Gokhan et al., J Clin Case Rep 2012, 2:16
DOI: 10.4172/2165-7920.1000227

Journal of Clinical Case Reports

Case Report Open Access

A Primary Insular Type Carcinoid Tumor Arising in a Mature Cystic Teratoma of the Ovary: A Case Report

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Abstract

Background: Carcinoid tumors are rare neoplasms of the diffuse peripheral neuroendocrine system which produce biological amines and various peptides. Carcinoid tumors most commonly occur in the gastrointestinal tract. Primary ovarian carcinoids are rarely seen; they consist less than 0.1% of all ovarian cancers and 0.3% of all carcinoid tumors.

Case: Here, we present a rare case of primary carcinoid tumor, insular type, arising in a mature cystic teratoma of the ovary.

Conclusion: Surgical excision of the tumor is usually sufficient treatment for most of the patients.

Keywords: Carcinoid tumors; Mature cystic teratoma; Insular type carcinoid tumors

Introduction

Carcinoid tumors are rare neoplasms of the diffuse peripheral neuroendocrine system which produce biological amines and various peptides [1]. Carcinoid tumors most commonly occur in the gastrointestinal tract. The second most common place where carcinoid tumors may appear is the lungs and also ovarian carcinoid tumors may be primary or metastatic [2]. Primary ovarian carcinoids are rarely seen; they consist less than 0.1% of all ovarian cancers and 0.3% of all carcinoid tumors. They are usually unilateral and mostly seen in the perimenopausal and postmenopausal women. Generally, they occur in association with mature cystic teratomas or mucinous tumors [3]. There are four histological types of primary ovarian carcinoid tumors; insular, trabecular, strumal and mucinous. The most common type is insular [4]. Here, we present a rare case of primary carcinoid tumor, insular type, arising in a mature cystic teratoma of the ovary.

Case Report

A 56 year old woman, gravida 3, para 3, visited the gynecology clinic with the complaint of lower abdominal pain. The patient was menopausal for the last 6 years and had no prior surgery. No pathologic sign was found during her physical examination. The transvaginal ultrasound revealed a right adnexial lobulated ovarian mass measuring 71×66 ×61 mm, which was solid with occasional internal scattered fluid areas. No free fluid was seen in abdominal or pelvic cavity.

The patient's serum CA125, CA19-9 and CEA levels were within the normal range. The patient was diagnosed with postmenopausal complicated adnexal mass and total abdominal hysterectomy and bilateral salpingo-oophorectomy was performed. During the operation, uterus, left fallopian tube and left ovary were observed normal. A mobile, solid, smooth surfaced, 6-7 cm right ovarian mass was detected and sent to frozen section. The result of frozen section was reported as mature cystic teratoma.

The right ovary was 6.5×6×6 cm in size; the cut surface was cystic with hair structures and keratinous material. Microscopical examination revealed cartilage, mature bone, epithelial structures (respiratory and squamous), mucinous glands together with mature neuronal elements (Figure 1). Among these structures, incidentally a small microscopical focus of monomorphic cells was seen. These cells were arranged in round nests, had round uniform nuclei with granular/salt & pepper chromatin. No mitoses were seen. Immunohistochemical studies showed diffuse synaptophysin and chromogranin positivity (Figure 2), while Ki-67 proliferation index was below 1% (not shown). The lesion was a carcinoid tumor of insular type arising from a mature cystic teratoma. The right ovary was totally submitted for microscopical examination. The left ovary was normal.

The patient had not showed carcinoid syndrome symptoms and the laboratory study for serotonin, vasoactive intestinal peptide and 24 hours urine 5-hydroxy-indole acetic acid levels were normal. The patient had no clinical symptoms and the laboratory studies showed no abnormal value in the follow up period.

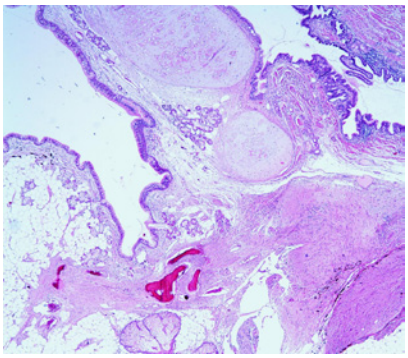

Figure 1: Cartilage, bone, respiratory epithelium, several glanular structures and neural tissue (lower right) are readily recognizable in the mature cystic teratoma (H-E, X4).

