# A Case Report of a Limited Stage Small Cell Carcinoma of the Vulva

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## Introduction

Extra-pulmonary small cell carcinomas (ESCCs) are rare and often aggressive malignancies, accounting for less than 2% of all gynaecological malignancies. These tumours are diverse in origin, are thought to arise from multipotent stem cells present in all tissues[1] and occur most commonly in the genitourinary and gastrointestinal systems[1-3]. There are limited case reports in the literature about ESCCs that mimic Bartholin's gland cysts[4-6]. Treatment approaches are extrapolated from available data about small cell lung carcinomas (SCLCs) and, in general, include surgery with adjuvant chemoradiotherapy[2]. ESCCs are generally considered to have poor prognosis, though prolonged survival has been achieved in patients with localised disease through aggressive multimodal therapy[7-8].

## Aim

To present a rare case of vulval small cell carcinoma.

## Case Report

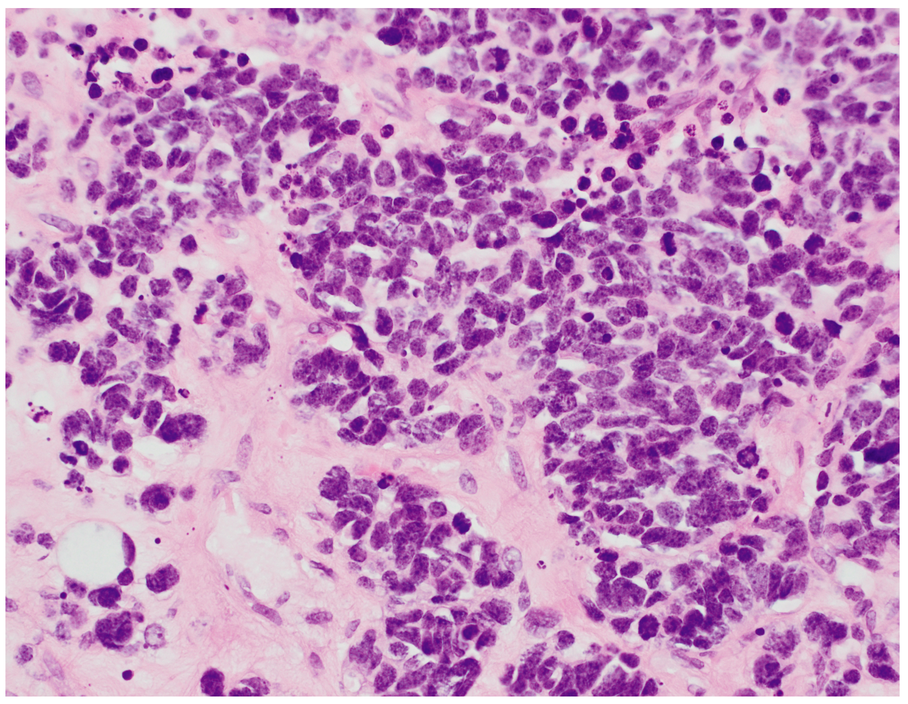
A 65 year-old P5 post-menopausal woman presented with a 3-month history of a painful and enlarging right labial lump that appeared to arise from the right Bartholin's gland. Clinical examination revealed a firm right labial nodule with a 20mm diameter with otherwise normal external genitalia. Ultrasound demonstrated a heterogenous mass with internal septations and vascularity. The patient's past surgical history included a total abdominal hysterectomy, a laparoscopic ovarian cystectomy, a gastric band, two lower uterine segment caesarean sections, and previous incision of a contralateral Bartholin's cyst. Her medical history was also significant for a current smoking status with 15 pack-year history.

Over the two-week period between initial review and surgical excision, a modest increase in size was noted, with the mass measuring 40 x 29 x 41mm. Histopathology revealed a high-grade malignant tumour with morphological features favouring small cell carcinoma. It was composed of sheets and vague cords of basaloid cells with minimal discernible cytoplasm, marked hyperchromasia and lacking chromatic detail and nucleoli (Image 1). On immunochemistry, the cells were chromogranin and CD56 positive, with diffuse strong p16 staining (Image 2). The tumour infiltrated fibrous tissue, skeletal mass and adipose tissue and there were positive margins. Staging CT did not reveal any obvious metastatic disease, although noted a stable 7mm right apical spiculated lung lesion (seen on a previous CT in 2015 and felt to be suggestive of lung fibrosis), and hepatic cysts. A PET scan subsequently showed localised disease only (Image 3). The patient completed four cycles of chemotherapy (cisplatin/etoposide) with concomitant radiotherapy (total dose received 50.6 GY/22##). Ongoing surveillance is planned – the most recent PET scan, approximately 8-months after resection, demonstrated complete metabolic response.

