Primary Signet Ring Cell Carcinoma of Prostate - A Rare Occurrence

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ABSTRACT

BACKGROUND
Primary signet ring cell carcinoma of prostate is an extremely rare variant of prostatic adenocarcinoma having a grave prognosis. It is characterized by an intracytoplasmic vacuole compressing the nucleus into a crescent shape at the cellular level. Although SRCC is primarily found in the stomach and colon, it can also be found in the pancreas, breast, thyroid, bladder, and prostate. Establishing a diagnosis of SRCC of the prostate requires histopathologic examination of the tissue, a negative gastrointestinal (GI) work-up (including computed tomography of the abdomen, a colonoscopy, and an esophagogastroduodenoscopy), and various stains. A PubMed based literature search was made regarding the incidence, clinical manifestations, microscopy, histological grade and stage of this tumour.

Signet ring cell carcinoma, a rare and aggressive subtype of adenocarcinoma of the prostate, requires careful diagnosis via thorough evaluation and specialized staining processes. Although the rarity of this disease does not allow for the establishment of absolute recommendations, treatment is often similar to traditional radical management of cancer of the prostate, with emphasis on an aggressive multimodal approach. Patients with SRCC of the prostate have an ominous prognosis and should be treated aggressively and followed up closely. Primary signet ring cell carcinoma is a rare variant of adenocarcinoma and therefore a detailed clinico-radiological and pathological work up should be done in order to rule out primary from other common sites.

KEYWORDS
Primary, Signet, Carcinoma, Prostate, PubMed

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INTRODUCTION

Signet ring cell carcinoma (SRCC) of the prostate is a rare tumour characterized by an intracytoplasmic vacuole compressing the nucleus into crescent shape at the periphery. Although SRCC is more commonly found in the stomach and colon, it can also be found in the pancreas, breast, thyroid, bladder, and prostate. Our review of the English-language literature identified only 46 cases of SRCC of the prostate through case reports and case series. Establishing a diagnosis of SRCC of the prostate requires histopathologic examination of the tissue, a negative gastrointestinal (GI) work-up (including computed tomography of the abdomen, a colonoscopy, and an esophagogastroduodenoscopy), and various stains that help localize the primary site of cancer to the prostate. The prognosis of patients with SRCC of the prostate is poor and inappropriate management is still not discovered because of the rarity of this tumour. This study reviews the established pathologic criteria, treatment, and outcomes of prostatic SRCC.

A PubMed search was then performed of the English literature published from January 1, 2000, to January 1, 2018, using the key words signet ring cell and prostate. Only cases describing SRCC with a primary prostate tumour were included. Individual cases were outlined, and the articles were summarized regarding patient age, survival, and treatment. When available, the stage, grade, prostate-specific antigen (PSA) level, Gleason sum, histologic characteristics (special staining), and GI work-up were evaluated.

REVIEW OF LITERATURE

Twenty-five articles were identified in the English-language literature. 46 patients with SRCC of the prostate were included in the review. The PSA level varied widely, from 1.9 to 536 ng/mL. More patients (33%) presented with stage 4 cancer than with any other stage of cancer. The most common Gleason sum was 8 (33%), and a combination of surgery, radiation, and hormonal therapy was most often used (41%), followed closely by hormonal therapy alone (31%). The minority of patients 31% (16/51) had a documented GI work-up to determine the primary SRCC source. For pathologic diagnosis, the most common stains performed were the periodic acid–Schiff stain, performed in 70% of patients; the prostate-specific acid phosphatase (PSAP) stain, performed in 70% of patients; and the Alcian blue stain, performed in 62% of patients.

These were positive in 50% (18/36), 87% (34/39), and 44% (14/32) of cases, respectively. Carcinoembryonic antigen (39%) and mucicarmine (39%) were less commonly used and were positive in 20% (4/20) and 40% (8/20) of patients, respectively. The extent of signet ring cell involvement of the specimen was reported in 76% (39/51) of cases. Of these, 84% (33/39) documented more than 20% of the tumour-containing signet ring cells. The mean overall survival was 28 months in the compiled group.

DISCUSSION

Signet ring is a term used to describe the histologic appearance of a tumour cell characterized by compression of the nucleus into the form of a crescent by a large cytoplasmic vacuole. Signet ring cell changes were first described in 1981 and are estimated to occur in 2.5% of cases of adenocarcinoma of the prostate. SRCC is often found in the presence of other high-grade prostatic adenocarcinoma patterns. Regardless of histologic classification, the grim prognosis and rarity of SRCC of the prostate warrant closer investigation.

Reviewed data showed median age of prostate SRCC is 68 years (range, 50-85 years), which is comparable to the previously reported 68.2 years with mean survival of 29 months. Reports indicate that up to 75% of cases present with locally advanced or metastatic disease at the time of diagnosis. In a study done at Mayo Clinic a grading system was analysed in which a Gleason sum of 8 is equal to a grade of 3 or 4, whereas a Gleason sum of 7 approximates a grade of 2 or 3. We found that more patients presented with non–stage IV than with stage IV disease (66% vs 33%). Fujita et al. Presented a case series of 42 patients with SRCC, which demonstrated that stage IV is a poor predictor of survival. Interestingly, neither PSA levels nor treatment modalities were predictive of survival in that group of patients.

Multiple tests have been established to assist in the diagnosis of SRCC of the prostate. Because the GI tract is a more common location for signet ring cells, many of these tests focus on differentiating a primary tumour of the prostate from one located in the GI tract. In our review of the literature, only 31% of patients had a reported GI work-up. Histologic criteria for SRCC of the prostate are highly variable in the available literature. Some publications have reported that the cells should stain negative for leukocyte common antigen and alpha SMA.

Reports concerning whether cells should stain positive for PSA and PSAP have been inconsistent, although it appears most do. In this investigation, 87% of those actually tested for PSA and PSAP had positive stains. Other immunohistochemical stains that can be used to ensure a primary prostate tumour include o-methylacetyl coenzyme A racemase and cytokeratin 5/6. The cytoplasmic vacuoles can contain lipids or mucin and stain positive with mucicarmine in about 50% of cases, PAS in about 60%, and alcian blue in 60%.

CONCLUSIONS

Establishing a diagnosis of SRCC of the prostate requires histopathological examination of the tissue, a negative gastrointestinal (GI) work-up (including computed tomography of the abdomen, a colonoscopy, and an esophagogastroduodenoscopy), and various stains that help localize the primary site of cancer to the prostate. The prognosis of patients with SRCC of the prostate is poor and inappropriate management is still not discovered because of the rarity of this tumour. This study reviews the established pathologic criteria, treatment, and outcomes of prostatic SRCC.

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tomography of the abdomen, a colonoscopy, and an esophagogastroduodenoscopy), and various stains that help localize the primary site of cancer to the prostate. The prognosis of patients with SRCC of the prostate is grim, and recommendations for treatment are sparse because very few cases are reported.

REFERENCES