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Case Report

Benign germ cell tumour and epithelial carcinoma – Rare synchronous presentation in unilateral ovary on background of endometriosis

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ABSTRACT

Ovarian carcimomas are the second most common gynecological malignancy of which more than 90% arise from surface epithelial cells. Endometriosis-related ovarian neoplasms (ERONs) are described as a group of tumors which include clear cell carcinoma, endometrioid carcinoma, and seromucinous borderline tumor. Benign cystic teratomas are commonest benign ovarian neoplasms derived from germ cells. Coexistence of benign cystic teratoma, endometrioid adenocarcinoma and clear cell carcinoma in the same ovary is very unusual. Our case presented with well differentiated endometrioid adenocarcinoma grade 2 with squamous metaplasia and a nodule of clear cell carcinoma on the background of endometriosis and benign cystic teratoma in the same ovary. To the best of our knowledge, this is the first case with such an unusual combination to be reported in the medical English literature.

1. Introduction

Most tumors of the ovary can be classified based on the anatomic structure into one of three major categories: surface epithelial tumors, sex cord stromal tumors and germ cell tumors. The malignant potential of these tumors are as follows:

1. Surface epithelial tumors -90%.
2. Sex cord stromal tumors -7%
3. Germ cell tumors -5%.[1]

15–20% of ovarian epithelial neoplasms are Borderline ovarian tumors. The majority of borderline tumors are serous and mucinous neoplasms seen most commonly in women of reproductive age.

Endometriosis is defined as presence of endometrial tissue consisting of both endometrial glands and stroma, outside of endometrium and myometrium.[2]

Endometriosis is an important precursor of epithelial ovarian cancer such as endometrioid and clear cell ovarian tumors.[3]

The risk of malignant transformation in endometriosis has been estimated to be 0.3–0.8%. Endometriosis Related Ovarian Neoplasms (ERON) include endometrioid carcinoma, clear cell carcinoma and seromucinous borderline tumor. Ovarian endometrioid carcinoma (OEC) is the most common type of ERON, occurring in approximately 75% of cases.[4,5]

Synchronous tumors/lesions represent simultaneous occurrence of different tumors/lesions in more than one organ at the same time. The most common Synchronous tumors of the female genital tract are malignancies of endometrium and ovary accounting for 50% to 70% of all cases.

The most frequent combination is that of endometrioid adenocarcinoma of endometrium and ovarian endometrioid adenocarcinoma. Synchronous tumors of completely different histological types and of different histogenesis are

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very rare.[6]

Here, we describe an unusual combination of benign germ cell tumour and epithelial carcinoma in the same ovary on the background of endometriosis.

2. Case History

A 50-year old female patient was admitted with complaint of irregular menses and intermittent spotting since 1 year. Per abdomen examination revealed a 20-week cystic mass.

MRI of abdomen and pelvis showed multiple intramural uterine fibroids and a pedunculated mass prolapsing through internal os. Right ovary showed simple hemorrhagic cyst. A large complex multilocular left ovarian cyst of size 12x15x17cm with focal wall calcification and intraluminal polypoidal nodules noted. Another cystic lesion of left adnexa of size 5cm in greatest dimension with focal fat intensity locules noted. A probable diagnosis of ? left ovarian teratoma with endometrial polyp was given. We received specimen of total abdominal hysterectomy with bilateral ovarian cystectomy.

2.1. Gross examination

Uterus 7.5 x6.5 x5.5 cm. Rt. ovarian cyst 4 x3.5x1 cm. 2cysts noted at Left adnexal site. Smaller Left. cyst 4 x2.5x1.5 cm, containing hair and adipose tissue. Larger Left ovarian cyst measured 15 x12 x5 cm. C/s of Left larger cyst showed intraluminal polypoidal projections largest measuring 3 cm. Luminal surface of both Right and Left ovarian cysts showed chocolate brown contents. C/s endometrial cavity showed a polyp of size 2.5cm. Rest of the endometrium showed focal hyperplasia. Myometrium showed multiple fibroids.

