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Clear Cell Carcinoma of Ovary - Rare Subtype with Poor Prognosis: A Case Report

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ABSTRACT

Ovarian clear cell carcinoma is a rare subtype of epithelial carcinoma of ovary having poor prognosis. This tumor has been found to be associated with endometriosis and has similarities with the pathogenesis of endometrioid carcinoma. We report a case of 37 years female presenting with the pain in right lower abdomen. Computerized Tomography revealed right ovarian cystic mass. Histopathology showed tubulocystic pattern of arrangement of tumor cells with few foci showing hobnail cells which confirmed the diagnosis.

Key words: Clear cell carcinoma, Ovary, Poor prognosis

INTRODUCTION

Ovarian clear cell carcinoma is a rare epithelial ovarian tumor with poor prognosis and high resistance to chemotherapy. This tumor has unique biological and clinical characteristics. It occurs more commonly in perimenopausal women and comprises approximately 15% to 25% of ovarian cancers which is higher when compared to 5% to 10% of ovarian cancer in North America and Europe. This might probably be due to genetic and environmental differences. [1] Most of them present as unilateral ovarian mass and are usually diagnosed in the early stages (i, e., in stage I / II). These tumors have drug resistant phenotype and the response rate to chemotherapy is lower when compared to other histologic types of ovarian cancer. [2] Numerous studies have proposed that this tumor is associated with endometriosis and has been thought to be malignant transformation of ectopic benign endometrium in ovary. [3] We are presenting a case of 37 years female with right ovarian mass.

CASE REPORT

37 years female presented with pain in the right iliac fossa since 3 months. On clinical examination 17X17cms lump was palpable in right lower abdomen. Contrast tomography of abdomen revealed well defined unilocular large cystic lesion with peripherally enhancing solid component arising from right adnexa which is displacing bowel loops superiorly and laterally, compressing right distal ureter causing moderate hydronephrosis. Clinical diagnosis was made as possibility of mucinous adenocarcinoma. Hematological parameters were within normal range except serum alkaline phosphatase which was 75IU/L. Total abdominal hysterectomy with

bilateral salpingo-oophorectomy was done. During surgical procedure adhesions were noted with posterior right ovarian cyst wall and anterior rectal wall.

We received ovarian cyst measuring 17.5X17.5X2cms attached with fallopian tube measuring 5.5cms in length. Cut section showed unilocular cyst filled with 150ml of greenish yellow colored serous fluid. Focal yellow solid and papillary excrescences measuring 14.7X13.5X3.5cms were seen (Figure 1). Also received uterus with cervix measuring 6X5X3cms, left ovary measuring 3.5X1X1cms and left fallopian tube measuring 3cms in length.

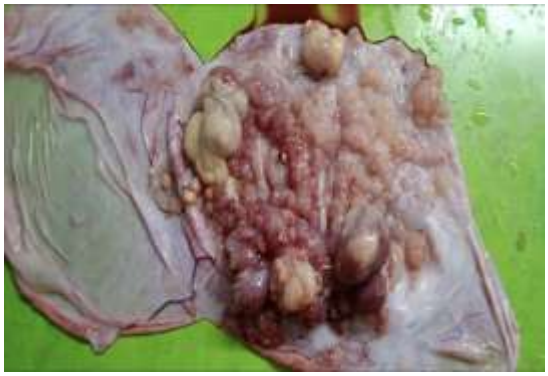

Figure 1: cut section of the ovarian cyst showing focal yellow solid and papillary excrescences

Microscopic examination of right ovarian cyst showed tumor cells arranged in tubulocystic and papillary pattern (Figure 2,3). Tumor cells were cuboidal to polygonal with mild nuclear pleomorphism and clear to eosinophilic cytoplasm (Figure 4). Few foci showed hobnail pattern of arrangement (Figure 5). In some foci cystic spaces were filled with eosinophilic material. Histopathological diagnosis of clear cell carcinoma was made.

