosis was noted.

Microscopic examination revealed a cellular tumor composed of uniform round to polygonal cells with abundant eosinophilic to clear cytoplasm. The nuclei were eccentrically placed, imparting a signet ring-like appearance. These vacuolated cells were arranged in sheets and nests separated by fine fibrous septa. No mitotic figures or atypia were identified. Importantly, there was no evidence of mucin within the cytoplasm on periodic acid-Schiff (PAS) and mucicarmine staining.

Immunohistochemical Profile: The tumor cells were strongly positive for inhibin, calretinin, and FOXL2. They were negative for cytokeratin AE1/AE3, CK20, CDX2, and EMA. These findings supported a diagnosis of a sex cord-stromal tumor with signet ring cell morphology.



Postoperative recovery was uneventful. Given the benign features and complete excision, no adjuvant therapy was advised. The patient remained disease-free at one-year follow-up.

DISCUSSION

Ovarian tumors with signet ring cell morphology usually raise the suspicion of metastatic adenocarcinomas, especially Krukenberg tumors originating from the gastrointestinal tract [Ulbright TM, 2000]. However, primary ovarian tumors with signet ring-like stromal cells, though rare, have been reported and pose significant diagnostic dilemmas [McCluggage WG, 2016].

The signet ring cell stromal tumor represents a morphologic variant of ovarian sex cord-stromal tumors. Unlike true signet ring cells that contain mucin, these vacuolated cells contain lipid or other non-mucinous material, as evidenced by negative mucin stains [Zhang X et al., 2018]. This key histochemical distinction is critical in avoiding misdiagnosis.

Immunohistochemistry plays a pivotal role in differentiating these tumors from metastatic carcinomas. The expression of sex cord-stromal markers such as inhibin and calretinin, along with absence of epithelial markers, is strongly supportive of a primary ovarian stromal neoplasm [Irving JA et al., 2006]. Additionally, FOXL2 gene mutations, seen in adult granulosa cell tumors, may be present, further supporting the stromal origin [Shah SP et al., 2009].

Clinically, most signet ring cell stromal tumors present as incidental findings or with nonspecific symptoms such as abdominal discomfort or pelvic pressure. Radiologic imaging often reveals a well-circumscribed solid mass without features suggestive of malignancy, as was seen in our case.

The behavior of these tumors appears to be benign based on the limited number of cases reported in the literature. However, long-term data are lacking. Complete surgical excision remains the mainstay of treatment, and no adjuvant therapy is indicated unless malignant transformation or recurrence is observed [Young RH, 2005].

This case emphasizes the necessity for thorough histopathologic evaluation supported by immunohistochemistry to arrive at an accurate diagnosis. Given the mimickers in the differential diagnosis,

awareness and recognition of this entity are essential for appropriate clinical management.

CONCLUSION

Signet ring cell stromal tumor of the ovary is a rare and diagnostically challenging neoplasm that can mimic metastatic malignancy due to its unusual morphology. Accurate diagnosis relies on histopathological assessment combined with a detailed immunohistochemical panel. Recognizing this benign variant among ovarian neoplasms is critical to prevent overtreatment. This case highlights the importance of considering signet ring cell stromal tumors in the differential diagnosis of ovarian tumors with signet ring morphology and contributes valuable data to the limited existing literature.

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