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Case Report
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Ovarian Carcinoid Tumor with Nodal Metastases: Case Report

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Abstract

Primary ovarian carcinoid tumors are very rare, they represent less than 0.1% of all ovarian cancers. The insular type is the most common, followed by the stromal, trabecular and mucinous types. A woman of 47 years old presented with lower abdominal pain, ultrasound evaluation revealed a voluminous pelvic mass on the right side. The patient underwent debulking surgery, it was diagnosed a carcinoid of the left ovary with prevalent trabecular and partly cribriform and insular pattern. Our patient had periaortic lymph node metastases, in this respect there is no evidence of involvement of lymph nodes for primary ovary, especially in the trabecular form, except for intestinal and pulmonary carcinoids. Therefore, this is the first case of mixed primary ovarian carcinoid, particularly in predominantly trabecular form, with lymph node metastases, as described in the literature.

Introduction

Primary ovarian carcinoid tumors are very rare, they origin from neuroendocrine tissue and represent less than 0.1% of all ovarian tumors (De et al., 2004). There are different histological patterns: insular, trabecular, strumal and mucinous (Davis et al., 1996). The insular pattern is the most common (26-53%), followed by stromal (26-44%), trabecular (23-29%) and mucinous (1.5%) type (Dotto et al., 2008). Unlike other histotypes, the trabecular one is characterized by a clinical carcinoid syndrome and a favorable prognosis. Generally, carcinoid of the ovary shows a single histological type, however, some cases of primary mixed carcinoid, characterized by two or more subtypes, have been reported (Mungen et al., 1997). Furthermore, over 90% of cases are unilateral, distant metastases have been described only for subtypes with high proliferative activity, such as mucinous and insular (Kopf et al., 2005). We present a rare case of ovarian mixed, trabecular and insular, carcinoid with lymph node metastases.

Case Presentation

A woman of 47 years old has come to the Obstetric and Gynecology Emergency Room at the Cannizzaro Hospital of Catania, reporting lower abdominal pain for about 1 month. The patient underwent pelvic examination and ultrasound revealed a heterogeneous and hypervascular neoplasm on the right side (cm 12 x 6 x 11), ascites appears as a collateral finding. The patient underwent also abdominal CT, which showed "extended expansive mass of probable adnexal origin (10 x 12 cm), with more lobes, assuming inhomogeneous enhancement. This mass compresses the bladder wall on the right. There is evidence of abundant perihepatitis and perisplenic ascites, that extends to the Douglas. Serum markers values were elevated (Ca125: 128 U / ml, CA19.9: 414 U / ml Ca15.3: 89.6 U / ml, CEA: 9:57 ng / ml), while preoperative routine laboratory tests were within the normal range. The endoscopic evaluation of esofagus and stomach was normal. Colonoscopy was not performed because of refusal by the patient. Prior ECG, chest X-ray, laboratory tests and preoperative informed consent, the patient underwent to surgery: longitudinal xipho-pubic laparotomy was performed. Inspection of the abdominal cavity is highlighted with evidence of: abundant ascites (1000 cc) which was aspirated and sent for examination; extensive omental adhesions with the parietal peritoneum; the uterus and the right annexe appeared regular in volume and surface, contrary the left annexe presented increased fallopian tube and ovary with a white-yellowish surface, of tense elastic consistency, with size of 15 cm. At inspection of the abdominal cavity, diaphragm was macroscopically free, like that liver and the other abdominal organs. It was performed resection of the left annexe, the specimen was sent for histological examination that reported: "malignant neoplasm with invasive carcinoma infiltrating the capsule". It is therefore completed debulking surgery with total hysterectomy, resection of the right annexe, total omentectomy, systematic pelvic and aortic lymphadenectomy. Residual tumor was absent. The final histopathology reported: "absence of malignant cells in ascites, uterus (cm 10 x 6 x 5) with proliferative endometrium and intramural leiomyomas; right ovary characterized by stromal hyperplasia, evidence of small adeno-fibroids on surface and edematous fallopian tube, left ovary appeared entirely occupied by solid neoformation vegetating on the outer surface". It was diagnosed a carcinoid of left ovary with prevalent trabecular and partly cribriform and insular pattern; cells arranged in thin dies with large, eosinophilic cytoplasm; there was tumor necrosis, infiltration and overcoming ovarian capsule, while the tube was free. The examination of other structures resected showed: omentum was congested, 8 common and external iliac lymph nodes on the right side were free from malignancy, three right obturator lymph nodes were sane, 6 peri-aortic lymph nodes on 12 were metastatic for trabecular subtype, 1 aortic bifurcation lymph node was free from disease, 1 left iliac lymph node was sane, 6 obturator lymphnodes were regular". Immunohystochimic examination showed positivity for EMA, Chromogranin A, synaptophysin, CD99. Final diagnosis revealed a "neuroendocrine carcinoma of the left ovary with chromogranin + +." The patient didn't need a transfusion. The postoperative Hb values were equal to 13.6 g / dl. The postoperative course was smooth, the discharge occurred 5 days after surgery. In relation to the clinical status, the pathology, the surgical intervention performed and the histology, after collegial discussion, the patient was sent to medical oncology consultation, which scheduled antiblastic therapy following scheme

