A case report of primary prostate intravascular large B cell lymphoma presenting as prostatic hyperplasia

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Abstract

Rationale: Intravascular large B-cell lymphoma (IVLCL) is a rare condition with a poor prognosis. The clinical presentation of primary lymphoma of the prostate is non-specific and it is difficult to distinguish from other prostatic diseases. The primary prostate IVLCL is very rare, the diagnosis and treatment of which remains unclear. We reported a rare case to explore the diagnosis and treatment for the primary prostate IVLCL.

Patients concerns: This report described a case of a 71-year-old male diagnosed as primary prostate IVLCL who presented with prostatic hyperplasia.

Diagnosis: The patient first visited an outpatient clinic of urinary surgery because of urinary urgency and frequency and was diagnosed as benign prostatic hyperplasia in about January 2010. Four years later, the symptoms worsened quickly within two months. The diagnosis was still prostatic hyperplasia according to the physical examination and imaging. However, histopathology showed IVLCL of prostate after transurethral resection of the prostate.

Interventions: With the clear diagnosis of primary prostate stage I IVLCL, the patient received immunochemotherapy of R-CHOP (rituximab, cyclophosphamide, adriamycin, vincristine, and prednisolone) for 4 cycles and intensity-modulated radiation therapy (IMRT) including the region of prostate with the dose of 45Gy/25f.

Outcomes: The response was complete remission after all treatment. The last follow-up time of the patient was June 20th, 2019, and no evidence of disease progression was observed. The progression-free survival of the patient was about 49 months until now.

Lessons: The biopsy of prostate by prostatectomy plays an important role in the diagnosis and removal of the original lesion of primary prostate lymphoma. There is no consensus on therapeutic modalities for the treatment of primary prostate IVLCL till now. Individual treatments include immunomotherapy and/or radiotherapy according to the National Comprehensive Cancer Network (NCCN) practice guideline of diffuse large B cell lymphoma (DLBCL) based on the performance status and tumor staging of the patient. Timely and accurate diagnosis as well as the appropriate treatment may improve the clinical outcome.

Abbreviations: CT = computed tomography, CTV = clinical target volume, NCCN = National Comprehensive Cancer Network, DLBCL = diffuse large B-cell lymphoma, H&E = hematoxylin and eosin, IMRT = intensity modulated radiation therapy, IPI = International Prognostic Index, IVLCL = intravascular large B-cell lymphoma, LDH = lactate dehydrogenase, MRI = Magnetic resonance imaging, MUM-1 = multiple myeloma oncogene 1, NHL = non-Hodgkin’s lymphoma, PET = positron emission tomography, PSA = prostate specific antigen, R-CHOP = rituximab, cyclophosphamide, adriamycin, vincristine, and prednisolone, TURP = Transurethral resection of the prostate, WHO = World Health Organization.

Keywords: immunochemotherapy, intravascular large B cell lymphoma, prostate, prostatic hyperplasia

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1. Introduction

IVLBCL is a rare subtype of extranodal DLBCL with poor prognosis, which is highly aggressive and may involve any tissue and organs.\(^1\)–\(^4\) Central nervous system,\(^5\) bone marrow,\(^6\) and skin\(^7\) are the most common systems involved with IVLBCL. It is different from the majority types of lymphoma presenting as the enlargement of lymph nodes. The histopathology of IVLBCL is characterized by the proliferation of neoplastic cells within the vascular lumen, especially within capillaries. So, the diagnosis of IVLBCL is often difficult because of the absence of specific clinical presentation, laboratory and imaging findings. Primary lymphoma of prostate is a rare condition that only accounts for 0.09% of all prostate neoplasms and 0.1% of all non-Hodgkin’s lymphoma (NHL).\(^8\) The prostate has been reported as one of the involved sites by IVLBCL in few cases.\(^9\)–\(^11\) However, primary prostate IVLBCL was reported only in three cases according to a comprehensive literature search from the electronic databases PubMed with the keywords of “intravascular large B cell lymphoma” and “prostate”.\(^12\)–\(^14\) It is lack of the accurate diagnosis and treatment strategy of primary prostate IVLBCL until now. Here we report a case of long-term survival with primary prostate IVLBCL in a 71-year-old male who presented with prostatic hyperplasia.

