Case Report

Primary ovarian carcinoid in mature cystic teratoma: A rare entity

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ABSTRACT

Primary ovarian carcinoids are rare, accounting for 0.3% of all carcinoid tumors; however, carcinoid tumors metastatic to the ovary are common. A majority of primary ovarian carcinoids occur in association with mature cystic teratoma. In this paper, we present a case of carcinoid tumor developing in a mature cystic teratoma in a 50-year-old female.

Key words: Carcinoid, ovary, teratoma

INTRODUCTION

Carcinoid tumors arising from the reproductive organs, such as primary ovarian carcinoids, are rare and sparsely documented. Stewart *et al.*[1] reported the first case of carcinoid tumor arising in an ovarian teratoma in 1939. Mature cystic teratomas (dermoid cyst) make up almost 20% of all ovarian neoplasms. Although they are almost always benign tumors, the rare development of cancer deserves emphasis.[2] The most common malignant change in a dermoid cyst is squamous cell carcinoma, followed by adenocarcinoma and carcinoid tumor.[3] Primary ovarian carcinoid tumors are uncommon and the majority of them are associated with mature cystic teratomas.[3] Robboy *et al.*[4] divided these tumors into three types: The insular type, trabecular type, and strumal carcinoid type. We report herein a case of trabecular carcinoid tumor arising from a mature cystic teratoma, which was examined by histological and immunohistochemical methods.

CASE REPORT

A 50-year-old woman presented with lower abdominal pain for 1 month. The ultrasonography revealed a heterogeneous mass measuring 7.5 cm × 3.7 cm × 2.4 cm containing echogenic reflectors. CA-125 was within normal limits. The vaginal examination revealed first degree cervical descent, a retroverted uterus and a left adnexal mass. A clinical diagnosis of mature cystic teratoma was kept and hysterectomy with bilateral salpingo-oophorectomy was performed. Grossly the uterus, cervix and right sided ovary were unremarkable. The left sided ovary was converted into a mass measuring 8 cm × 4 cm × 3 cm. The outer surface was smooth gray-white with distended blood vessels. Cut surface revealed solid-cystic areas. The cystic area was filled with pultaceous material along with tuft of hair. The solid area was homogeneous, tan-brown to yellow in color. Microscopic examination revealed a cyst lined by stratified squamous epithelium with skin appendages [Figure 1]. The sub-epithelial zone showed keratin debris surrounded by giant cell reaction to keratin, smooth muscle cells, blood vessels, and chronic inflammatory cell infiltrate. The sections from the solid part revealed tumor cells arranged in acini, tubules, ribbons, cords and trabeculae. The tumor cells had round to oval nuclei with stippled chromatin and granular eosinophilic cytoplasm [Figure 2]. No mitotic figures were identified. On the basis of nuclear features, a neuroendocrine tumor was suggested. Immunohistochemistry was carried out with chromogranin A which showed cytoplasmic positivity. A diagnosis of mature cystic teratoma with primary carcinoid ovary (trabecular type) was given. The patient is kept on radiological surveillance of abdomen and pelvis at 3 month interval.

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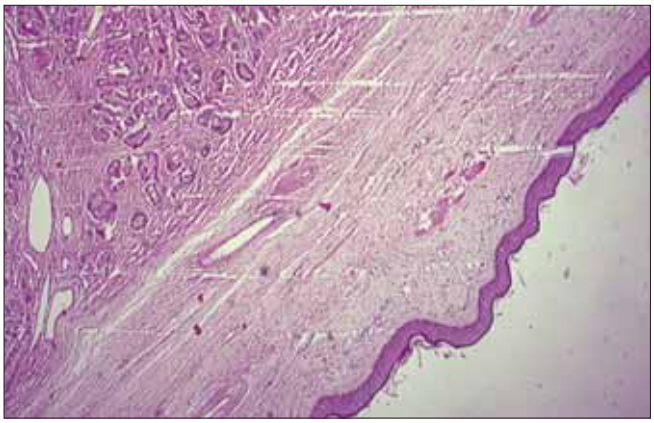

Figure 1: Ovarian carcinoid lined by stratified squamous epithelium component of mature teratoma (H and E, ×10)

