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Case Report

Primary ovarian carcinoid tumor: a report of 4 cases

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ABSTRACT

Ovarian carcinoid tumor is a rare neoplasm that account for 0.3% of all ovarian tumors. It is commonly seen in perimenopausal and postmenopausal women. The aim of this study is to investigate the clinical features and to evaluate the treatment of this rare tumor. The study reviewed retrospectively the clinical and pathological findings of 4 cases of primary OCT treated in Salah Azaiez Institute between 1994 and 2012. The median age was 50 years (ranging from 28 to 75 years). All the patients underwent radical surgery. Two patients had carcinoid tumors occurring in mature cystic teratoma. Three patients had stage I diseases, and one patient had stage III disease, this patient had adjuvant chemotherapy. The follow up median was 23 months. There was one case of bone and lung metastasis diagnosed in the patient who had stage III disease, fourteen months after the surgery but the patient died of her disease four month later. The other patients were alive without evidence of disease recurrence. Primary Ovarian carcinoid tumors are most of the time associated with good clinical results. Surgical excision of the tumor is usually sufficient treatment for most of the patients

Keywords: Carcinoid, Ovary, Primary, Teratoma

INTRODUCTION

Carcinoid tumors are neuroendocrine tumors arising from the embryologic primitive gut derivation. A carcinoid tumor usually appears in the gastrointestinal tract.[1] Primary ovarian carcinoid tumors (OCTs) are uncommon. The majority of them are associated with mature cystic teratomas. Most tumors are seen in peri- or postmenopausal women with symptoms of enlarging mass, or are incidental findings. These tumors are divided into four main types: insular type, trabecular type, strumal type and mucinous type.

CASE REPORT

The median age was 50 years (ranging from 28 to 75 years). Two patients complained of abdominal pain, one patient complained of dysuria and one patient presented acute peritonitis. On pelvic examination, there were a pelvic mass in three cases that were also revealed on ultrasonic examination. All the patients underwent radical surgery. Two patients had lombo-aortic lymphadenectomy and there was a lymphatic involvement in one case of them. At operation, three cases were stage I, and one case was stage III.

The tumors ranged from 3 cm to 16 cm in diameter (average 9.7 cm). Two patients had carcinoid tumors occurring in mature cystic teratoma, and there was a mature cystic teratoma in the controlateral ovary in one case. Histologically, there were two insular OCTs, one trabecular and one mucinous OCT. The patient who had stage III disease received adjuvant chemotherapy (CT)

with tree cycles of PVeBV regimen (Etoposide-Cisplatin- Vinblastine- Bleomycin).

The follow up median was 23 months (ranging from 4 to 47months). There was one case of bone and lung metastasis diagnosed in the patient who had stage III disease, fourteen months after the surgery. The metastases were treated by CT (BEP: Bleomycin-Etoposide-Cisplatin) but the patient was died of her disease four months later. The other patients were alive without evidence of disease recurrence. The clinicopathological characteristics and the treatment outcome related to the four cases were recapitulated in Table1.

Table 1: Clinicopathological characteristics and treatment outcome.

	Case 1	Case2	Case3	Case4
Age (years)	50	28	47	75
FIGO Stage	I	III	I	I
Tumor size (cm)	10	16	3	10
Histological Subtype	insular	mucinous	trabecular	insular
Treatment	RS	RS+CT	RS	RS
Outcome	Favorable	Died (16 months after surgery)	Favorable	Favorable
Follow-up (months)	47	16	3	26

DISCUSSION

Ovarian carcinoid tumors constitute only 0.3% of all ovarian carcinomas.[1] They account for 0.5% of all carcinoid tumors. They may remain undiagnosed until the time of surgery. Carcinoid tumor can be seen in the ovary as a metastasis of a primary tumor located in gastrointestinal tract or elsewhere.[2-3] Insular carcinoid is the most common type of primary OCT.[4] The majority of these tumors occur as a component of mature cystic teratoma, but a considerable number present in pure form. In our report, there were two cases where the tumor was associated with a mature cystic teratoma.[5]

According to the World Health Organization classification, OCTs represent a group of ovarian monodermal teratomas.[6] Most tumors are seen in peri- or postmenopausal women as cystic or solid ovarian mass, generally about 10 cm in maximal dimension. They are virtually always unilateral, but in up to 15% of cases there is also a mature cystic teratoma or mucinous tumor in the contralateral ovary. In this report, a mature cystic teratoma in the control lateral ovary was found in one case. Patients with primary OCTs show symptoms of an abdominal mass, or the tumor is an incidental finding.

The unique complications derived from the secretion of serotonin are known as carcinoid syndrome, characterized by episodic cutaneous flushing, cyanosis, abdominal cramps, diarrhea, carcinoid heart disease, bronchoconstriction.[7]

Women with primary OCTs develop carcinoid syndrome in 43%.[8] There was no case of carinoid syndrome in our report. Even though carcinoids can cause the carcinoid syndrome, with diarrhea as one of the symptoms, several cases of strumal carcinoid have been reported in patients exhibiting severe constipation.[9] Ovarian carcinoids exhibiting estrogenic or androgenic manifestations are extremely rare. In our cases, no one of the patients had hirsutism or estrogenic manifestations. Microscopically, primary OCTs present a variety of patterns, including insular, trabecular, strumal and mucinous.[10]

A mixed type has also been reported, which is composed of any combination of the pure types. Primary ovarian carcinoids also arise in association with mucinous tumors; therefore, a preoperative diagnosis is extremely difficult.[11] The most common histological subtype being insular, followed by trabecular and mucinous.[12]

The histological appearance is similar to that of carcinoid tumor elsewhere. There are tumors with an insular pattern of growth similar to those seen in the appendix and small bowel, tumors with a trabecular appearance similar to that seen in the rectum, and tumors with mucinous appearance similar to those seen primarily in the appendix.[13]

Insular carcinoids are best considered as tumors of low malignant potential and trabecular carcinoids are very infrequently associated with metastases. Clinical course of mucinous carcinoids tends to be more aggressive. The liver is the most common site for carcinoid tumor metastasis. After spreading to the liver, the tumor can metastasize to the lungs, bone, skin, or almost any organ.[14] In the present report the metastasis site was the bone and the lung. No effective treatment exists for OCTs, with the exception of surgical resection. Furthermore, most cases are diagnosed in the early stage and curable by excision of the affected adnexa alone. Bilateral salphingo-oophorectomy and total abdominal hysterectomy are the treatments of choice at pre-and

postmenopausal state, while unilateral salphingo-oophorectomy usually suffices for younger patients.[15] In our report, three patients underwent a radical surgery even though they had a stage I disease, because they had a pre- and postmenopausal state, whereas the patient who was aged 28 years underwent a radical surgery because of her advanced staged disease.

There is no evidence on the benefits of adjuvant therapy or the type of drug

CONCLUSION

Primary OCTs are rare neoplasms of the ovary and they are most of the time associated with good clinical results. It is necessary to know the clinical characteristics of primary ovarian strumal carcinoid tumors to ensure their appropriate diagnosis and management. Surgical excision of the tumor is usually sufficient treatment for most of the patients. In cases with a high risk of recurrence, particularly in patients with carcinoid syndrome at diagnosis, careful follow-up examinations must be continued for an extended period of time.

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