

Clear Cell Adenocarcinoma of Ovary: A Rare Case Report and Review of Literature

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Abstract: Clear cell carcinoma of ovary comprises 5-12% of ovarian cancer. These are usually large tumors ranging from 10 to 30 cm in diameter with solid and partly cystic nodular lesion on their surfaces. It is usually associated with endometriosis, but paraneoplastic syndrome, hypercalcemia, venous thrombosis may occur occasionally. Prognosis of clear cell carcinoma of ovary is poor comparable to other common epithelial tumors as it does not respond to chemotherapy. Here we are reporting a case of clear cell carcinoma of ovary with distinct clinic-pathological and histological pattern in a 38 year old lady presented with pain abdomen and mass in lower abdomen underwent hysterectomy with bilateral salpingoophorectomy.

Keywords: clear cell adenocarcinoma, endometriosis, hypercalcemia, oophorectomy, ovary

I. Introduction

Clear cell adenocarcinoma of ovary is a type of the surface epithelial-stromal tumor comprising of 5-12% of all ovarian cancer having distinct clinico-pathological and morphological entity^{1,2,3}. These are usually seen in 5th decade of life and associated with exposure of diethylstilbestrol (DES) in utero. These are usually large size tumors mainly composed of clear cells but sometimes having hyperchromatic nuclei mimicking hobnail^{1,4,5}. However tubular, cystic, papillary, solid pattern or mixed pattern may be seen. In 1967 Scully and Barlow³ presented convincing evidence of the association of the so-called mesonephric carcinoma with endometriosis and endometriomata, suggesting the "Mullerian" nature of this epithelium. Atypical endometriosis is known to be precursor of clear cell carcinoma. Paraneoplastic syndrome, hypercalcemia, deep vein thrombosis, pulmonary embolic episodes are also associated with it. Recently mutations of ARID1A gene have been detected in clear cell carcinoma of ovary⁶.

The lesion became identified as a separate pathologic entity and was called clear cell carcinoma to avoid the histogenetic confusion implied in the term mesonephroma. Large aggregate of intra-cytoplasmic glycogen coalescing in to clear cell and stacked lamellae of granular endoplasmic reticulum are characteristic in electron microscopy.⁷ The tumors were diagnosed as clear cell tumor on the basis of a histological pattern that was either tubular, cystic, papillary, or solid, and in which the cells were either clear or hobnail. These tumors have relatively low mitotic activity and poor prognosis as it does not responds to cisplatin based chemotherapy⁸.

II. Case Presentation

Here we are presenting a case of 38 years old female presented with pain in right lower abdomen and whitish vaginal discharge for 5 years. On clinical examination, 16 weeks size lump was palpable in right lower abdomen. Ultrasound was suggestive of right ovarian dermoid cyst. Level of CA 125 and AFP were 39.5IU/ml and 0.46IU/ml respectively. Elective trans-abdominal hysterectomy with bilateral salpingoophorectomy was done. On gross examination, size of uterus 6cmX5.5cmX5cm, size of right ovary 10cmX7cmX4cm while left ovary was normal in size. Right ovary had both solid and cystic areas. Two nodular masses of size 4cmX3cmX3cm each with dirty yellow colour were seen. On cut section, solid area was grayish white in colour and firm in consistency. On microscopic examination, there were mainly tubulo-cystic pattern and hobnail pattern.

