Case Report

Malignant Mixed Epithelial Tumour of Ovary-Serous Papillary Cystadenocarcinoma and Transitional Cell Carcinoma with Tubo-Ovarian Torsion: A Rare Tumour with Rare Presentation

Shashikant Adlekha¹, Tandra Chadha²

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- Department of Pathology, Sree Narayana Institute of Medical Sciences, Ernakulam-483594, Kerala, India
- ² Department of Microbiology, Sree Narayana Institute of Medical Sciences, Ernakulam-483594, Kerala, India

Abstract

Ovarian torsion can be associated with various pathophysiological factors. Most commonly, benign epithelial ovarian tumours present with torsion. We present an unusual case of mixed malignant epithelial ovarian tumour with a predominant component of high-grade serous cystadenocarcinoma (85%) and transitional cell carcinoma (TCC) (15%) in a patient who presented with acute lower abdomen. The tumour was associated with tubo-ovarian torsion.

Keywords: malignant epithelial neoplasms, torsion abnormality, acute abdomen, serous carcinoma, transitional cell carcinoma

Introduction

Ovarian torsion is one of the significant causes of acute lower abdomen in women. The clinical presentation is vague and non-specific and necessitates urgent diagnosis and surgical intervention to save the adnexa from getting infarcted. Ovarian tumours, more commonly benign and rarely malignant, are implicated in torsion. Mixed epithelial tumours account for less than 4% (reported incidence varies from 0.5% to 4%) of all ovarian epithelial tumours, and incidence of mixed malignant epithelial tumour is much rarer (1). As the behaviour of the tumour depends on the predominant component in mixed malignant tumours, it is imperative to rule out a mixed carcinoma in the ovarian neoplasm specimen by sectioning multiple areas and evaluating them microscopically.

Case Report

A 55-year-old post-menopausal female presented with sudden onset acute lower abdominal pain. Greyscale and colour Doppler ultrasonography of the abdomen showed a "whirlpool appearance" and suggested left tubo-ovarian torsion with no evidence of enlarged lymph nodes or any lesion in the liver, kidney,

or bladder. Emergency surgery was performed and the left tubo-ovarian mass was sent for histopathological examination. The left ovary measured $6 \times 2.5 \times 1$ cm and showed marked haemorrhage and infarction on its surface. On sectioning the ovary, a unilocular cyst, with the major portion infarcted, and a grey-white solid area (Figure 1) were seen amidst the infarcted area. The solid area did not show any papillary processes. Multiple sections were taken from the grey-white solid areas of the ovary. The fallopian tube measured 4 cm in length and was grossly infarcted over the majority of its area. Sections (Figure 2,3) from the infarcted portion of the fallopian tube and ovary and the grey-white solid areas of the ovary showed the tumour to be a mixed malignant epithelial ovarian tumour (highgrade serous and high-grade transitional cell carcinoma (TCC), associated with tubo-ovarian torsion. The serous carcinoma component showed fine and irregularly branching papillae lined with malignant cuboidal to columnar epithelial cells of high nuclear grade and presence of mitoses. Occasional slit-like or irregular glands and solid components of serous adenocarcinoma were also seen. The TCC component showed undulating bands, a diffuse pattern, and occasional broad

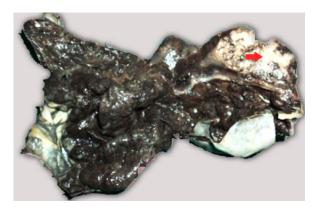


Figure 1: Cut open specimen of ovary showing marked degree of infarction and focal grey white solid area (red arrow).

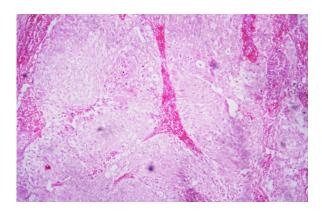


Figure 2: Section showing closely packed malignant transitional cell nests (Haematoxylin and eosin, 100× magnification).

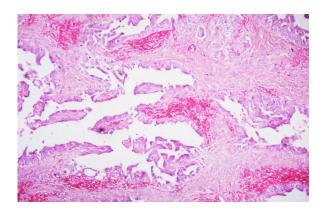


Figure 3: Section showing a cystic cavity lined by proliferating papillae and micro papillae of serous adenocarcinoma with invasion of stroma by solid nests and glandular structures-(Haematoxylin and eosin, 100× magnification).

papillae lined with cells with clear to oxyphilic cytoplasm, a marked degree of anisonucleosis, and prominent nucleoli in many of the cells. Serous carcinoma accounted for 85% and TCC accounted for 15% of the mixed epithelial ovarian tumour. Immunohistochemical studies of the sections revealed positive staining for CK7, WT1, and CA125, and negative staining for CK20, thus ruling out metastasis from extra-ovarian urothelial tumours. Benign transitional cell nests were not seen. Psammoma bodies were seen in a few sections. The patient was referred to the oncology unit for further management.

