A pure primary transitional cell carcinoma of the ovary: A rare case report with literature review

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Abstract

Primary transitional cell carcinoma (TCC) of the ovary is a rare and recently recognized subtype of ovarian surface epithelial-stromal cancer. Pure forms of the TCC ovary account for only 1% of surface epithelial carcinomas. The clinical presentation is indistinguishable from other types of ovarian cancers. They have a favorable response to chemotherapy than other surface epithelial cancers. We report a case of 55-year-old woman who presented with a hard mass in the abdomen. Computed tomography-diagnosed it as a carcinoma of the ovary. Tumor was immunoreactive with Wilms’ tumor protein-1 and nonreactive with cytokeratin 7 (CK7) and CK20. Histopathology diagnosis of primary TCC of the ovary was made. These tumors are needed to be differentiated from metastatic TCC from other sites and undifferentiated carcinomas of ovaries. Clinical features and immunohistochemistry are helpful. Surgical resection is the primary therapeutic approach followed by standardized chemotherapy.

Key words: Cytokeratin 20, cytokeratin 7, ovary, transitional cell carcinoma, Wilms’ tumor protein-1

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Introduction

A primary transitional cell carcinoma (TCC) of the ovary is rare and recently recognized subtype of ovarian surface epithelial-stromal cancer.[1-4] Criteria for diagnosis and especially spectrum of its morphology are not clearly established because studies of its morphology are few.[5] The clinical presentation is indistinguishable from other types of ovarian carcinoma. The recognition of such tumor is important because of a favorable response to chemotherapy than other surface epithelial cancers.[6]

Case Report

A 55-year-old postmenopausal woman presented with abdominal distension since 2 months. She gave a history of pain in abdomen, dyspepsia, and generalized weakness. Physical examination revealed firm to hard abdominal mass (15 cm × 12 cm) with an irregular surface and restricted mobility. Free fluid was absent. In vaginal examination, the hard mass was felt through all fornices, more on the right side and infiltrated the anterior rectal wall.

Computed tomography (CT) scan of abdomen and pelvis showed a well-defined, lobulated mass (10.7 cm × 10.4 cm × 12 cm) in the right pelvis with heterogeneous enhancement. Mass was extending superiorly in the umbilical region and posterior to the urinary bladder. There was no calcification. Mass effect on adjoining anterior wall of rectum was seen. Other abdominal and pelvic organs were normal except hydronephrosis of the right kidney. There were no lymph nodes enlarged. Amongst other laboratory investigations, CA-125 levels were raised. A clinical diagnosis of carcinoma of right ovary was made.

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Surgical specimens of tumor mass and uterus, cervix, left fallopian tube with ovary and a segment of large intestine (9 cm) were received separately. Tumor mass (13 cm × 12 cm × 10 cm) had lobulated external surface; the cut section was white mostly solid with few cystic areas [Figure 1a]. Large intestinal wall was infiltrated by whitish tumor at places and mucosa was normal.

Microscopy of the tumor sections showed elongated bands of pseudostratified neoplastic epithelial cells resembling transitional epithelium separated by the variable amount of necrotic material and fibrovascular tissue [Figure 1b]. Papillae with fibrovascular core lined by similar neoplastic cells were seen [Figure 1c]. Tumor cells arranged in solid sheets, traversed by delicate fibrovascular septa were seen [Figure 1d]. Numerous well-demarcated, small, and empty micro spaces were seen in the bands of the neoplastic epithelium [Figure 1e]. Tumor cells were relatively monomorphic with a round to oblong nuclei and many showing nucleoli. Cytoplasm was moderate, granular, or vacuolated. Occasional nuclei showed nuclear grooves. 8–10 mitotic figures per high power field were seen [Figure 1f]. On immunohistochemical (IHC) staining, tumor cells were cytokeratin 7 (CK7), CK20 negative [Figure 2a and b] and Wilms’ tumor protein-1 (WT1) positive [Figure 2c]. A diagnosis of pure primary TCC of right ovary was made.

