Extrapulmonary small cell carcinomas are rare tumors that originate from many different body sites including the gastrointestinal tract, larynx, and urogenital system.[1, 2] Prostatic small cell carcinoma (PSCC) is a rare and aggressive malignancy of neuroendocrine cell lineage arising in the human prostate, with an estimated incidence of 0.5% to 2% of all primary prostatic tumors.[3] The mean age at presentation is 65 years. The median cancer specific survival of patients with small cell carcinoma of the prostate in 259 men according to the SEER database from 1973-2004 was 19 months. 60.5% of men presented with metastatic disease with a decreased survival related to stage 2 and 5 year survival rates of 27.5% and 14.3%, respectively.[4] Pure PSCC accounts for 50% of cases and the remainder demonstrate an admixture of PSCC and adenocarcinoma.[2] In contrast to most prostatic adenocarcinomas, PSCC is rarely a primary presentation. Presenting symptoms may be related to metastases and rarely to paraneoplastic syndromes such as ectopic ACTH production[5], inappropriate ADH release[6], or myasthenic syndrome.[7] Compared to adenocarcinoma, a relative decrease in PSA production and AR expression is observed.[8] It causes lytic bone lesions, and is characterized as a rapidly growing and highly metastatic lesion.[9, 10] Pure PSCC arising *de novo* with no or minimal adenocarcinoma component behaves more aggressively than mixed variety or adenocarcinoma showing cells with neuroendocrine differentiation.[11]

PSCC is seen frequently in patients with a prior diagnosis of prostatic adenocarcinoma. It is postulated that androgen-ablation therapy enhances the progression of neuroendocrine differentiation, as these particular cells lack androgen receptors, and leads to a progressive, hormone-refractory malignancy.[12] As the number of cases so far is limited, optimal therapy for PSCC is yet to be defined. Given the high rate of occult metastases, clinically localized small cell prostate cancer is typically treated aggressively, often with multi-modality therapy with chemotherapy and radiation similar to limited stage small cell lung cancer. Metastatic small cell carcinoma of the prostate is treated with platinum based combination chemotherapy with regimens similar to those used to treat small cell lung carcinoma.[13-15]

In conclusion, PSCC is an aggressive malignancy, often presenting with advanced metastasis at the time of diagnosis, and may develop in patients with previously diagnosed adenocarcinoma who have received androgen ablation therapy. Pure prostatic small cell neuroendocrine carcinoma should be distinguished from prostatic carcinoma with focal neuroendocrine differentiation. This distinction carries important prognostic and therapeutic implications.