Case Report

DOI: 10.7860/JCDR/2016/15861.7780

Pathology Section

Clear Cell Adenocarcinofibroma Ovary – A Rare Histopathological Variant with Unusual Presentation

MANJIT KAUR[1], NAVTEJ SINGH[2], AMRIT PAL SINGH RANA[3]

ABSTRACT

The clear cell adenocarcinoma of ovary is a distinctive tumour. Stroma rich variants (adenocarcinofibroma) of these types of tumours are uncommon. We report a rare case of clear cell adenocarcinofibroma of ovary in 37 years of female presenting with bilateral ovarian masses. The tumour contained fibrous component and was radiologically indistinguishable from soft tissue malignancy. The serum level of CA-125 was raised. Histopathology and immunohistochemistry revealed the rare microscopic type of ovarian tumour. Because of differences in the clinical course as well as management, possibility of this unusual type of carcinoma must be ruled out.

Keywords: Surface epithelial tumor, Pancytokeratin, Malignancy

CASE REPORT

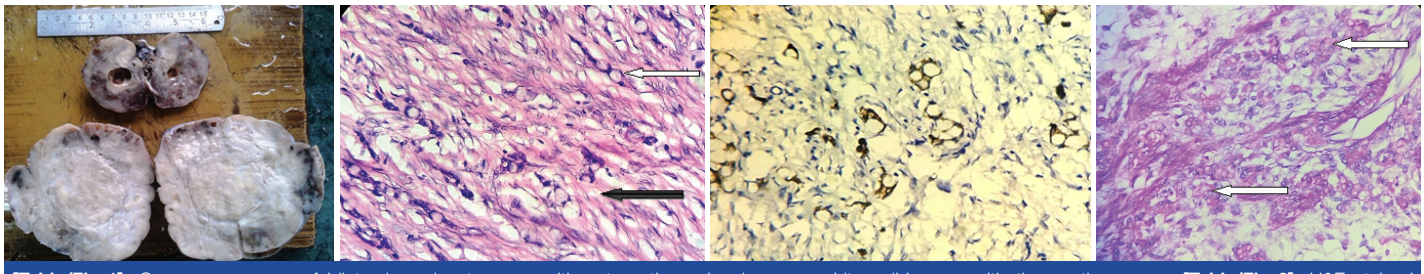
A 37-year-old female presented with pain right hypochondrium with amenorrhea. Serum level of CA-125 was raised up to 133.8 U/ml (normal levels <35 U/ml). Serum CEA levels were normal. Computed tomography examination of lower abdomen and pelvic cavity showed homogenously enhancing soft tissue density lesion suggestive of malignancy. Cytological smears from peritoneal fluid showed only neutrophils against the proteincaeous background, there was no evidence of atypical cells. Specimen of bilateral ovaries with fallopian tubes was grossed. One large ovarian mass measuring 12x9.5x9cm and small ovarian mass measuring 6x4x3cm in size were identified. External surfaces of both the ovaries were smooth. Cut surfaces of both the ovarian tumours were solid [Table/Fig-1] with multiple tiny cystic areas filled with clear fluid. Histopathological examination on light microscopy revealed clear cell type of epithelial tumour cells with abundant fibrous tissue stroma [Table/Fig-2]. On Immunohistochemistry (IHC), tumour cells showed positivity with pancytokeratin. So epithelial origin of tumour cells was confirmed [Table/Fig-3]. Tumour cells also showed cytoplasmic positivity for Periodic Acid Shiff (PAS) staining [Table/Fig-4].

DISCUSSION

The ovarian neoplasms are classified by World Health Histological Organization and are separated according to tissue of origin. The most neoplasms fall under category of tumours of surface epithelium. Surface epithelial tumours are further classified based on differentiation (serous, mucinous and endometrioid mainly) and extent of proliferation of the epithelium [1]. Clear cell adenocarcinoma is 6% of all surface epithelial malignant ovarian tumours. Common age at presentation is 5th-6th decade of life with 10% bilaterality [2,3]. It may arise from glandular epithelium of endocervix or from endometrioid foci [4]. Depending upon stromal component it is divided further as clear cell adenocarcinofibroma and cystadenocarcinofibroma [5,6].