2.2. Microscopic impression

Larger left ovarian cyst showed intracystic, polypoidal, well differentiated Endometrioid adenocarcinoma Grade II with Squamous metaplasia and a nodule of clear cell carcinoma on background of borderline endometrioid tumor and endometriosis. No capsular or vascular invasion seen. Smaller left ovarian cyst showed a mature cystic teratoma. No immature elements seen. Right adnexal mass showed endometriotic cyst. Uterus showed benign endometrial polyp, endometrial hyperplasia with no atypia, adenomyosis and multiple fibroids.

3. Discussion

Endometriosis is very commonly seen in woman of reproductive age affecting upto one out of ten females.[7]

Various theories have been proposed to explain endometriosis. Some of them are

1. Retrograde menstruation theory: during menses endometrial lining cells travel backwards through Fallopian tubes reach the peritoneal cavity, proliferate and cause chronic inflammation with formation of adhesions.
2. Coelomic metaplasia theory: Coelomic cells lining the pelvic peritoneum undergo metaplastic transformation to form endometrial tissue.
3. Induction theory: a combination of the first 2 theories.[8]

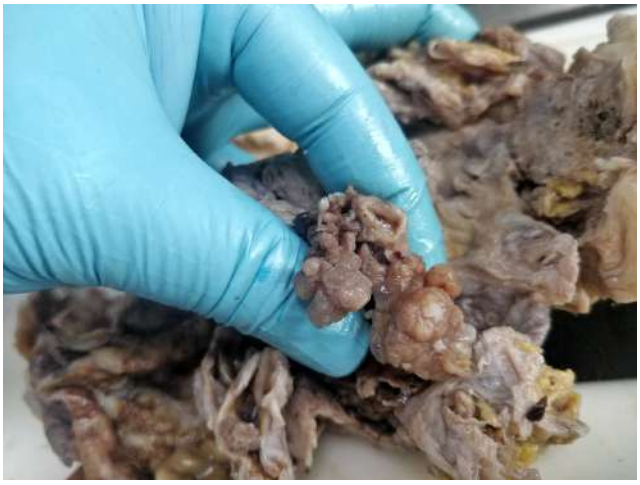

Fig. 1: Intracystic polypoidal excrescences

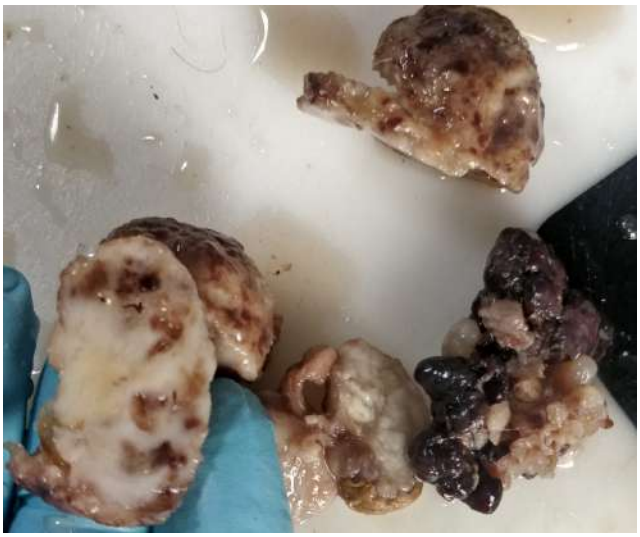

Fig. 2: Intracystic polypidal excrescences with foci of endometriosis

Molecular evidence suggests that the presence of ARID1A mutation is regarded as the most important genetic alteration leading to malignant transformation of endometriotic tissue. Malignant tumors commonly associated with endometriosis are endometrioid carcinoma (EC) and clear cell carcinoma (CCC).[9,10]

