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Case report

Primary small cell ovarian cancer of pulmonary type: A case report

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ABSTRACT

Background: Primary small cell ovarian cancer of pulmonary type (SCCOPT) is a rare aggressive ovarian tumour with an incidence of < 1%, usually occurring in perimenopausal or postmenopausal women and known to have a poor prognosis. Current treatment is platinum based but has not resulted in long term survival.

Case presentation: We report a case of a 77-year old Caucasian woman who presented initially with a one-week history of abdominal discomfort with raised inflammatory markers and Ca125 of 50 μ/ml. Calcium levels were normal. She underwent primary debulking surgery, and histology showed a tumour comprising areas of classical small-cell carcinoma morphology. 6 cycles of adjuvant chemotherapy with carboplatin was offered. Relapsed/progressive disease was noted after 3 months of chemotherapy and patient died 7 months after treatment completion.

Conclusions: SCCOPT is a rare aggressive malignancy with majority of the women having an overall survival of 2 years. There is no clear consensus for the diagnosis and optimal treatment.

1. Introduction

Primary ovarian neuroendocrine malignant neoplasms are extremely rare, accounting for < 2% of all ovarian cancers, and usually related to the large cell type (Reed et al., 2014). Primary small cell ovarian cancer was first described in 1979 (Reed et al., 2014). It is a highly aggressive tumour with an incidence rate of < 1% of all ovarian cancers and a poor outcome (Reed et al., 2014; Eichhorn et al., 1992). Two tumour variants are described; the small cell cancer of hypercalcemic type (SCCOHT) and the pulmonary type (SCCOPT) with SCCOHT occurring in younger women and SCCOPT in older women respectively (Reed et al., 2014; Atienza-Amores et al., 2014). They are not easily distinguished pre-operatively from common epithelial ovarian cancers and the differential diagnosis will include germ cell and granulosa/sex-cord tumors (Reed et al., 2014). To our knowledge, only 22 cases in total of SCCOPT have been described in the literature with majority of these arising in mature cystic teratomas (Eichhorn et al., 1992; Rubio et al., 2015; Ikota et al., 2012; Lim et al., 1998; Chang et al., 1992; Grandjean et al., 2007; Kurasaki et al., 2013; Mebis et al., 2004; Suzuki et al., 2007; Lo Re et al., 1994; Tsolakidis et al., 2012; Reckova et al., 2010). However, from these, only 8 cases are described as "pure" primary SCCOPT (Eichhorn et al., 1992; Kurasaki et al., 2013; Suzuki et al., 2007; Lo Re et al., 1994; Tsolakidis et al., 2012; Reckova et al., 2010) (Table 1).

We report a clinical case of unilateral primary SCCOPT presented in a 77-year old woman, the current treatment options and clinicopathological considerations.

2. Case presentation

A 77-year old woman presented to Accident and Emergency with 1-week history of low abdominal pain, intermittent difficulty initiating micturition and constipation. Clinical examination showed no ascites and a pelvic mass was found in the pelvis. Ultrasound of the pelvis demonstrated a 11.7 × 9.6 × 11.8 cm complex mass in the midline of the pelvis extending to the right adnexa (Fig. 1). Her blood results were satisfactory, except for a slight increase in inflammatory markers and Cancer Antigen (CA)-125 of 50 μ/ml, calculating the Risk of Malignancy Index (RMI) at 450 (Davies et al., 1993).

Hence, Computed tomography (CT) of chest, abdomen and pelvis was performed which revealed within the pelvic cavity a large 14 × 8.2 × 9.5 cm heterogeneous multi-septated mass with solid and cystic components originating from the right adnexa and mostly keeping with ovarian tumour (Fig. 1). There was no evidence of disease

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Table 1
Case reports and clinopathological characteristics of primary ovarian small cell carcinoma of pulmonary type.

Case reports	Age (years)	FIGO stage	Origin and size (cm)	Treatment	Postoperative adjuvant treatment	Recurrence (months)	Overall survival (months)
Case 1: Eichhorn et al. (1992)	76	IIIB	ROV/NA LOV/NA	STAH, BSO	NA	NA	12
Case 2: Eichhorn et al. (1992)	64	IIIB	ROV/5.5 LOV/4.5	TAH, BSO, OMT, AP	Cisplatin and cyclophosphamide	8	NA
Case 3: Eichhorn et al. (1992)	49	IIIB	LOV/16	LSO, COL, LYM	Cisplatin, cyclophosphamide and doxorubicin	NA	13
Lo Re et al. (1994)	16	IIIA	LOV/9.5	TAH, BSO, OMT	Vinblastine, cisplatin, cyclophosphamide, bleomycin doxorubicin and etoposide	NA	13
Suzuki et al. (2007)	49	IC	LOV/15	TAH, BSO, OMT, LYM	Paclitaxel and carboplatin	NA	≥36
Reckova et al. (2010)	67	IV	ROV/6	TAH, BSO, OMT, AP	Carboplatin and etoposide	NA	24
Kurasaki et al. (2013)[a]	54	IIIA	NA	TAH, BSO, OMT	Paclitaxel and carboplatin	NA	NA
Tsolakidis et al. (2012)	55	IIIC	LOV/8	TAH, BSO, OMT, LYM	Carboplatin and etoposide	NA	≥21
Current case	77	II	ROV/15	TAH, BSO, OMT	Carboplatin	3	7