Although ovary is a frequent site of metastases with bilateral involvement in more than 80% of the cases and predominant presentation being cystic mass formation [7,8], but in the current case report patient was 37-year-old with bilateral solid ovarian tumours. In the study carried out among 46 cases of secondary neoplasm ovary, surface involvement by tumour was appreciated in 21 cases followed by surface rupture in 7 cases. The nodular growth pattern and hilar involvement were less frequently seen and uniform solid pattern was not detected at all [8,9]. It is mentioned in the literature that elevated serum CA-125 ovarian tumour with few bowel symptoms can be seen in association with metastases. Careful gross/microscopic assessment of the external ovarian surface and absence of bowel symptoms was helpful to rule out metastatic deposits hence krukenberg tumour, in the present case report [9].

It is considered that clear cell adenocarcinoma is of surface epithelial type. Although in the literature very little is mentioned about adenocarciofibroma of clear cell type. Ookura N and fellows have suggested that adenocarcinofibroma (serous type) had different course and prognosis from carcinoma. Also, due to excess fibrous component, it is difficult to make final diagnosis on clinical as well as radiological grounds. Histopathological examination suggested malignant epithelial cells with clear cell change along with abundant benign fibrous tissue stroma. IHC showed positivity for keratin, consequently epithelial origin was



[Table/Fig-1]: Gross appearance of bilateral ovarian tumours with cut sections showing grey white solid areas with tiny cystic spaces. **[Table/Fig-2]:** H&E stained section (40X) shows clear cells (white arrow) along with stromal component (Black arrow). **[Table/Fig-3]:** Section shows clear cells stained for pancytokeratin (40X). **[Table/Fig-4]:** Section (40x) shows cytoplasmic PAS positivity (white arrow).

confirmed and possibility of mesenchymal origin was ruled out. Furthermore cytoplasmic positivity with PAS stain was also noted which favors the epithelial serous origin of the tumour rather than mucinous type. Keeping in view the efficacy of serum CA 125 tumour marker in relation to IHC, its raised levels confirmed the primary origin and ruled out the possibility of krukenberg tumour [10-13].

In recent years understanding of the underlying pathogenesis and molecular events in the different tumour subtypes has been increased [2,4]. Now there is much more realization that the various subtypes have different behavior and prognosis. So it's very important to classify and diagnose the type of tumour as early as possible [11].

In the emerging era of new chemotherapeutic agents with associated targeted therapies. The histopathological typing of various rare variants of primary ovarian tumours such as clear cell carcinoma, mucinous carcinoma and low grade serous carcinoma is necessary. These types are resistant to traditional chemotherapy with poor clinical outcome. So it is essential to apply the modern approach to ovarian cancer histogenesis and origin to screen, detect and treat these lesions [2].

The present case report suggests that clear cell adenocarcino fibroma is a rare tumour and being different in course and presentation, should be differentiated from other tumours to reach the correct diagnosis.

CONCLUSION

Clear cell adenocarcinofibroma of ovary is a very rare entity. Early recognition of the type is essential for staging and prognosis. Hence histopathology remains the gold standard for identification. Careful gross inspection and histopathological examination with special staining techniques is an excellent approach to reach the definitive diagnosis.

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PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Pathology, Guru Gobind Singh Medical College, Faridkot, India.
2. Professor and Head, Department of Pathology, Guru Gobind Singh Medical College, Faridkot, India.
3. Assistant Professor, Department of Surgery, Guru Gobind Singh Medical College, Faridkot, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Amrit Pal Singh Rana,
218, Medical Campus, Sadiq Road, Faridkot, India.
E-mail: dramritpalsingh@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Jul 23, 2015**
Date of Peer Review: **Oct 08, 2015**
Date of Acceptance: **Jan 31, 2016**
Date of Publishing: **May 01, 2016**