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Carboplatin AUC6 + Taxol 175 mg / m q21 (the first administration was performed 4 days after surgery and it was well tolerated) for a total of 6 cycles.

Conclusions

Primary ovarian carcinoid tumors are rare neoplasms, representing less than 0.1% of all ovarian cancers and less than 1% of carcinoid tumors (Kuscu et al., 2003). They affect mainly postmenopausal women, the mean age of presentation is 50 years (Dotto et al., 2008). Most carcinoids occur in the GI tract, particularly in the appendix and then ileum, stomach, rectum, colon and duodenum (Athavale et al., 2004). The respiratory system is affected in 25% of cases, the ovaries are rarely involved. Ovarian carcinoids are characterized by neuroendocrine cells with round or cuboidal core and uniform granular chromatin and small nucleoli. The cytoplasm is usually abundant and eosinophilic. We recognize four histologic types: insular, trabecular, mucinous and stromal. The insular is the most common subtype, represented in 26-53% of cases followed by stromal (26-44%), trabecular (23-29%), and mucinous (1.5%) type (Dotto et al., 2008). The mixed form consists mainly of trabecular and insular subtypes and it is classified according to the predominant pattern. It occurs in pure form in 15% of cases, or mainly combined with other elements as a dermoid cyst or a struma ovarii. Regarding immunohistochemistry, carcinoids are immunoreactive to neuroendocrine markers such as chromogranin, Leu-7 and the sinaptosina or, lesser extent, peptide hormones such as serotonin, gastrin, insulin, glucagon. Carcinoids occur predominantly with nonspecific symptoms such as pain and bloating. Carcinoid syndrome is characterized by flushing, diarrhea, bronchospasm and hypertension, it is related to the secretion and release of metabolites by 5-hydroxytryptamine, kinins and prostaglandins by the tumor, it has been reported in 30% of insular carcinoid and in a few cases among trabecular and stromal forms. The primary ovarian carcinoid is tipically a unilateral disease. bilateral involvement is very rare or is indicative of secondary spread within the peritoneal cavity, which is a feature of carcinoid of the gastrointestinal tract (Mungen et al., 1997; Resl et al., 2003). The survival rate for metastatic carcinoid is 65% at 1 year and 25% at 5 years (Dotto et al., 2008). In most cases, primary ovarian carcinoids arise in the early stages of disease and only few cases reported early distant metastases. In case of metastatic disease, both ovaries are involved and more than 90% of patients have metastases outside the ovary (Dotto et al., 2008). A study by Davis et al (Davis et al., 1996) described 17 patients with primary carcinoid of ovary and showed the presence of lung and liver metastases in 6 patients, only for the insular subtype. For trabecular type, no metastatic case has yet been reported (Timmins et al., 2000).

In our case, the patient suffers from unilateral mixed primary carcinoid of the ovary, with predominant trabecular component than insular. Typically the two patterns occur in isolation, but the few cases reported also described a predominance of trabecular subtype with some small foci of insular cells. Our patient had periaortic lymph node metastases, there is no evidence of involvement of lymph nodes for primary ovary, in the trabecular form. Lymphonode involvement is reported for intestinal and pulmonary carcinoid. Therefore, this is the first case of mixed primary ovarian carcinoid, particularly in predominantly trabecular form, with lymph node metastases, as described in the literature.

Concerning therapy, surgical excision is recommended for cancers at early stages, while management in advanced forms is rather doubtful. In some cases, radical surgery remains the best treatment in terms of survival and palliation (Athavale et al., 2004). There is no evidence on the benefits of adjuvant therapy or the type of drug; our patient was therefore referred to in the standard chemotherapy regimen for epithelial ovarian cancer. Further studies and close follow-up for the correct outcome evaluation of the patient are recommended.

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