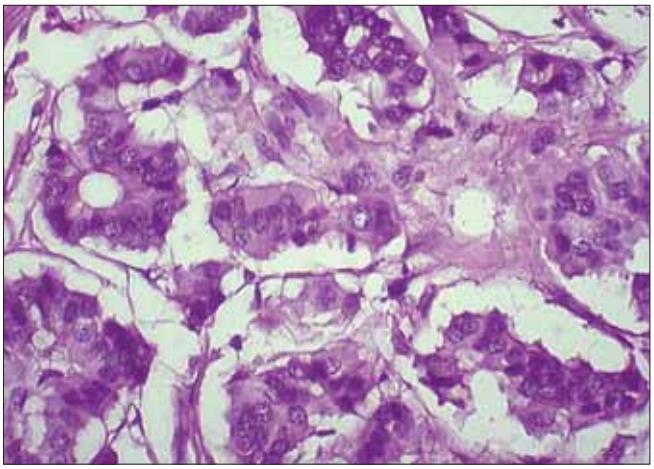

Figure 2: Tumor cells are arranged in acini and trabeculae having round to oval nuclei, stippled chromatin and abundant granular eosinophilic cytoplasm (H and E, ×40)

DISCUSSION

Mature cystic teratomas constitute 15-20% of all ovarian tumors.[2] Although dermoid cysts of the ovary are almost always benign tumors, primary malignancy developing in these cysts rarely encountered about 1.5%, despite the presence of embryonic structures in these neoplasms.[2] The most common malignant change in mature cystic teratoma is squamous cell carcinoma.[3] Carcinoid tumors constitute only 0.3% of all ovarian neoplasms; thus they may remain undiagnosed until the time of surgery.[4,5] Carcinoid tumor can be seen in the ovary as a metastasis of a primary tumor located in gastrointestinal tract or elsewhere,[6] as a component of mature cystic teratoma or as a primary pure neoplasm of this organ. Carcinoids typically arise in the intestine, with the appendix most frequently involved or more rarely from the thymus, bronchus, stomach, or pancreas. The large majority of primary ovarian carcinoids are unilateral, but in 16% of cases the contralateral ovary is involved by a cystic teratoma or a mucinous neoplasm.[4] In contrast, most metastatic carcinoid tumors to the ovary are associated with peritoneal metastasis. The prognosis in primary carcinoid (whether pure or a component of mature cystic teratoma) is very good, whereas metastatic carcinoids have poor outcome.

Carcinoid tumors secrete a wide variety of neurohumoral substances such as serotonin, histamine, tachykinin, bradykinin, kallikrein, corticotrophin, substance P, motilin, and prostaglandins.[7] Persistent systemic exposure to large quantities of these hormones and biogenic amines can result in carcinoid syndrome, the classical triad of flushing of upper extremities and face, wheezing, and diarrhea. Normally, systemic exposure does not occur with an intestinal carcinoid until it has metastasized, because of efficient hepatic metabolism of secreted substances. However, primary ovarian carcinoids can cause these symptoms directly, because their venous drainage bypasses the portal venous system.[4] In our case, the patient did not present with any features of carcinoid syndrome. Microscopically, the appearance is similar to that of carcinoid tumor elsewhere, in that they recapitulate the various patterns of this well-differentiated neuroendocrine tumor as seen in various sites. Thus, 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/goblet cell appearance similar to those seen primarily in the appendix.[8] The similarities also apply to their histochemical features (argentaffinity or argyrophilia depending on the type), and to the consistent presence of neurosecretory granules at electron microscopic level.[9] Chromogranin A, neuron specific enolase, 5-HT and variety of peptide hormones like peptide YY have been demonstrated immunohistochemically, particularly in tumors of trabecular type.[10] It is not possible to separate primary from metastatic carcinoid tumors on morphologic grounds; however, if the ovarian carcinoid is admixed with areas of teratoma, the chances are overwhelming that it is of primary origin. Excision of primary carcinoid of insular or trabecular type is usually curative. The behavior of mucinous type is more aggressive as in appendix.[8]

This case adds to the rare reports in the literature of a carcinoid occurring in a mature cystic teratoma. It also signifies the importance of extensive sampling of the solid parts of a dermoid cyst to increase the data about primary ovarian carcinoids arising in a mature cystic teratoma.

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