Primary insular carcinoid of the ovary

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Summary

Primary ovarian carcinoids are very rare tumors that account for less than 5% of all carcinoids and 0.1% of all ovarian malignancies. We present a rare case of a primary, non-functioning, insular carcinoid of the left ovary in a 44-year-old woman originating from the outer surface of a mature cystic teratoma. After an uneventful unilateral salpingo-oophorectomy, the patient had no sign of recurrence with computed tomography and 5-HIAA evaluation at 3-year follow-up. Although rare, primary ovarian insular carcinoid tumors that are confined to the ovary and treated with surgery are expected to have an excellent overall outcome..

Key words: Primary insular ovarian carcinoid; Mature cystic ovarian teratoma; Ovarian neoplasms; Salpingo-oophorectomy; Non-functioning carcinoid.

Introduction

Primary ovarian carcinoids are very infrequent tumors. They account for less than 5% of all carcinoids and 0.1% of all ovarian malignancies [1, 2]. The clinical carcinoid syndrome has been described in 43% of the insular type and 25% of the mature teratoma-associated insular carcinoids [3, 4]. There is a strong correlation with the size of the neoplasm to the manifestation of the carcinoid syndrome [3]. Although rare, primary ovarian carcinoid tumors treated with surgery alone and found to be confined to the ovary can be expected to have an excellent overall outcome [4]. We present a very rare case of a primary, non-functioning, insular carcinoid of the left ovary originating from the outer surface of a mature cystic teratoma in a 44-year-old woman.

Case Report

A 44-year-old woman with an unremarkable medical history presented to our department with a six-month history of lower abdominal pain and distension. On clinical examination, a non-tender, well-circumscribed mass was noted in the left lower abdomen. Blood tests including neoplasmatic markers such as carbohydrate antigen 19-9 (CA 19-9), carcinoembryonic antigen (CEA), and carbohydrate antigen 125 (CA 125) were normal. Abdominal ultrasound (US) revealed a 10 cm cystic mass in the left ovary and a 4 cm solid mass adjacent to the cyst; these findings were confirmed by contrast-enhanced abdominal computed tomography (CT) and, subsequently, laparotomy. The solid lesion was firmly attached to the cyst. Unilateral salpingo-oophorectomy was performed.

The solid, yellow-tan, lesion measured 4 x 3.5 x 0.4 cm and the cyst 10 x 10 cm. Histopathological examination showed the lesion to be a primary ovarian carcinoid characterized by a well defined insular pattern of uniform cells with round nuclei and abundant chromatin (Figure 1). There was no atypia and no mitosis noted. Fibrous acellular stroma separated the islets of carcinoid cells. The cystic structure was a mature cystic teratoma lined by epidermis with skin appendages (hair follicles). The tumor cells were stained strongly positive to chromogranin A (Figure 2A). In addition, immunopositivity to neuron specific enolase (NSE) and synaptophysin was identified (Figures 2B and C). Consequently, diagnosis of a primary insular ovarian carcinoid originating from the outer surface of a mature cystic teratoma was made.

After an uneventful recovery, the patient was discharged on the fifth postoperative day. After a follow-up period of three years, our patient has had no sign of recurrence with CT and 5-HIAA evaluation.

Discussion

Primary ovarian carcinoids are slow growing malignant neoplasms. They are classified as insular, trabecular, strumal, mucinous, and mixed. Insular carcinoids represent the most common type. They rarely metastasize while they are commonly associated with carcinoid syndrome. Size over 10 cm is well associated with carcinoid syndrome [3].

The differential diagnosis should include several other ovarian tumors such as granulosa cell tumor, Sertoli-Leydig tumor, and Brenner tumor. Immunopositivity to chromogranin A documents the diagnosis [5]. In case the tumor is incidentally found, it should be treated as a germ cell tumor with bilateral salpingo-oophorectomy and hysterectomy or unilateral salpingo-oophorectomy if preservation of fertility is desired [5].

A metastatic carcinoid to the ovary should also be excluded. Thorough investigation for synchronous gastrointestinal or mesenteric masses should be carried out pre- and intraoperatively. A metastatic carcinoid is likely if liver or peritoneal dissemination is identified. Additional clues are the presence of bilateral disease and multiple nodules in contrast to a single solid mass in the primary ovarian carcinoid [6]. The presence of teratoma-

Revised manuscript accepted for publication February 19, 2008

Fig. 1

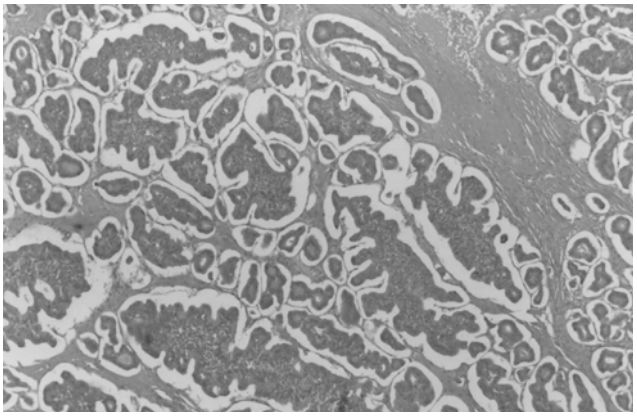

Figure 1. — Nests of round uniform neoplastic cells are separated by an acellular fibrous stroma. No atypia or mitosis is noted (HE x 100).

Fig. 2

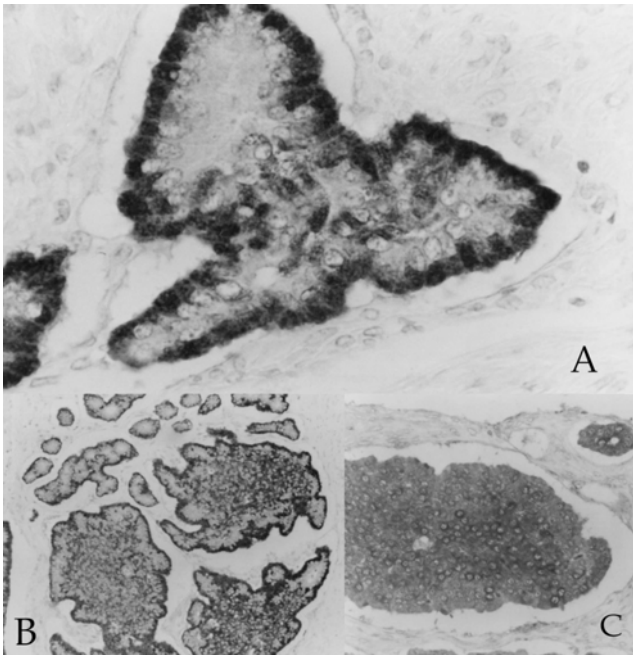

Figure 2. — A) Neoplastic cells exhibiting strong immunopositivity to chromogranin A (x 200). B) Immunopositivity to NSE is identified (x 200). C) Positivity to synaptophysin is also evident (x 200).

tous elements in association with an ovarian carcinoid confirms the primary ovarian origin of the lesion [5].

After a follow-up of three years, our patient has no sign of recurrence of the disease. Survival for patients with primary insular carcinoid of the ovary is excellent with 10-year survival of nearly 100% if the disease is confined to the ovary and treated with surgery [4].

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