A Case Report of Ovarian Carcinoid Complicated with Mucinous Cystadenoma and Review of Relevant Literature

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Abstract: Ovarian mucinous cystadenoma is a common tumor of the female reproductive organs, but ovarian carcinoid is very rare, accounting for about 0.1% of all ovarian tumors [1]. A case of ovarian carcinoid complicated with mucinous cystadenoma was reported. The clinical manifestations, histological features, diagnosis, treatment and prognosis were discussed.

Keywords: Ovarian Carcinoid; Carcinoid Syndrome; Mucinous Cystadenoma

1. Case report

The patient is a 45-year-old female. Admitted to hospital because of "heavy menstruation with progressive dysmenorrhea for two years". The ultrasound showed cystic mass in the bilateral adnexal area (about 4.6x3.3cm on the left and 7.6x4.8x7.1cm on the right). During the operation, the specimen was examined and resectomized. A small nodular tissue was found inside the left ovarian cyst, while the right ovarian cyst was a cyst with smooth cyst wall and no protrusion was found. Left ovarian neoplastic lesions, the final results to be confirmed by paraffin wax routine extensive sampling and immunohistochemistry. Postoperative examination: 1. (left) ovarian carcinoid complicated with mucinous cystadenoma; Immunohistochemistry showed carcinoid components (214610-2#) : PCK (+), CK7 (+), CK20 (-), the vera.ttf - 1 (+), Tg (+), CgA (+) oven, Syn (+), CD56 (+), Caretenin (-), inhibin (-), PAX - 8 + (part), Ki - 67 (LI: 3%), PAS staining mucous. 2. (Right) serous cystadenoma of ovary.

2. Discussion

Carcinoids are neuroendocrine tumors with specific histological, biological, and clinical features. Although they are most commonly found in the gastrointestinal and bronchopulmonary systems, a significant proportion of these tumors can occur in less common anatomical localisation, which can result in tumors remaining undetected until they have metastasized or exhibit secretory activity [2]. Ovarian carcinoid is a very rare tumor, only 5% of carcinoids are of ovarian origin [3]. Due to its low incidence, there is little literature on case reports or describing primary ovarian carcinoid.

Clinical Features of ovarian carcinoid Among reported cases of ovarian carcinoid, patients ranged in age from 31 to 83 years old, but most patients were postmenopausal or perimenopausal [4]. The incidence of ovarian carcinoid is mostly unilateral, some of them are solid masses, or they are often combined with benign teratoma or mucinous tumor. Some people believe that the clinical symptoms of carcinoid cancer are related to the size of the tumor [3]. Most of the patients had no obvious symptoms, and a few had abdominal discomfort or abdominal pain due to large tumor growth and pelvic cavity pressure.

Histopathological features of carcinoid Primary ovarian carcinoid can be divided into four types: island-like, trabecular, mucous and stromal (trabecular carcinoid mixed with thyroid tissue) [9]. ① The most common is the island type, which is characterized by small acinar and polygonal cell nests with round or oval nuclei and uniform chromatin distribution. ②

Trabecular type is rare, and the cells are arranged in trabeculae or bands [10]. (3) The mucinous type, also known as adenoid or goblet cell adenocarcinoma, is morphically similar to the goblet cell adenocarcinoma of the appendix. The glands are dense or sieve, nest or sheet, may be accompanied by necrosis and goiter, and the mucous component may be malignant and contain signet ring cells. The stromal type consists of different proportions of carcinoid components and goiter components, among which carcinoid components are mostly banded or trabecular. Ovarian carcinoids, like other neuroendocrine tumors, also express one or more neuroendocrine markers: chromogranin, synaptophysin, or CD56 [9].

Diagnosis of ovarian carcinoid Several imaging techniques such as ultrasound, CT, magnetic resonance imaging (MRI) and methoxyguanidine can be used to diagnose carcinoid. Other imaging techniques, such as positron emission tomography (pet), can also be of great help in determining the stage of the disease, discovering hidden tumors, and determining treatment options [12]. However, the imaging findings of ovarian tumors are not well described in the literature reported so far. Preoperative diagnosis of ovarian carcinoid is difficult due to its low incidence and lack of typical clinical manifestations and sensitive test methods. As ovarian carcinoid tumors are solid tumors, there is no significant difference between them and solid primary ovarian cancer or metastatic tumors in imaging [13]. At present, the diagnosis is mainly determined by postoperative pathological examination, and no definitive preoperative diagnosis has been reported in the existing literature. To date, there is no consensus on the formal staging of ovarian carcinoid cancer, instead the International Federation of Obstetrics and Gynecology (FIGO) ovarian cancer staging is used [14-15].

Treatment of ovarian carcinoid Most ovarian carcinoids are localized to the ovaries and are at an early stage, and can usually be cured by surgical removal alone. The standard treatment for primary ovarian carcinoid cancer is to remove it completely by oophorectomy or salpingo-oophorectomy. Adjuvant chemotherapy or radiotherapy is not usually recommended or required. Primary ovarian carcinoid has a low degree of malignancy and is generally considered a benign disease if it is confined to one ovary. The 10-year survival rate is close to 100%. For advanced patients, the 5-year survival rate is close to 33% [3]. Unlike primary carcinoids, metastatic carcinoids are more aggressive, with 1/3 of patients dying within 1 year of initial diagnosis and 3/4 dying within 5 years [4]. Therefore, it is of great significance to distinguish metastatic carcinoid from primary carcinoid. Metastatic carcinoid is mostly bilateral, while primary ovarian carcinoid is usually unilateral [3]. Macroscopically, metastatic carcinoid consists of tumor nodules, while primary ovarian carcinoid forms a single homogeneous mass, and the presence of other teratoma elements associated with ovarian carcinoid confirms that it is ovarian primary. Although island-like carcinoids of the ovary are considered malignant, they grow slowly and are rarely associated with metastasis, which in at least 90% of cases presents as extra-ovarian disease [4]. For patients with abdominal recurrence after initial surgery, there is a lack of strong evidence related to treatment due to the small number of cases reported in the existing literature.

Prognosis of ovarian carcinoid In patients with carcinoid, the appearance of carcinoid syndrome is associated with length of survival. The median survival time of patients is 3.5-8.5 years [19]. In two different studies, the 5-year survival rate after symptom onset was 30% and 67%, respectively . Obviously, the prognosis of patients with carcinoid cancer varies greatly. Ki-67, based on the proliferation rate of tumor cells, is now routinely used as a grading system for neuroendocrine tumors, and is widely considered to be a marker of tumor aggressiveness and can be used to predict the prognostic behavior of tumor biology. Therefore, more research is needed on clinical and molecular predictors that affect patient prognosis.

3. Conclusion

To sum up, great progress has been made in the treatment of ovarian carcinoid in the past decades, but there is no effective treatment for ovarian carcinoid with distant metastasis and metastatic carcinoid, and there is no reliable preoperative diagnosis method for ovarian carcinoid in clinical practice. Carcinoid cancer is relatively rare, so more and better clinical screening indicators, biological and imaging indicators are needed to monitor all stages of the disease, so as to achieve early detection, early diagnosis and treatment as possible.

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