

A rare encounter with small cell carcinoma of the ovary, hypercalcemic type (SCCOHT): Diagnostic dilemma and management approach

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ABSTRACT

Introduction: Small-cell carcinoma of the ovary, hypercalcemic type, is a rare and aggressive ovarian malignancy accounting for less than 0.01% of all ovarian tumours. It predominantly affects young women, with a median age of 24 years, and is associated with hypercalcemia in approximately two-thirds of cases. The disease is driven by inactivating mutations in the SMARCA4 gene. Due to its rarity, there is no standardised treatment protocol, and long-term survival remains poor even in early-stage disease. **Case Description:** A 26-year-old woman with no significant medical history presented with a 2-week history of rapidly increasing abdominal distention. Examination revealed a firm suprapubic mass equivalent to a 28-week gravid uterus. CT imaging identified a large solid-cystic pelvic mass (10.6 × 21.8 × 23.6 cm) without ascites or metastases. Laboratory investigations showed asymptomatic hypercalcemia (4.91 mmol/L), which was managed with hydration and bisphosphonates. Surgical resection was performed. Histopathology was inconclusive for common ovarian tumours but suggested SCCOHT based on immunohistochemical findings (WT1, CD99, CD10, calretinin, p16, CKAE1/AE3, and synaptophysin positivity). Genetic testing for SMARCA4 was initiated. She received three cycles of BEP chemotherapy but developed an allergic reaction to etoposide. Residual FDG-avid disease was noted on PET scan. She completed six cycles of carboplatin and paclitaxel, later developing a pulmonary embolism treated with rivaroxaban. **Discussion:** This case illustrates the diagnostic and therapeutic complexity of SCCOHT. Despite aggressive management, the prognosis remains poor. Hypercalcemia in young women with adnexal masses should raise suspicion for SCCOHT. Multidisciplinary care and continued reporting of such rare cases are crucial to advance understanding and develop evidence-based treatment strategies.