Primary carcinoid tumor of the ovary: A case report

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Summary

Primary ovarian carcinoid tumors are rare. A 47-year-old woman presented to our emergency room with lower abdominal pain. Physical examination, pelvic ultrasonographic evaluation and abdominal computed tomography revealed a 10-cm mass in the right ovary containing cystic and solid components, as well as calcifications typical of a dermoid cyst. At laparotomy, a smooth-surfaced, firm and mobile right adnexal mass with solid and cystic portions was detected. Initially, right salpingo-oophorectomy was performed. Frozen-section examination identified the mass as a sex cord stromal tumor containing a mature cystic teratoma. Based on this finding, total abdominal hysterectomy, left salpingo-oophorectomy, omentectomy, appendectomy were performed, and the pelvic-paraaortic lymph nodes were also removed. All histological findings in the right ovary were similar to the features of cystic teratoma and trabecular carcinoid tumor. Examination of the resected lymphatic, omental, and appendiceal tissue indicated no tumoral invasion. The diagnosis was ovarian carcinoid Stage IA. Serum testing post-surgery revealed that the levels of cancer antigen (CA) 19-9 and CA125 were 18.5 u/ml and 10.5 u/ml, respectively. The patient was discharged on postoperative day 5. The report describes the clinicopathologic and immunohistochemical features of a primary ovarian carcinoid that contained a mature cystic teratoma.

Key words: Primary carcinoid; Ovarian carcinoid.

Introduction

Carcinoid tumors are neoplasms of the diffuse peripheral endocrine system, and they produce biogenic amines and various polypeptides. These masses most often arise in the gastrointestinal tract and less frequently in the bronchi, biliary tract, and ovaries [1]. Primary ovarian carcinoid tumors are rare, accounting for less than 0.1% of all ovarian carcinomas [2]. It mostly occurs in women over 50 years of age and constitutes less than 1% of all carcinoid tumors [3]. Ovarian carcinoid tumors are also called "specialized teratomas" due to the fact that they often contain mature or immature teratoid components. Patients with ovarian carcinoid may also present with clinical "carcinoid syndrome". This is especially common for large tumors. Carcinoid syndrome [4] (facial flushing, diarrhea, bronchospasms and edema) has been estimated to occur in one third of patients with ovarian carcinoid tumors [5, 6]. Theoretically, serotonin-like substances are released directly into the systemic circulation through the ovarian venous system, bypassing hepatic deactivation. Ovarian carcinoid tumors are categorized in four groups based on histologic patterns: insular, trabecular, strumal (struma ovari and carcinoid) and mucinous. All types may or may not contain an associated mature teratoma. Over 300 patients have been reported with this disease [7]. Past reports reveal that survival for patients with primary ovarian carcinoid is excellent if disease is confined to one ovary.

In this report, we describe the clinicopathologic and immunohistochemical features of a primary ovarian carcinoid that contained a mature cystic teratoma.

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Case Report

A 47-year-old, gravida 12, para 12 woman presented to our emergency room with lower abdominal pain. Physical examination revealed a smooth, non-painful, 8-10 cm mass in the right anterior pelvis. The mass was separate from the uterus. The patient had no signs or symptoms of carcinoid syndrome (flushing, diarrhea, cardiac murmur, hypertension, pedal edema). Palpation of the uterus and left adnexa revealed nothing abnormal. Pelvic ultrasonographic evaluation demonstrated a 10 cm in diameter mass in the right ovary that contained cystic and solid components. Computed tomography (CT) scanning of the abdomen and pelvis revealed a right adnexal mass that measured 10 cm in the largest diameter. It consisted of solid and cystic components and showed calcifications typical of a dermoid cyst. The CT scan also showed diffuse thickening of the peritoneum and omentum and a small amount of free fluid in the abdomen. Serum testing for tumor markers showed cancer antigen (CA) 125 99.7 u/ml, CA 19-9 534 u/ml, alpha-fetoprotein (AFP) 1.62 u/ml and carcinoembryonic antigen (CEA) 1.7 u/ml. There were no abnormal findings on a chest X-ray, colonoscopy or gastroscopy, and the patient's medical, surgical and family histories were unremarkable.