III. Figures



Fig1: Gross examination of right ovary



Fig2: Cut section of right ovary

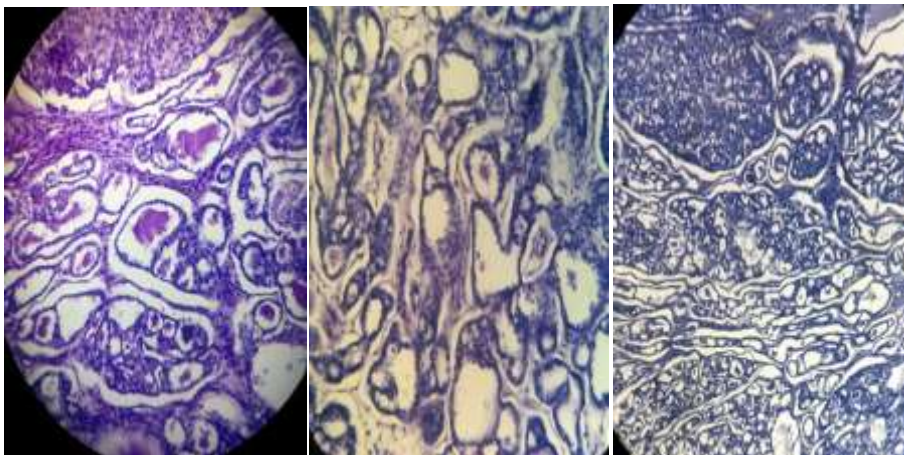


Fig 3: Tumor cells are arranged in tubulo-cystic pattern pattern, cysts contain eosinophilic secretions. **Fig 4:** Tumor cells arranged predominantly in tubulo-cystic pattern pattern, cysts contain eosinophilic secretions. Tubules are lined by cuboidal cells with atypical hyperchromatic nuclei. (H & E, 20X)

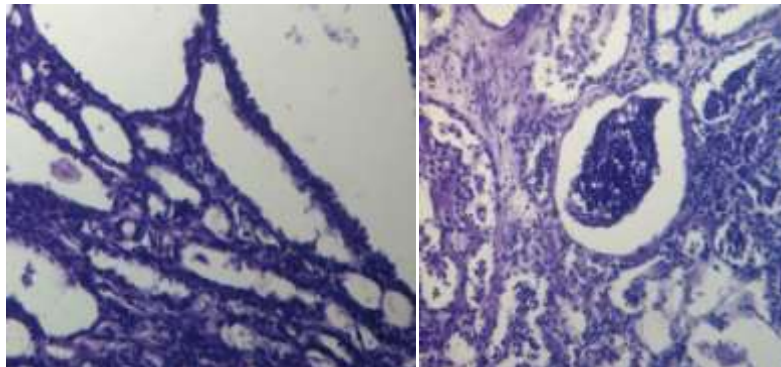


Fig 5: Tumor cells showing hob-nailing, hobnail cells are columnar cells and have either granular eosinophilic or clear cytoplasm and have hyperchromatic apical nuclei that bulge into the lumina. (H & E, 40X).

Fig 6: Vascular invasion by tumor cells.(H & E , 20X)

IV. Discussion

Ovarian clear cell carcinomas arise in endometrial tissue, either in areas of endometriosis or endometriomata as described by Scully and barlo, who found that 25% of their clear cell carcinomas arose in endometriomata and 53% were associated with pelvic endometriosis⁹. Unopposed high level of estrogen and in urero exposure of diethylstilbestrol (DES) are associated with ovarian clear cell carcinoma. Klemia and Gronroo found endometria with hyperplasia to carcinoma in 44% of their patients with clear cells carcinoma of the

ovary^{10,11}. However hormonal status of patients with clear cell carcinoma was clinically normal and there was no history of hormone therapy¹².

The histology of pure clear cell carcinoma of the ovary or endometrium, on the other hand, is distinct, consisting of clear or hobnail cells arranged in tubules, cysts, papillary formations, or sheets. Ultra-structurally, the neoplastic cells of clear cell carcinoma contain rough endoplasmic reticulum and large accumulations of glycogen^{13,14}. Clear cell carcinoma represents a variant of endometrioid carcinoma histo-genetically and histologically, as it is associated with a unique clinical presentation and prognosis.

V. Conclusion

Although clear cell carcinoma of ovary resemble with endometriosis and endometrioid carcinoma, but It should be considered as distinct entity on the basis of its etiological, morphological, clinico-pathological and histo-pathological peculiarity.

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