NA: Not available; ROV: Right ovary; LOV: left ovary; STAH: Subtotal abdominal hysterectomy; TAH: Total abdominal hysterectomy; BSO: Bilateral salpingoophorectomy; OMT: Omentectomy; LYM: Lymphadenectomy; AP: Appendicectomy.

[a] The patient was followed up for 22 months without recurrence.

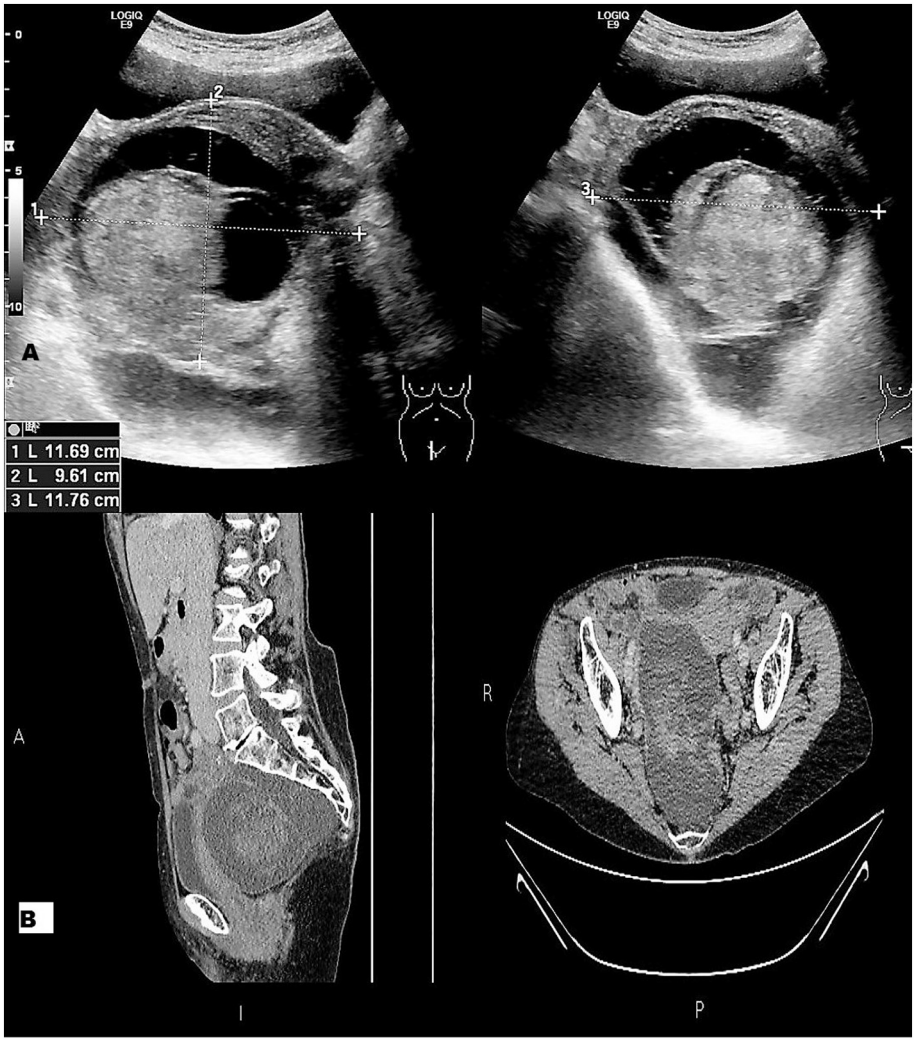

Fig. 1. Imaging findings: (A) Ultrasound of the pelvis demonstrating a 11.7 × 9.6 × 11.8 cm complex mass in the midline of the pelvis extending to the right adnexa (B) Abdominal CT scan demonstrating a 14 × 8.2 × 9.5 cm heterogeneous multi-septated mass with solid and cystic components originating from the right adnexa.