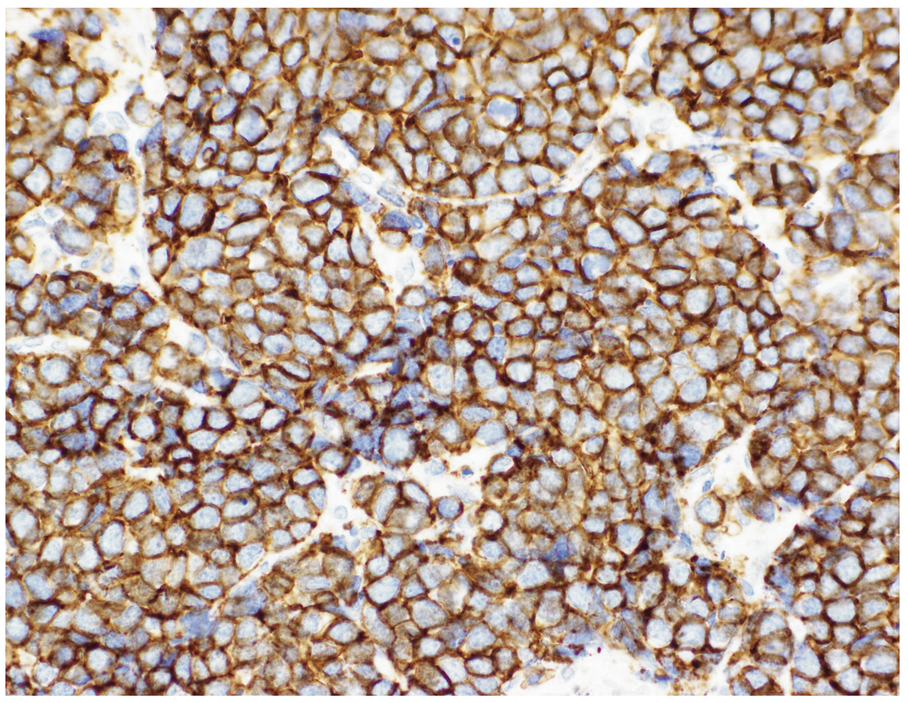
## Conclusion

This case highlights the importance of considering malignancy as a potential cause of vulval masses in post-menopausal women. If extra-pulmonary small cell carcinoma is detected, prompt referral is required as these are aggressive malignancies with significant risk of relapse, even in instances of localised disease treated with radical chemoradiotherapy. Given risk factors for these lesions are not well established, clinicians should strongly consider resecting or performing a biopsy of any vulval mass in a post-menopausal woman.

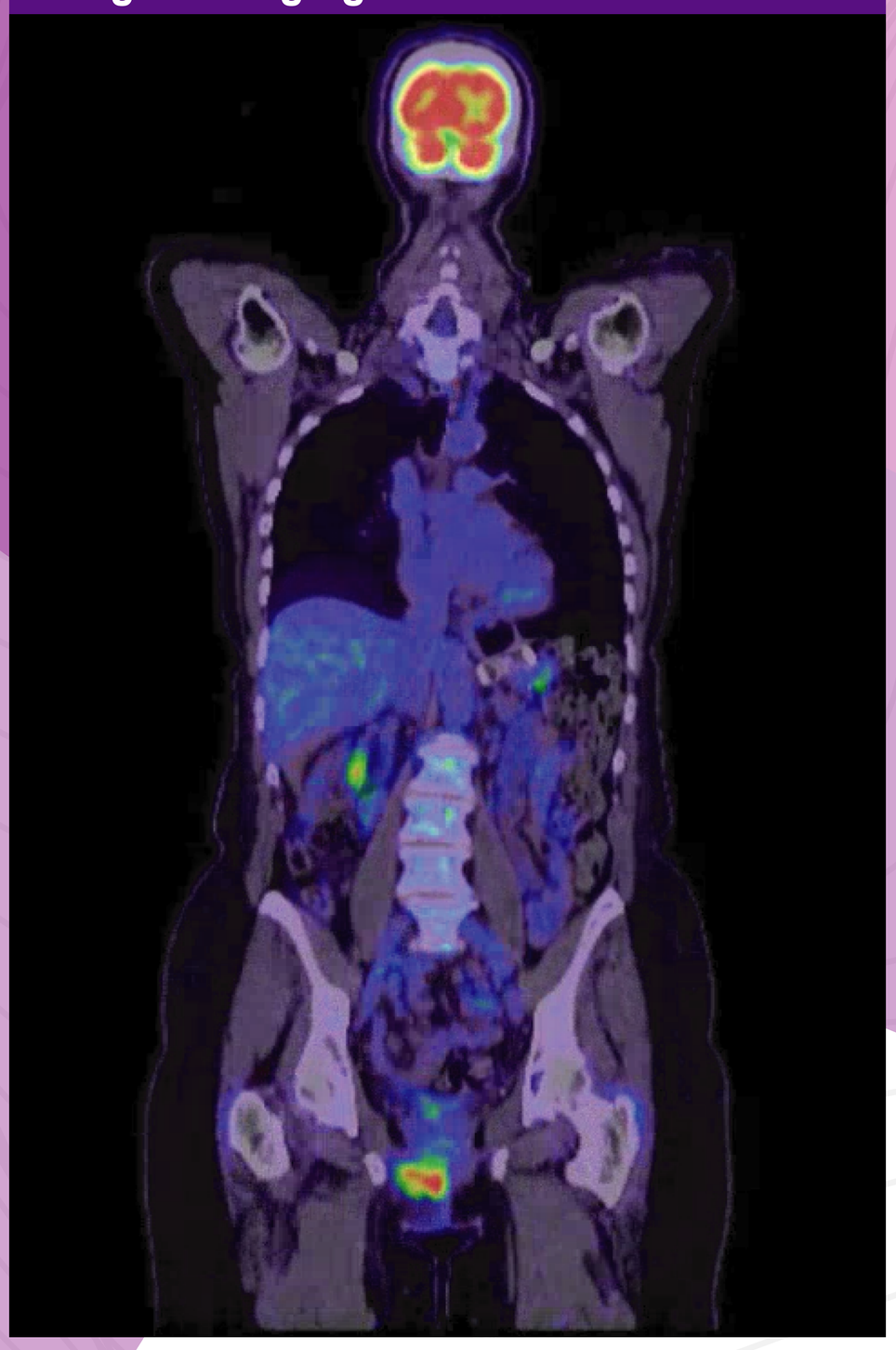
## Image 1. H&E staining



## Image 2. Chromogranin staining



## Image 3. Staging PET scan



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