Oncology

Lymphoepithelioma-like Carcinoma (LELC) of the Prostate

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ABSTRACT

Lymphoepithelioma-like carcinoma (LELC) is an aggressive tumor that rarely affects the prostate. Few cases are reported in the literature. We present a case report, pathologic description and review of the literature.

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Introduction

Lymphoepithelioma is an undifferentiated malignant carcinoma linked to Epstein-Barr virus and well-described in the head and neck literature.1 Tumors with similar histologic features and arising from other organ systems are referred to as lymphoepithelioma-like carcinomas (LELC). LELC may affect other sites including salivary glands, thymus, lung, skin, stomach, breast and the genitourinary tract.2 In the genitourinary tract, LELC most commonly affects the urinary bladder and prostatic involvement is rare.3,4

Case report

A 77 year old male with history of tobacco use, biopsy-confirmed retroperitoneal fibrosis and dyslipidemia presented to primary care with dysuria and gross hematuria for 6 weeks. He was treated with antibiotics followed by an alpha blocker with no improvement. No digital rectal exam was performed prior to urology referral. A computed tomography (CT) scan of the abdomen and pelvis without oral or intravenous contrast 6 months prior revealed an atrophic left kidney, retroperitoneal soft tissue surrounding the aorta and lung, skin, stomach, breast and the genitourinary tract. In the genitourinary tract, LELC most commonly affects the urinary bladder and prostatic involvement is rare.3,4

He underwent a standard 12 core transrectal ultrasound-guided prostate biopsy which demonstrated large volume LELC in the left base, left mid and left lateral apex; additionally, 5% Gleason 3 + 3 adenocarcinoma was identified in the right lateral base.

On pathologic examination, the tumor cells had irregular vesicular nuclei, prominent nucleoli and indistinct cytoplasmic borders with high nuclear/cytoplasmic ratio (Fig. 1). Immunohistochemistry showed tumor cells positive for prostatic carcinoma markers ERG and NKX3.1 (Figs. 2 and 3), epithelial marker CKAE1/3 and negative for prostatic carcinoma markers (PSA, p504S) and urothelial carcinoma marker (p63 and GATA3). EBV-encoded RNA (EBRE) was not identified by chromogenic in situ hybridization (CISH).

Staging CT of the chest, abdomen and pelvis with intravenous contrast revealed a 1.5 cm spiculated nodule in the lingula with non-specific mediastinal and right hilar adenopathy most likely consistent with a chronic inflammatory process and less likely metastatic disease. Furthermore, stable pelvic adenopathy and infiltrative changes were visualized in the retroperitoneum consistent with the patient’s history of retroperitoneal fibrosis.

Bone scan was negative for metastatic disease. The patient was taken to the operating room with the intent of radical prostatectomy. Intraoperatively, pelvic nodes were enlarged and firm and frozen section was benign. The prostate was large, hard and immobile with extraprostatic tumor involvement of the distal membranous urethra and anterior rectal wall. In addition, fibrosis and adherence likely related to patient’s history of retroperitoneal fibrosis was noted between the posterior aspect of the prostate and rectum, rendering dissection extremely difficult. Prostatectomy was aborted and patient referred for pelvic external beam radiation.

Funding: Geisinger Clinic.

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Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

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http://dx.doi.org/10.1016/j.eucr.2015.12.006
Postoperative pelvis MRI revealed carcinoma involving the right lobe centered in the mid gland fat with extension to the apex and base with gross extracapsular extension and invasion of the neurovascular bundle. He completed a 9 week course of external beam radiation therapy consisting of 8100 cGy in 45 fractions. He tolerated the radiation therapy without immediate side effects and at last follow up was without symptoms, his PSA was 0.03 ng/mL, and CT scan of chest, abdomen and pelvis revealed no evidence of disease. He will continue on surveillance with physical exam and PSA every 6 months.

Discussion

Lymphoepithelioma is most commonly observed in the nasopharynx (i.e. nasopharyngeal carcinoma). The term “lymphoepithelioma” derives from the tumor’s characteristic histologic appearance, in which large polygonal epithelial cells are often surrounded by lymphoid cells. An association between nasopharyngeal carcinoma and Epstein-Barr virus has been observed, with EBV latent genes contributing to alteration in cellular gene expression and growth. Treatment typically consists of radiotherapy with or without chemotherapy. The 5 year survival is 72% stage I, 64% stage II, 62% stage III, and 38% for stage IV.

LELC of the prostate is a rare, aggressive malignancy with an overall poor prognosis. Unlike nasopharyngeal disease, there is no clear association with Epstein-Barr virus. Adlakha et al described the first case of LELC of the prostate in a 66 year old man with obstructive urinary symptoms. The patient was treated with radical prostatectomy and followed for 15 months without complications. In a cases series of 5 patients with LELC of the prostate; all presented with obstructive urinary symptoms and an elevated PSA. The mean age at diagnosis was 76 years and the initial diagnosis was made on transurethral resection in 3 patients and radical prostatectomy in the remainder. The mean follow up period was 20.2 months; 1 patient was lost to follow up and 4 died of disease.

Histologic findings in the few reported cases of prostatic LELC include syncytial cell arrangement and vesicular nuclei, absence of lumen formation, polyclonal lymphoid stroma, and positive immunoreactivity with PSA and α-methylacyl coenzyme A racemase (AMACR) in specimens from radical prostatectomy or transurethral resection of prostate. While the presented case was immunoreactive for PSA and had a histologic pattern characteristic of LELC, comparison with prior case series is limited by needle biopsy specimen.

In summary, LELC of the prostate is a rare, aggressive malignancy with poor prognosis. An accurate diagnosis is essential as LELC of prostate tends to arise in the background of adenocarcinoma. Further research is needed to refine diagnosis and treatment for this rare, aggressive entity.

Conflicts of interest

No conflicts of interest.

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