Fig. 3: Benign cystic teratoma with hair and adipose tissue

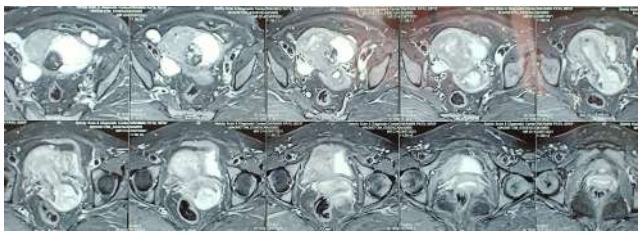

Fig. 4: Bilateral ovarian mass

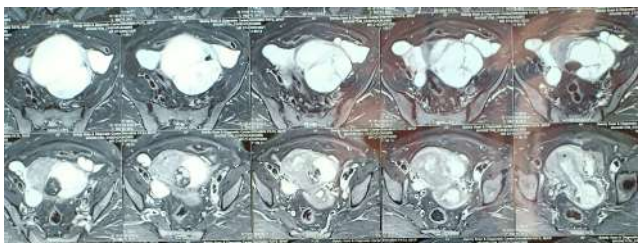

Fig. 5: Bilateral ovarian mass

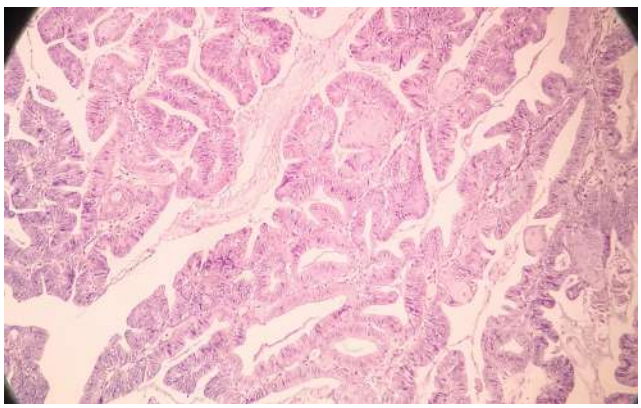

Fig. 6: Endometrioid adenocarcinoma, H&E 10X magnification

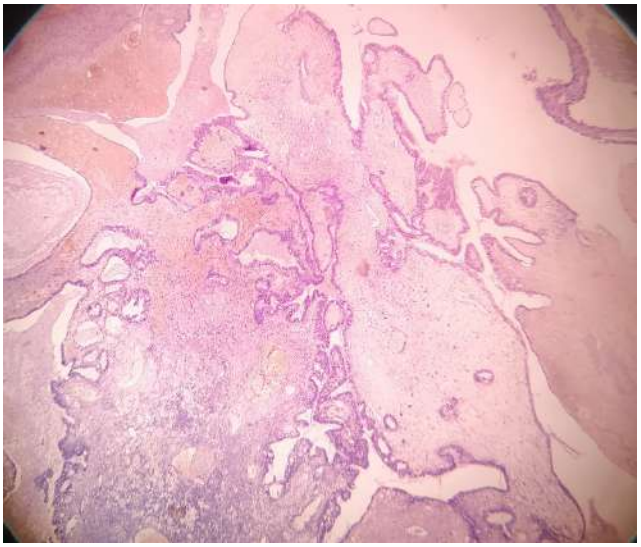

Fig. 7: Borderline tumour with endometriosis, H&E 10X magnification

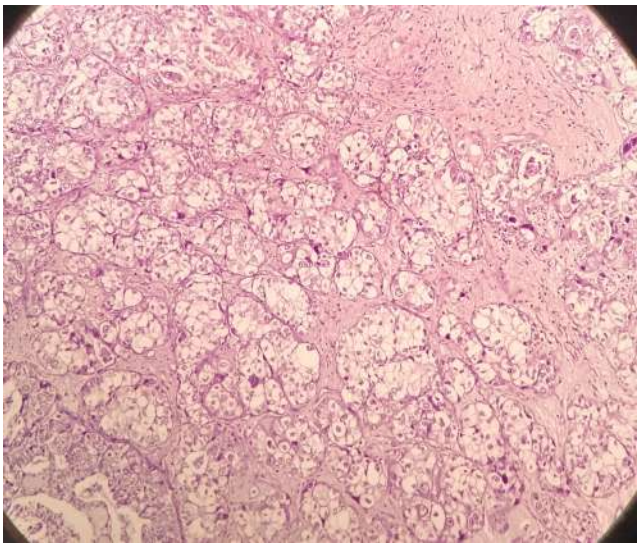

Fig. 8: Focus of clear cell carcinoma, H&E 40X magnification

Synchronous tumors of completely different histological types and of different histogenesis are very rare. This case constitutes a potpourri of tumors with unrelated histogenesis in a single ovary. Extensive review of literature did not reveal any case with a combination of such tumors in the female genital organ.