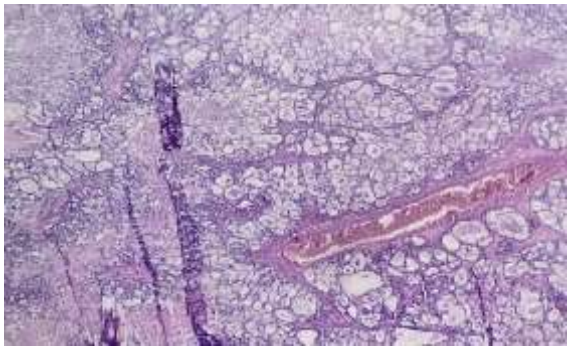

Figure 2: Tumor cells arranged in tubulocystic pattern (H&E,X40)

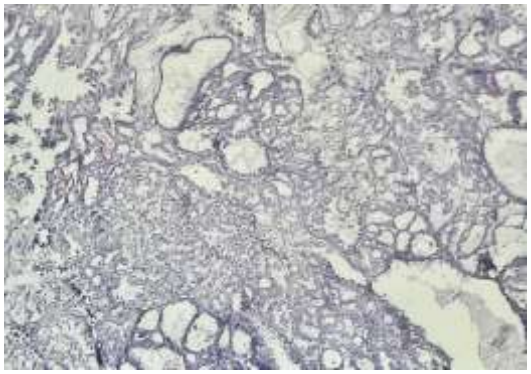

Figure 3: Tumor cells arranged in tubulocystic and papillary pattern (H&E,X100)

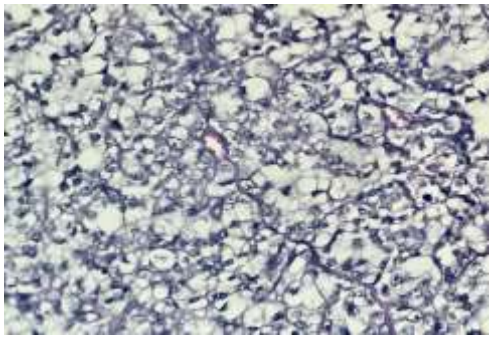

Figure 4: Tumor cells having clear cytoplasm and round to oval vesicular nuclei with prominent nucleoli (H&E,X400)

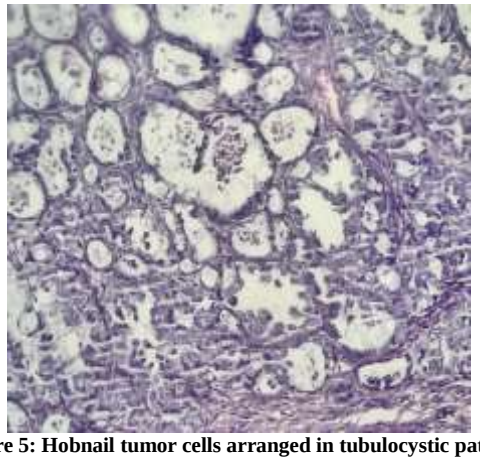

Figure 5: Hobnail tumor cells arranged in tubulocystic pattern (H&E,X200)

DISCUSSION

Ovarian carcinoma accounts for 2.5% of all malignancies in females and is one of the common malignancies of female reproductive system. [4] Ovarian carcinoma has poor prognosis with 5-year survival rate of 47.6% due to late stage diagnosis [5]. Histological type of ovarian cancer is an important prognostic factor and determines the response to chemotherapy. Among ovarian cancers, epithelial ovarian cancer accounts for 90% of ovarian malignancies. They are further subdivided into 5 main

histological subtypes. [6] Among the epithelial ovarian cancers, high grade serous carcinoma accounts for 70% to 80%. Other rare subtypes include low grade serous carcinoma (5%), endometrioid carcinoma (10%), mucinous carcinoma (6%), clear cell carcinoma (6%) and other rare and undefined cancers. [7]

Schiller in 1939, first named ovarian clear cell carcinoma as mesonephroma, as its origin was thought to be from mesonephric duct. [8] Later it was recognized as a distinct histologic subtype of ovarian cancer by World Health Organization in 1973 and was named as Ovarian clear cell carcinoma. [9]

Ovarian clear cell carcinoma was found to be associated with endometriosis. Pathogenesis of this tumor has shown similarities with endometrial carcinoma and has shown to have inactivation of tumor suppressor genes such as PTEN (Phosphatase and Tensin homologue), activation of PI3K/AKT/mTOR (Phosphoinositide-3 kinase/ protein kinase B/ mammalian) signal pathway, inactivation of ARID1A (AT-rich interactive domain 1A) and high microsatellite instability. [10]