Discussion

Mixed epithelial ovarian tumours, by definition, are composed of admixtures of two or more of the five major cell types: serous, mucinous, endometrioid, clear cell, and Brenner/transitional. According to the World Health Organisation, the minor component must account for at least 10% of the tumour in a microscopic examination in order to be classified as a mixed tumour (1).

The origin of mixed epithelial ovarian tumours has been controversial. Kurman et al. (2) and Malpica et al. (3) have stated that ovarian cancers are of de novo origin. They divided ovarian cancers into type I and type II, based on their clinical behaviour. Type I includes lowgrade endometrioid, clear cell, mucinous, and transitional carcinomas, which behave in an indolent manner and are confined to the ovary at the time of presentation. Type II tumours are highly aggressive, evolve rapidly, and present in advanced stages. They include high-grade serous carcinomas, undifferentiated carcinomas, and malignant mesodermal mixed tumours (carcinosarcoma). The frequent combinations encountered in mixed epithelial tumours include serous/endometrioid and serous/TCC types (4-7). The least differentiated component determines the tumour grade and the dominant cell type generally dictates behaviour (8).

It is quite important to identify the transitional cell component, as tumours with a predominant TCC component respond much better to chemotherapy compared to other surface epithelial tumours. Ovarian TCC is a rare subtype of epithelial ovarian cancer (9). Pure forms of TCC account for only 1% of surface epithelial carcinomas, mixed carcinomas with predominant TCC components account for 5%, and carcinomas with minor TCC components account for 3% (10). It is sometimes very difficult

to distinguish moderate-to-high-grade TCC from poorly differentiated serous carcinoma and undifferentiated carcinoma. TCCs are characterised by undulating thick bands and varied patterns, such as insular, trabecular, and even minor components of undifferentiated carcinoma. TCCs have broad papillae compared to the fine papillae found in serous carcinomas. In addition, TCCs are lined with cells, some of which have the transitional cell characteristics of pale and granular or oxyphilic cytoplasm, round to oblong nuclei with a single nucleolus or a longitudinal nuclear groove, and the presence of undulating bands lined with these cells. Immunostaining does not help to distinguish between TCC and poorly differentiated serous carcinomas, as TCC stains positively for all serous immunomarkers, such as CK7, CA125, and WT1, and negatively for all urothelial markers, such as uroplakin, thrombomodulin, and CK20. TCC also needs to be differentiated from the more common malignant Brenner tumour by the absence of benign Brenner tumour components/benign transitional cell nest and stromal calcification, compared to the presence of these components in malignant Brenner tumours.

Ovarian torsion usually occurs unilaterally in a pathologically enlarged ovary, and it is more common on the right side (60%). Torsion can involve the ovary alone, but it more commonly affects the ovary and oviduct (adnexal torsion). It frequently arises from one of many anatomic changes. Torsion is associated with ovarian tumour in 50-60% of cases. Dermoid tumours are the most common benign tumours associated with torsion. Malignant tumours are much less likely to be associated with torsion compared to benign tumours (11,12) due to the presence of cancerous adhesions that fix the ovary to surrounding tissue. Adnexal torsion is not limited to women of reproductive age (13); postmenopausal women with an adnexal mass may be affected with torsion. Approximately 17% of ovarian torsion patients present with sudden onset of severe unilateral lower abdominal pain that worsens intermittently over many hours. Early diagnosis of ovarian (adnexal) torsion is imperative, and colour Doppler ultrasonography has a vital role in the examination of women with lower abdominal and pelvic pain (14,15).

This case showed a varied and rare presentation of the much rarer surface epithelial ovarian tumour.

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Conflict of Interest

None.

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Authors' Contributions

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Correspondence

Dr Shashikant Adlekha MD Pathology (RDVV), DNB Pathology (National Board, New Delhi) Department of Pathology Sree Narayana Institute of Medical Sciences North Kuthiyatode, Chalakka-683594 District: Ernakulam Kerala, India

Tel: +9172-9305 2109 Fax: +048-4361 5523

Email: shuntyad@yahoo.co.in

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