

Adult Granulosa Cell Tumor of the Ovary: A Case Report and Review of The Literature

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Abstract

Case Report

Background: Adult granulosa cell tumors (AGCTs) are rare ovarian neoplasms, representing 2–5% of malignant ovarian tumors. Their clinical presentation is often related to estrogenic activity, commonly causing postmenopausal bleeding and endometrial abnormalities. Preoperative diagnosis remains challenging due to nonspecific symptoms and inconclusive imaging. **Case presentation:** We report the case of a 56-year-old postmenopausal woman who presented with abundant vaginal bleeding. Pelvic MRI demonstrated significant endometrial thickening but no identifiable ovarian mass. She underwent total hysterectomy with bilateral salpingo-oophorectomy. Histopathological examination revealed atypical endometrial hyperplasia associated with an adult granulosa cell tumor of the left ovary, confirmed by immunohistochemistry. Postoperative staging, including imaging and tumor markers, showed no evidence of metastasis or recurrence. The case was discussed in a multidisciplinary tumor board, and a strategy of clinical and biological surveillance was adopted. **Conclusion:** This case illustrates the diagnostic difficulties of AGCTs when imaging findings are noncontributory. It emphasizes the central role of histopathology and immunohistochemistry in establishing a definitive diagnosis, the importance of multidisciplinary management, and the need for long-term follow-up given the risk of late recurrence.

Keywords: Adult granulosa cell tumor; Ovary; Postmenopausal bleeding; Endometrial hyperplasia; Long-term follow-up.

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INTRODUCTION

Adult granulosa cell tumors (AGCTs) are rare ovarian neoplasms, accounting for approximately 2–5% of malignant ovarian tumors [1], and representing the majority (around 70%) of sex cord–stromal tumors [2]. They typically occur in middle-aged to postmenopausal women, with a peak incidence between 50 and 55 years [3]. Clinically, AGCTs are characterized by estrogenic activity, leading to manifestations such as postmenopausal bleeding, atypical endometrial hyperplasia, or even associated endometrial adenocarcinoma [4]. Although their evolution is usually indolent, they carry a risk of late recurrence, which requires long-term surveillance [5]. Diagnosis is based on histopathological examination, supported by immunohistochemistry. The FOXL2 (C134W) mutation, identified in more than 95% of cases, is considered a specific diagnostic marker of AGCT [6]. Biologically, serum inhibin B measurement is useful both for diagnosis and for postoperative surveillance [7]. The

standard treatment is surgery, most often hysterectomy with bilateral salpingo-oophorectomy. Chemotherapy is reserved for advanced or recurrent disease. Because of the risk of late recurrence, sometimes up to 20 years after initial treatment, prolonged clinical, radiological, and biological follow-up is essential [8]. We report the case of a patient with an AGCT of the ovary, illustrating the diagnostic and therapeutic challenges of this rare entity.

CASE PRESENTATION

A 56-year-old woman, married and mother of five, menopausal for three years, with no significant medical, familial, or gynecological history and no use of oral contraception, presented with a three-year history of heavy postmenopausal bleeding. No leucorrhea or other associated symptoms were reported, and the condition evolved in a context of preserved general status.

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Clinical examination revealed an ECOG performance status of 1, with no palpable pelvic mass and only mild hypogastric tenderness.

Pelvic MRI demonstrated a slightly enlarged uterus with an endometrium thickened to 25 mm, irregular outer margins, and homogeneous enhancement after contrast injection. Multiple intramyometrial microcysts suggestive of adenomyosis were also noted. No ovarian cyst was detected. The bladder, rectum, pelvic fat, and Douglas pouch were unremarkable.

Hysteroscopy with endometrial curettage was performed, and histopathological analysis revealed glandular and polypoid hyperplasia without atypia or

malignancy. The patient was placed under clinical and ultrasound follow-up with symptomatic treatment.

However, the symptoms progressively worsened, which justified performing a total hysterectomy with bilateral salpingo-oophorectomy.