**DISCUSSION**

Primary TCC of the ovary was first described by Austin and Norris in which definite urothelial features are present but no benign, metaplastic and/or proliferating Brenner tumor can be identified.\[7,8\] It arises from pluripotent surface epithelium of ovary and forms cells with urothelial potential. Pure forms of the TCC ovary account for only 1% of surface epithelial carcinomas, mixed carcinomas with a predominant TCC for 5% and those with minor TCC component for 3%.\[3\]

Much of the interest in TCC of ovary has been focused on whether it has independent prognostic or therapeutic significance while others focused on its IHC profile.\[3\] TCC has various histological and IHC patterns.\[3\] Although its morphologic features have been described, the overall literature on this aspect is limited.\[3,5,8‑11\] It has a better prognosis than all other types of ovarian carcinoma following standardized chemotherapy.\[5\] Some investigators believe, variation in the interpretation of morphologic features is one possible explanation for better prognosis and response to chemotherapy than other ovarian carcinomas.\[3,10\]

Clinical presentation of TCC is indistinguishable from other types of ovarian carcinomas. It is important to recognize TCC because it is uncommon; there is some overlap in its morphology with that of the other surface epithelial carcinomas and improved patient survival after a favorable response to chemotherapy.

The 1999 (World Health Organization) described TCC as an invasive tumor that lacks a component of benign Brenner tumor and is characterized by the presence of papillae lined by malignant cells of transitional cell type or nests of such cells in the fibrous stroma.\[5\]

Silva et al.\[3\] suggested “thick papillary proliferations, a smooth luminal border and projection into empty spaces” are the requirements for a diagnosis of TCC. Another less common but distinctive feature they observed was the presence of micro spaces in neoplastic epithelium.

Eichhorn and Young\[5\] studied 100 cases of ovarian carcinomas with transitional cell component. The most frequent histological...
features observed were, undulating thick bands of cells lining the cysts (93%) followed by micro spaces (87%) large cystic spaces (73%), large blunt papillae (63%), necrosis (57%), slit-like fenestrations (49%), bizarre giant cells (35%), small filiform papillae (18%), gland-like spaces (17%), squamous differentiation (13%), and psammoma bodies (4%).

In our case, bizarre giant cells, gland-like spaces, squamous differentiation, and psammoma bodies were not seen. Both undulating and diffuse patterns were seen in our case. Tumor was infiltrating the wall of rectum. Features observed by Eichhorn and Young[3] such as large eosinophilic nucleoli (69%) and longitudinal grooves (48%) were not seen prominently in our case.

Metastatic TCC from the bladder should be differentiated from primary ovarian TCC for appropriate treatment choice which is usually facilitated by clinical findings.[3] Similar observations were made in our study. IHC staining may be also helpful.

In IHC studies, authors emphasized: (1) The presence of CK7/CK20 always indicate a urinary tract origin, (2) Ovarian TCC are negative for CK20, thrombomodulin, and uroplakin, Vimentin, CA-125, WT1 are positive in primary ovarian TCC.[2,12,13] Tazi et al.[7] found primary ovarian TCC was positive for CK7, CA-125, and negative for CK20. In our case, the tumor was immunoreactive for WT1 and nonreactive for CK7 and CK20.

When the tumor has extensive slit-like manifestations and giant cells, a problem can occur in differentiating from poorly differentiated serous carcinoma and undifferentiated carcinoma. Neoplastic cells recognizable as transitional cells, papillae, and thick bands of neoplastic cells of the same type and micro spaces favor the diagnosis of primary TCC of the ovary. The absence of benign or borderline Brenner elements ruled out malignant Brenner tumor.

Higher survival rates can be achieved by surgical resection followed by cisplatin-based chemotherapy. After surgery, the patient was referred to oncology center for further treatment.

**Conclusion**

Pure primary TCC is rare. Clinical presentation is indistinguishable from other types of ovarian carcinomas. Neoplastic epithelial cells recognizable as transitional epithelium arranged in well-demarcated bands from the cystic spaces, papillary projections, and micro spaces within neoplastic epithelium are important histological features. Clinical features and IHC features are helpful in differentiating from metastatic TCC. Surgical resection is the primary therapeutic approach followed by standardized chemotherapy.

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**Conflicts of interest**

There are no conflicts of interest.

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