

Primary Ovarian Small Cell Carcinoma of Pulmonary Type: An Exceptionally Rare and Aggressive Tumor with Poor

Prognosis.

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Introduction

Primary small cell carcinoma of the ovary is an extremely rare tumor. There are two types of small cell carcinoma described in the literature: hypercalcemic type and pulmonary types. The pulmonary type often affects older women and is often diagnosed at stage 3-4.¹ Current platinum-based treatment has not resulted in long term survival.¹

Aim

Our study aims to investigate the clinicopathological findings and treatment of SCCOPT in a young 31-year-old woman.

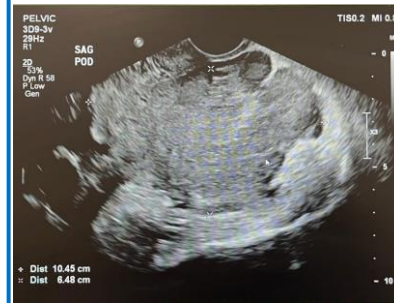
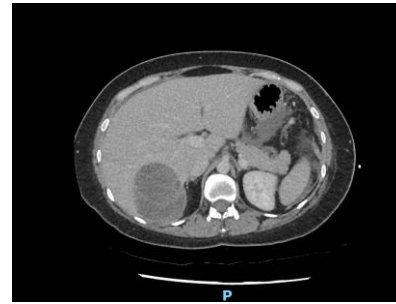


Figure A. Left adnexal mass



B. Liver metastasis

Case report

She presented with one month history of worsening lower abdominal discomfort. Pelvic ultrasound demonstrated a 10.4 x 6.5 x 7.3cm left adnexal mass inseparable from the left ovary, and a large volume of ascites. CA-125 was moderately elevated at 379. CTCAP redemonstrated the pelvic findings without evidence of extraperitoneal disease. A subsequent diagnostic laparoscopy confirmed extensive peritoneal disease. Ascitic fluid cytology was suspicious for metastatic small cell ovarian cancer.

Initially, the provisional diagnosis was small cell ovarian cancer of hypercalcaemic type, and she was treated with the PAVEP regimen (Cisplatin, Doxorubicin, Etoposide, Cyclophosphamide).

CTCAP following her first cycle of chemotherapy, showed a mixed response with stable bulky intra-abdominal disease and new bilateral pleural effusions. The pleural fluid cytology unfortunately confirmed metastatic small cell ovarian cancer.

Meanwhile, negative SMARCA2 and SMARCA4 staining confirmed SCCOPT. Therefore, chemotherapy with a de-intensified cisplatin/etoposide regimen was started as per consensus.

Unfortunately, the cancer progressed very fast and within a mere two months since her disease presentation she succumbed to death.

Discussion

SCCPT has microscopic features and neuroendocrine profile of pulmonary small cell carcinoma. It is composed of small cells with scanty cytoplasm, oval to spindle shaped nuclei with inconspicuous nucleoli.² The available treatments include different regimens of chemotherapy drugs before or after extensive pelvic surgery based on the extensiveness of the disease. Whatever treatment is employed, generally the patient surviving more than two years is low.

In conclusion, data on this type of tumor is scarce. There is no optimal therapy, and the prognosis remains poor.

References:

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2. Mebis, J., De Raeve, H., Baekelandt, M., Tjalma, W. A., & Vermorken, J. B. (2004). Primary ovarian small cell carcinoma of the pulmonary type: a case report and review of the literature. *Eur. J. Gynaecol. Oncol*, 392, 2936.