Postoperative histological examination revealed atypical endometrial hyperplasia with a suspicious focus of invasion, as well as a round-cell proliferation in the left ovary, raising suspicion of a granulosa cell tumor and warranting immunohistochemistry. Immunohistochemical analysis confirmed atypical endometrial hyperplasia without invasion and an adult granulosa cell tumor of the left ovary.

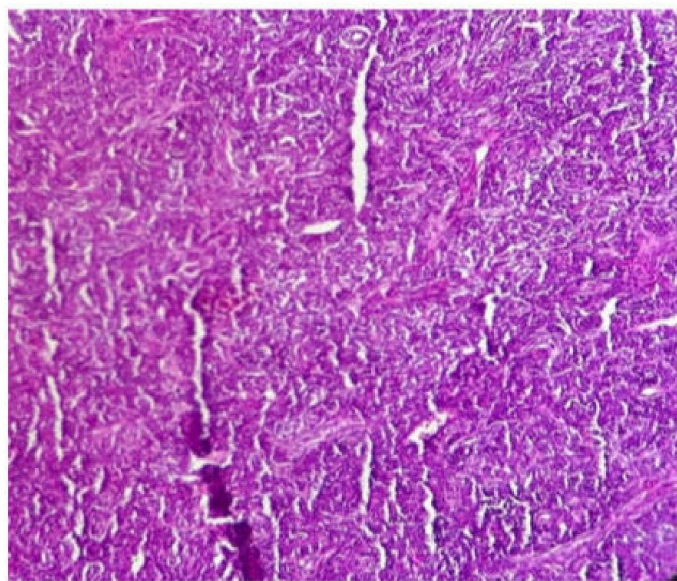


Figure 1: Hex10: Tumor proliferation of round cells with diffuse architecture

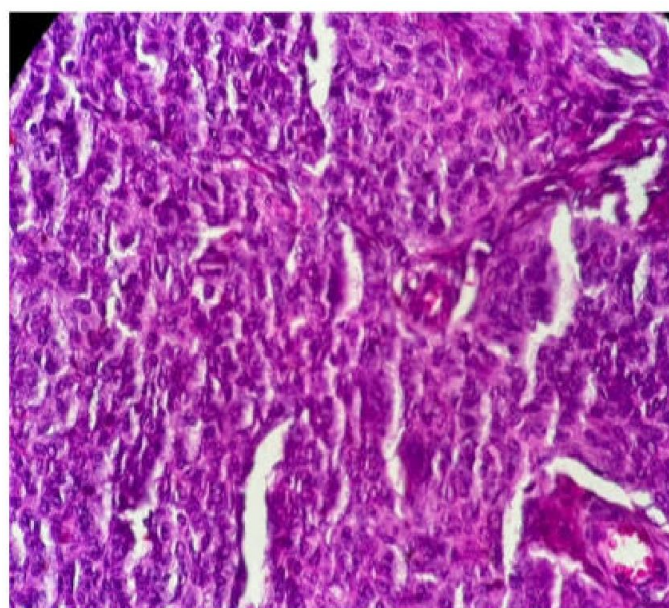


Figure 2: Hex40: Uniform tumor cells with angular nucleus

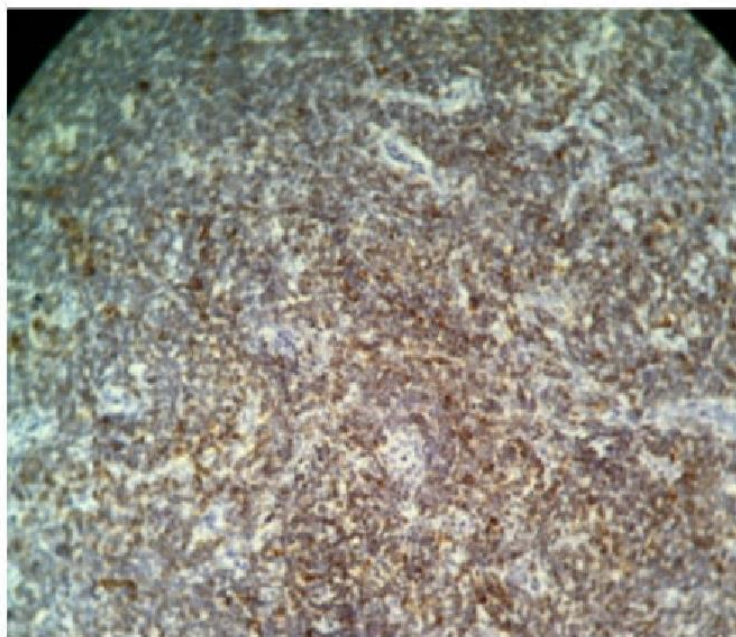


Figure 3: Diffuse staining of tumor cells with anti-inhibin antibody

Postoperative thoraco-abdomino-pelvic CT scan showed no abnormalities or evidence of recurrence. Tumor markers including CA125, AFP, and inhibin B were within normal limits. The case was discussed at the multidisciplinary gynecology-oncology tumor board, and a decision for clinical and biological surveillance only was adopted.

DISCUSSION

AGCTs are rare tumors, accounting for 2–5% of malignant ovarian neoplasms [1]. They arise from sex cord–stromal cells, which retain endocrine activity, mainly estrogen secretion, leading to a distinct clinical and biological profile. They typically occur in middle-aged to postmenopausal women, consistent with our patient's profile [2,3].

The clinical presentation of AGCT is most often dominated by estrogen-related manifestations, in particular postmenopausal bleeding. Endometrial changes such as hyperplasia, atypical hyperplasia, or carcinoma may occur due to prolonged estrogenic stimulation [4]. Indeed, 5–25% of patients with AGCT are reported to develop associated endometrial lesions, making systematic endometrial biopsy mandatory in cases of postmenopausal bleeding [4]. Our case perfectly illustrates this association, with initial endometrial thickening misinterpreted as adenomyosis, later confirmed as atypical hyperplasia.

Preoperative diagnosis of AGCT is often challenging because the tumors are usually small or moderate in size and may be missed by imaging. Pelvic MRI, although useful, may fail to detect the ovarian lesion, as in our case [4]. This highlights the importance

of combining imaging, clinical assessment, and histopathology.