At laparotomy, we found a smooth-surfaced, firm, mobile, right adnexal mass with solid and cystic components. We also noted approximately 200-300 ml of free fluid in the abdomen and thickened omentum. The left ovary contained a serous cyst that was 4-5 cm at its largest diameter. There were no remarkable findings in the uterus, gastrointestinal tract or upper abdomen.

Initially, we performed a right salpingo-oophorectomy. Frozen section examination revealed a sex cord stromal tumor containing a mature cystic teratoma. Based on these results, we also performed total abdominal hysterectomy, left salpingo-oophorectomy, omentectomy, appendectomy and removed the pelvic-paraaortic lymph nodes. The tumor in the right ovary measured 10 cm at its largest diameter. The cut surface of the tumor showed a solid and multilocular cystic appearance. The

solid area of the tumor was yellowish-brown in color and the cysts contained tufts of hair, yellow grumous material and mucoid fluid. Also firm, white cartilage, gritty bone and intact teeth were observed. Microscopically the tumor composed of sheets and islands of neoplastic cells surrounded by fibrous stroma. The cells had been grown in ribbons in a trabecular pattern in most areas (Figure 1). The cells had regular, round nuclei and moderate amounts of eosinophilic cytoplasm and they immunostained with synaptophysin, cytokeratin, chromogranin-A and neuron specific enolase (NSE) (Figure 2). Adjacent to the carcinoid tumor cystic structures were observed (Figure 3). The inner surface of the each cyst was lined by squamous or columnar epithelium. One of the cysts contained luminal mucin and numerous glandular structures were noticed on the wall of the cyst. Finally all the histological findings in the mass in the right ovary were compatible with trabecular carcinoid tumor and cystic teratoma. There was no evidence of tumoral invasion of the lymphatic, omental or appendiceal tissue. Also, no tumor cells were detected in the ascites fluid. The diagnosis was ovarian carcinoid Stage IA. Postoperative serum testing showed a CA 19-9 level of 18.5 u/ml and a CA 125 level of 10.5 u/ml. The patient was discharged on postoperative day 5.

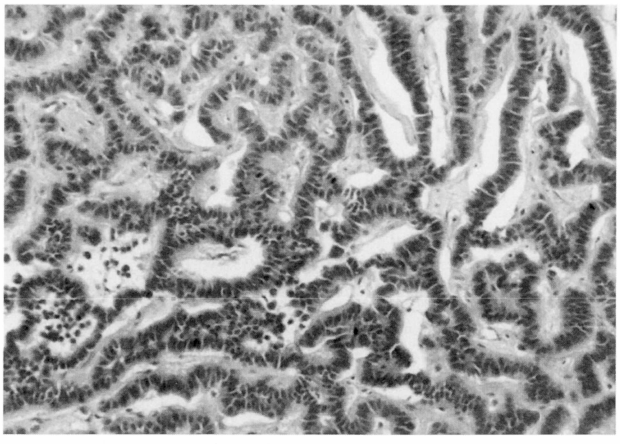

Figure 1. — The tumor is composed of long ramifying cords of tumor cells surrounded by dense fibrous stroma. The cords or trabeculae are composed of cells that contain hyperchromatic nuclei and light chromatin.

