A case report of rare Ovarian Neuroendocrine carcinoma

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Abstract

“Unlike lung cancer, which arises from the lung, or a breast cancer that arises from the breast, neuroendocrine tumors arise from a cell type. These neuroendocrine cells can really arise from [almost] anywhere in the body.” – Dr. Lowell Anthony

Historically misunderstood to be rare and relatively benign, neuroendocrine tumors (NETs) are increasingly common and are now regarded as malignant neoplasms that can cause debilitating symptoms and potentially life-threatening issues for patients.

Neuroendocrine tumors are a heterogeneous group of separate clinico-pathological entities that share a common characteristic, i.e., expression of endocrine differentiation potential. In the ovary, the term “neuroendocrine” relates mainly to widely-known carcinoids, but it may also be applied to neuroendocrine carcinomas of non-small-cell type and small cell carcinomas of pulmonary type. Ovarian carcinoids develop in pure form or in association with other tumors, mainly teratomas. They originate from endocrine cells, either of teratomatous origin or possibly also indigenous. Ovarian neuroendocrine carcinomas belong most probably to surface epithelial neoplasms, which express endocrine pathway of differentiation. The neuroendocrine carcinomas of non-small-cell type are characterized by the presence of islands, sheets, and trabeculae with little intervening stroma (organoid growth pattern) and cellular homogeneity. However, they are higher-grade than carcinoids. Primary ovarian small cell carcinomas of the pulmonary type do not differ histologically from their counterparts in other organs. They are composed of small cells with scanty cytoplasm and oval to spindle-shaped nuclei.

Keywords: NETs, neuroendocrine tumor, .

Introduction

Neuroendocrine tumors (NETs) are neoplasms that arise from cells of the endocrine and nervous systems. Many are benign, while some are malignant. It is most common in the intestine, where they are often called carcinoid tumors, but they are also found in the lung and the rest of the body. Although there are many kinds of NETs, they are treated as a group of tissue because the cells of these neoplasms share common features, such as looking similar, having special secretory granules, and often producing biogenic amines and polypeptide hormones.

In the ovary, the term “neuroendocrine” relates mainly to widely-known carcinoids, ovarian carcinoids develop in pure form or in association with other tumors, mainly teratomas. They originate from endocrine cells, either of teratomatous origin or possibly also indigenous. Ovarian neuroendocrine carcinomas belong most probably to surface epithelial neoplasms, which express endocrine pathway of differentiation. The neuroendocrine carcinomas of non-small-cell type are characterized by the presence of islands, sheets, and trabeculae with little intervening stroma (organoid growth pattern) and cellular homogeneity. Primary ovarian small cell carcinomas of the pulmonary type do not differ histologically from their counterparts in other organs. They are composed of small cells with scanty cytoplasm and oval to spindle-shaped...
nuclei. To date about 13 cases of this tumor type in the ovary have been reported. Some of them developed in pre-existing benign or malignant ovarian tumors. The prognosis for this tumor type is poor (2).

Case description

We describe a rare case of a Neuroendocrine carcinoma of the ovary of a 27-year-old woman who presented in 2006 with pain, discomfort in the lower part of the abdomen and rectal bleeding. A CT scan revealed a large tumor in abdomen, with involvement of the liver omentum. The patient underwent several investigations including colonoscopy with biopsy. The biopsy from rectum revealed NET differentiated grade 2, highly positive for NSE, EMA, CK, S100, Chromogranin A. and Ki-67 was between 9% and 11%.

In November 2007 she underwent surgery and a large tumor, 21 x 14 x 6 cm, weight 1708 grams was removed. A liver biopsy and omentum pathology revealed neuroendocrine differentiation with primary localization, probably from ovary. The patient started treatment with Temozolomide, Capecitabine and Bevacizumab. She was also started on monthly injection of Sandostatin LAR.

Due to insurance problems the patient returned to her home country for unconventional treatment. She returned back a year later with discomfort in the lower abdomen, rectal bleeding and repeat investigations revealed lung metastases, progression in the liver and recurrent tumor in the pelvis. The patient started second line chemotherapy with Doxorubicin, Cyclophosphamide, Vincristine and Bevacizumab but stopped treatment after 5 cycles due to severe side effects.

In October 2010 a CT scan revealed disease progression and a huge abdominal distension was noticeable (from 19 cm to 27 cm), (Figure 1). The patient had also bilateral hydronephrosis, right iliac adenopathy. She was commenced on the m-Tor inhibitor Everolimus and Sandostatine LAR and continued treatment for almost 16 months with good tolerability but still progressed throughout the treatment.

In March 2012, she was operated upon as a relief measure and had laparotomy, debulking surgery and oophorectomy. A huge circumscribed solid cystic mass weighing 8540 grams and measuring 30 x 25 x 20 cm was removed (Figure 2). Patient continued to progress and passed away in April 2012.

Discussion

To our knowledge, this case report is one of few involving a primary neuroendocrine carcinoma of the ovary occurring in association with a mucinous neoplasm. Mixed mucinous and neuroendocrine carcinoma of the ovary may represent a rare neoplasm with extremely aggressive behavior.
Primary Neuroendocrine carcinoma of non-small cell type in the ovary is an extremely rare neoplasm. They differ histologically from ovarian carcinoma of the small cell pulmonary type. Only few cases have been reported worldwide and were associated with a surface epithelial-stromal neoplasm which may be benign, borderline or malignant. NET’s are always unilateral and age range is between 22 and 77 years. Prognosis is extremely poor with metastases to peritoneum, liver and other abdominal organs\(^{(3,5)}\).

**References**


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