Most of the patients are in the mean age of 50 – 53 years, presenting usually as abdominal or pelvic mass. This tumor is associated with vascular thrombotic events and paraneoplastic syndrome like hypercalcemia. Tumor may grow up to 30cms and is usually unilateral (8% are bilateral). Grossly tumor is uniloculated thick walled cyst with multiple yellow fleshy protrusions into the lumen. Microscopically tumor may exhibit different histologic patterns like tubulocystic, papillary or solid pattern. Tumor cells have clear cytoplasm which is due to the presence of glycogen and at times intracytoplasmic mucinous inclusions. Some of the cells have granular eosinophilic cytoplasm. Tumor also contains hyaline globules which are PAS positive. Mitotic activity is low. Tumor may also show necrosis, psammomatous calcifications and stromal lymphocytic infiltrate. Immunohistochemically tumor cells are positive for CK7, EMA, PAX 8, HNF 1β, Napsin A, AMACR. Tumor cells are negative for ARID1A, WT1, ER, PR, SALL4, OCT4, CD 10, and p16.

Clear cell carcinoma must be differentiated from low grade and high grade serous carcinoma, endometroid carcinoma, metastatic clear cell renal cell carcinoma, dysgerminoma and yolk sac tumor. Low grade and high grade serous carcinomas with clear cell features are positive for WT1 and ER, but negative for Napsin A and HNF 1β. Endometrioid carcinoma with clear cell features are positive for ER and PR and negative for Napsin A and HNF 1β. Metastatic clear cell carcinoma are positive for CD10 and negative for CK7, ER, PR, Napsin A and AMACR. Dysgerminoma and yolk sac tumors are positive for SALL4, AFP and Glypican 3, but negative for CK7.

CONCLUSION

Ovarian clear cell carcinoma is rare epithelial ovarian carcinoma having distinct biological and molecular characteristics. Immunohistochemistry helps us to identify this rare subtype. Identifying this subtype is important as it has poor prognosis and shows resistance to chemotherapy.

Declaration by Authors

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Conflict of Interest: The authors declare no conflict of interest.

REFERENCES

1. Schnack T H, Hogdall E, Nedergaard L, Hogdall C. Demographic clinical and prognostic factors of primary ovarian adenocarcinoma of serous and clear cell histology- A comparative study. Int. J. Gynecol. Cancer 2016;26(1):82-90
2. Jin Y, Li Y, Pan L. The target therapy of ovarian clear cell carcinoma. Onco Targets Ther. 2014;7:1647-1652
3. Shin H Y, Yang W, Chay D B, Lee E J, Chung J Y, Kim H S et al. Tetraspanin 1 promotes endometriosis leading to ovarian clear cell carcinoma. Mol. Oncol. 2021;15(4):987-1004

4. Torre L.A, Trabert B, De Santis C.E, Miller K.D, Samimi G, Runowicz C.D, et al. Ovarian cancer statistics. CA Cancer J. Clin 2018;68(4):284-296
5. Eisenhauer E.A, Real-world evidence in the treatment of ovarian cancer. Ann Oncol 2017;28:viii61-viii65
6. Lheureure S, Gourley C, Vergote I, and Oza A M. Epithelial ovarian cancer. Lancet 2019;393(10177):1240-1253
7. Barnes B.M, Nelson L, Tighe A, Burghel G.J, Lin I.H, Desai S et al. Distinct transcriptional programme stratify ovarian cancer cell lines into 5 major histologic subtypes. Genome Med. 2021;13(1):140
8. Saavedra J.A, Sandow J. Clear cell adenocarcinoma of ovary (Saphir and Lackner) and so called mesonephroma ovarii (Schiller) as combined tumors. Review of literature and case report. Arch. Gynakol 1968;206(2):131-153
9. Korenaga T.R, Ward K.K, Saenz C, Mc Hale M.T, Plaxe S. The elevated risk of ovarian clear cell carcinoma among Asian pacific Islander women in the united states is not affected by birth place. Gynecol. Oncol. 2020;157(1):62-66
10. Willis B.C, Sloan E.A, Atkins K.A, Stoler M.H and Mills A.M. mismatch repair status and PD-L1 expression in clear cells carcinoma of the ovary and endometrium. Mod Pathol 2017;30(11),1622-1632

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