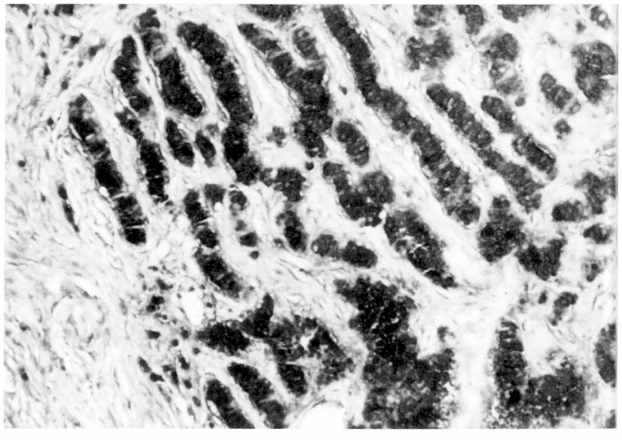

Figure 2. — Tumor cells have abundant cytoplasm and show diffuse and strong positivity with neuron specific enolase (NSE).

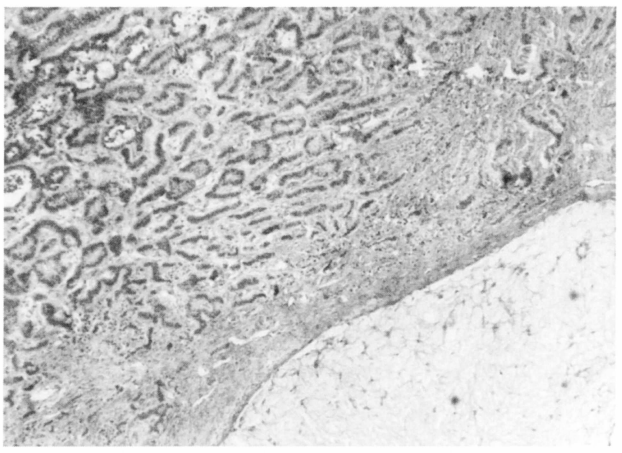

Figure 3. — A typical mucinous cyst and adjacent to the cyst a tumoral process can be observed. The tumor is composed of cords and trabeculae.

Discussion

Primary ovarian carcinoid is very rare. The literature from the past 56 years documents only 316 cases [3, 5, 7, 8] and detailed information is available for only 157 of these patients [2, 4, 5, 8-20]. Carcinoid syndrome was noted in 21 (13%) of these 157 cases. In our case, the ovarian carcinoid showed the trabecular pattern and the patient did not exhibit carcinoid syndrome. As described above, this syndrome is characterized by skin changes (flushing, telangiectasia, pellegra dermatosis), abdominal pain and secretory diarrhea (without tumoral bowel involvement), pulmonary and cardiovascular effects (hemodynamic derangement, wheezing, and structural damage to the heart) [21] called carcinoid heart disease.

Some reported cases of ovarian carcinoid have featured excessive pain or pressure with defecation [4, 20]. This may be due to inhibition of intestinal motility by the tumor-producing gut hormone peptide YY. The major complaints that led to hospitalization in these cases were pain with defecation and abdominal pain. The level of gut hormone peptide YY was not measured in any of these patients.

Other authors have documented normal CA 125 levels in two cases of advanced ovarian carcinoid and one case of early-stage disease. Our case was also early (Stage I) disease. After surgery, the patient's tumor marker levels were in the normal range.

The reported 5-year survival rate for patients with early stage disease is excellent, however, the corresponding rate for advanced-stage patients is low, at 33% [22]. Early-stage ovarian carcinoid tumors confined to one ovary should be treated with surgery alone and excellent outcomes can be expected. For patients with advanced disease, postoperative systemic combination chemotherapy is recommended. These patients must also be thoroughly investigated for non-ovarian primary tumors, such as the more common tumors of gastrointestinal origin. Experience with therapy for advanced-stage patients is limited and further studies are needed.

Conclusion

Primary ovarian carcinoid is a very rare neoplasm. Our case featured the trabecular pattern and the patient did not exhibit carcinoid syndrome. The treatment for early-stage ovarian carcinoid tumors confined to one ovary is surgery alone and excellent outcomes can be expected in these cases.

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