Histopathology and immunohistochemistry remain the gold standard for diagnosis. The discovery of the FOXL2 C134W mutation in nearly all AGCTs represents a major advance, helping distinguish them from other stromal or epithelial ovarian tumors [5,6]. In our case, immunohistochemistry confirmed the granulosa cell nature of the tumor.

Surgery remains the cornerstone of treatment. Total hysterectomy with bilateral salpingo-oophorectomy is the standard approach for postmenopausal women or when malignancy is suspected [7]. Lymphadenectomy is not routinely recommended, as lymph node metastases are rare [7]. For younger patients desiring fertility preservation, conservative surgery may be considered in early-stage disease.

Postoperative follow-up is based on clinical and biological surveillance, particularly inhibin B measurement, which is a sensitive and specific tumor marker for AGCT [7,8]. In our patient, normal postoperative inhibin B levels were reassuring and consistent with a decision for simple surveillance. Thoraco-abdomino-pelvic CT was also negative for distant spread.

The overall prognosis of AGCT is favorable, particularly when diagnosed at an early stage [8]. However, the main challenge remains the risk of late recurrence, which can occur more than 10 years after initial treatment [8,9]. This underscores the need for long-term surveillance, with clinical follow-up, targeted imaging, and biomarker monitoring.

This case highlights the necessity of suspecting AGCT in patients presenting with postmenopausal bleeding associated with endometrial abnormalities, the diagnostic limitations of preoperative imaging, and the essential role of a multidisciplinary approach combining surgery, histopathological assessment, and long-term oncological follow-up.

CONCLUSION

Adult granulosa cell tumors (AGCT) of the ovary are rare neoplasms, representing a small proportion of ovarian malignancies, and often present diagnostic challenges due to their nonspecific clinical manifestations and the limited sensitivity of imaging. Multidisciplinary management, integrating surgical intervention and thorough pathological and biological evaluation, is crucial for effective treatment and monitoring. Early favorable outcomes underscore the need for prolonged surveillance to identify potential late recurrences, which are characteristic of this tumor type.

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