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Received October 14, 2012; **Accepted** November 22, 2012; **Published** November 24, 2012

Citation: Gokhan B, İlker S, Gunes G, Alp U, Serdar GG (2012) A Primary Insular Type Carcinoid Tumor Arising in a Mature Cystic Teratoma of the Ovary: A Case Report. J Clin Case Rep 2:227. doi:10.4172/2165-7920.1000227

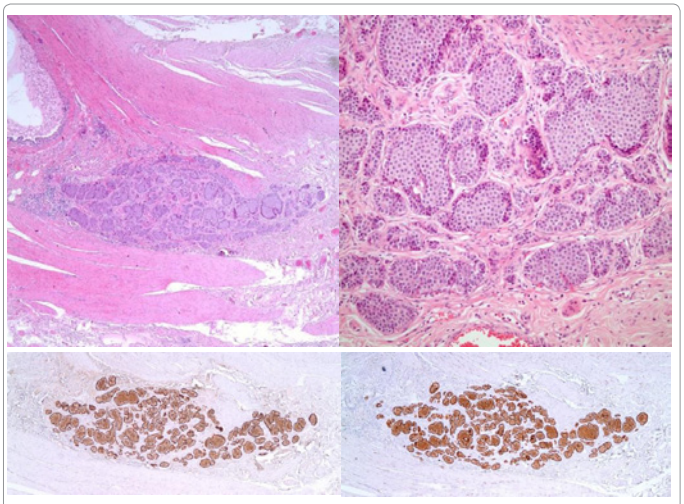

Figure 2: A microscopic focus of nests (upper left) (H-E, X4) consisting of polygonal, uniform cells (upper right) (H-E, X20) are seen. Immunohistochemical studies show diffuse cytoplasmic chromogranin (lower left, X4) and synaptophysin (lower right, X4) positivity.

Discussion

Ovarian carcinoid tumors originate from the totipotent cells of benign teratomas and found in association with these tumours. These tumors have low malignant potential. The histological diagnosis of carcinoid tumours depends on immunohistochemical identification of chromogranin a granules, enterochromaffin, argyrophil and argentaffin cells. There are four histological types of primary carcinoid tumors; insular, trabecular, mucinous and strumal. The most common type of primary ovarian carcinoid tumor is insular [1]. The most aggressive type of primary ovarian carcinoid tumor is mucinous. The insular carcinoid tumor could be confused with granulosa cell tumor, sertoli-leydig cell tumor, brenner tumor and metastatic carcinoids [1]. These tumors should be considered in the differential diagnosis.

Primary ovarian carcinoids should be distinguished from metastatic carcinoids. Nearly all primary ovarian carcinoids are unilateral. Metastatic ovarian carcinoids are usually bilateral and the most common primary origin is the gastrointestinal system. Macroscopically, the tumor consists of multiple nodules in metastatic ovarian carcinoids and a single mass in the primary carcinoids. Teratoma formation also supports the tumor as a primary focus [5]. The average age of the patients who were diagnosed with primary ovarian carcinoids is 57 (with the range of 44-77) [5]. Diagnosis of primary ovarian carcinoid tumor for our patient was supported with the facts that the age of the patient was 56, she was menopausal for 6 years, and also with the pathology result showing carcinoid tumor was unilateral with underlying mature cystic teratoma.

Most of the ovarian carcinoid tumors are asymptomatic. Carcinoid syndrome, which is characterized with flushing, abdominal pain, diarrhea, and pulmonary and cardiovascular changes, occurs only in 1/3 of the women with primary ovarian carcinoid tumor [6]. The presented case didn't have the clinical signs of carcinoid syndrome.

The treatment for ovarian carcinoid tumors is surgical excision [2]. Surgical staging is not required for the carcinoid tumors. If the patient is perimenopausal or postmenopausal, total abdominal hysterectomy and bilateral salpingo-oophorectomy should be performed. If the patient is young; unilateral or bilateral salpingo-oophorectomy should be performed [1]. According to the literature, if the disease is limited to one ovary, the prognosis is excellent in primary ovarian carcinoid tumors and ten year survival rates are approximately 100%. However, if the disease is in advanced stage, five year survival rates are approximately 33% [7]. Our patient was postmenopausal and the disease was limited to one ovary so we performed total abdominal hysterectomy and bilateral salpingo-oophorectomy with no surgical staging.

Conclusion

In conclusion, primary ovarian carcinoid tumors are rare neoplasms of the ovary and they are associated with good clinical results [2]. Surgical excision of the tumor is usually sufficient treatment for most of the patients [8]. In contrast to primary ovarian carcinoid tumors, metastatic carcinoid tumors behave more aggressively and 33% of the patients die within the first year of the diagnosis; 75% of the patients die within 5 years [9].

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