2. Case report

A 71-year-old male first visited an outpatient clinic of urinary surgery because of urinary urgency and frequency and was diagnosed as benign prostatic hyperplasia in about January 2010. He was treated with Tamsulosin which was an \(\alpha\)-blocker and the symptoms relieved after taking the medicine. But the symptoms worsened quickly within 2 months since September 2014. He also had hematuria occasionally during the past 2 months without fever, night sweats, weight loss or any other preceding symptoms. He visited an outpatient clinic of urinary surgery again in November 2014. Physical examination was normal. Ultrasound examination showed enlarged prostate without nodular surface which was measured as \(8 \times 4 \times 6 \text{cm}^3\). Magnetic resonance imaging (MRI) scan of the prostate showed the enlarged prostate without nodular surface in T2 enhanced weighted imaging (Fig. 1). His complete blood count, lactate dehydrogenase (LDH) and serum prostate-specific antigen (PSA) level were normal. He had no personal or family medical history of malignant neoplasm and urinary system infection. The diagnosis was still prostatic hyperplasia according to physical examination and imaging. Transurethral resection of the prostate (TURP) was performed in November 2014. The symptoms regressed after the operation. Astonishingly, the histological, and immunohistochemical studies of the prostatectomy showed an atypical, intravascular population of cells, not prostatic hyperplasia. The cells were large and featured. The atypical population showed positive staining for CD20, CD10, multiple myeloma oncogene 1 (MUM-1), Bcl-6, CD34, negative staining for CD3, ALK, CK, CD30, CyclinD1, and exhibited a high proliferation index as illustrated by Ki-67 staining (98% positive) (Fig. 2). The features were consistent with IVLBCL. The patient was diagnosed with IVLBCL in the prostate. He went to the department of lymphoma to continue the specialized therapy. A further \(^{18}\)FDG positron emission tomography-computed tomography (PET-CT) scan of the whole body showed no significantly increased metabolic activity in the region of the prostate gland. No other areas of increased metabolic activity were observed, either. Bone marrow biopsy showed no evidence of lymphoma infiltration. The final diagnosis of this patient was primary prostate IVLBCL, germinal center B-cell subtype, stage IA according to the Ann Arbor staging criteria, and International Prognostic Index (IPI) score was 1. The patient was classified as the low-risk group based on the IPI score. He received 4 courses of immunochemotherapy with R-CHOP. Followed by 2 courses and 4 courses of chemotherapy respectively, the repeated computed tomography (CT) scan from neck to pelvic cavity revealed that the response to the therapy was complete remission. He received intensity-modulated radiation therapy (IMRT) of the prostate after chemotherapy, and the dose of the clinical target volume (CTV) including the region of prostate was...
45 Gy/2.5 f. He finished all treatment in May 2015 and is still alive now with a good quality of life.

3. Discussion

IVLBCL is a rare subtype of NHL with a poor prognosis. IVLBCL was first described in 1959 by Pfleger and Tappeiner and was characterized by the diffuse proliferation of malignant B-cells in small- and medium-size vessels, especially in capillary and postcapillary venules. It can involve any organ of the body such as small- and medium-size vessels, especially in capillary and postcapillary venules. (original magnification × 400). Immunohistochemistry staining showed that the tumor cells (arrow) were CD20 positive confirming the presence of B lymphocytes. (original magnification × 400). CD34 immunohistochemistry staining highlighted the intravascular growth pattern of tumor cells (arrow). (original magnification × 400).

IVLBCL cases. Shimada reported that the addition of rituximab to CHOP or CHOP-like regimen revealed a better complete response rate, overall survival and 2-year progression-free survival than those of chemotherapy alone in the patients with IVLBCL. Despite the lack of standard therapy based on large sample of clinical trials, the chemotherapy of R-CHOP with or without radiotherapy remains the currently recommended therapy for IVLBCL according to the available data and NCCN practice guideline of DLBCL. The patient was diagnosed as primary prostate IVLBCL stage I according to the Ann Arbor staging criteria and was classified as the low-risk group based on the IPI score. The patient received 4 courses of chemotherapy of R-CHOP and IMRT including the area of the prostate with a total dose of 45 Gy to improve the local control according to the NCCN guideline of DLBCL. Although the reported prognosis of IVLBCL is very poor, the patient has survived for 4 years without disease progression until now. Therefore, timely and accurate diagnosis as well as the appropriate treatment may improve the clinical outcome.

In conclusion, primary IVLBCL of the prostate is rare and clinically difficult to distinguish from benign prostatic hyperplasia and prostate carcinoma as it occurs in the same age group of 6th decade and presents with the similar obstructive urinary
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