Such combination tumors are more commonly seen in association with hereditary mutation syndromes like nonpolyposis colon cancer (Lynch) syndrome, breast ovarian cancer syndrome, Li-Fraumeni syndrome etc. Our case had no clinical evidence of any syndromic association.[6]

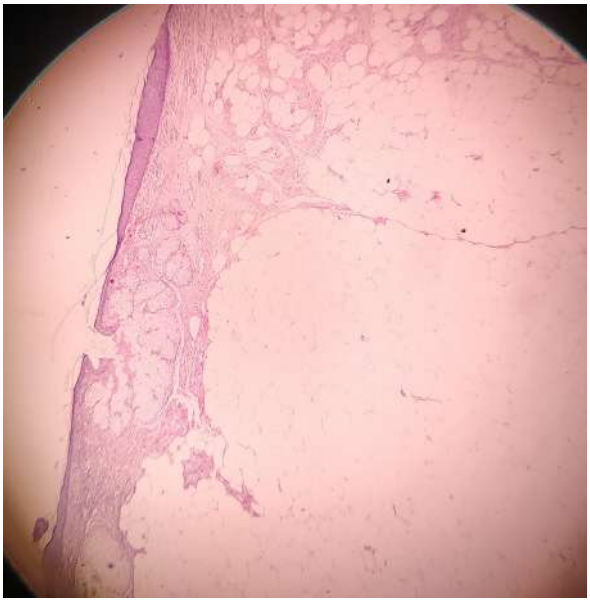

Fig. 9: Benign cystic teratoma, H&E 10X magnification

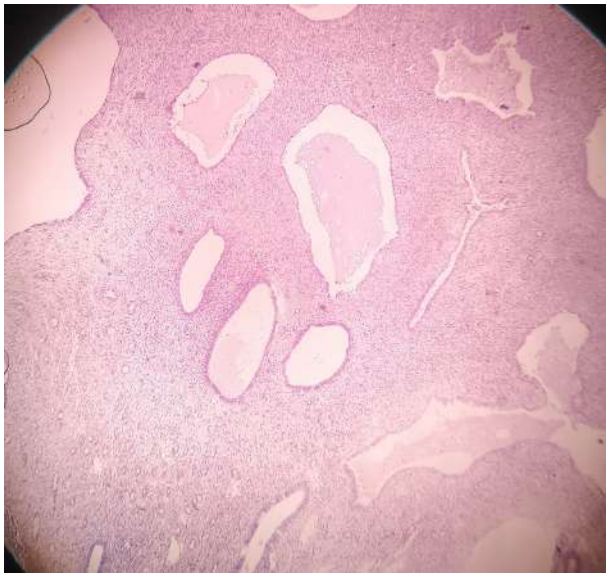

Fig. 10: Endometriosis, H&E 10X magnification

The incidence of synchronous tumors of different histological appearance is unexplainable. This probably may be a coincidental combination.

4. Conclusion

Hereby, we are reporting an unusual synchronous combination of tumors in the ovary of a 51-year old patient. This patient had a large endometriotic cyst with intracystic, polypoidal endometrioid adenocarcinoma, clear cell carcinoma and borderline endometrioid tumour –malignant epithelial tumours. Benign cystic teratoma of the same ovary- benign germ cell tumour. Apart from these tumors, patient also had adenomyosis, multiple fibroids and benign endometrial polyp of the uterus. Contralateral ovary showed endometriosis. To the best of our knowledge, such a combination is not reported in the English medical literature and this is the first case to be reported.

5. Conflict of Interest

The authors declare no conflict of interest.

6. Source of Funding

None.

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