

Case Report

Primary T-cell Non – Hodgkin lymphoma of the Urinary Bladder Presenting as Haematuria and Bilateral Hydronephrosis: Report of a Rare Case

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Abstract

Introduction: Primary T cell lymphoma of the urinary bladder is extremely rare disease. Most of the lymphomas of urinary bladder are B-cell lymphomas. We encountered a case of T-cell lymphoma of the urinary bladder presenting as haematuria and bilateral hydronephrosis.

Presentation of Case: A 42-year-old man was admitted to our hospital because of intermittent painful gross haematuria associated with bilateral loin pain and vomiting lasting for 3-4 weeks. Results of his general examination were normal. An ultrasonographic study and Computed Tomography (CT) of his abdomen and pelvis revealed bilateral gross hydro-ureteronephrosis with significantly thickened urinary bladder wall with large circumferential growth involving the entire bladder occluding both ureteric orifices with internal calcification. A cystoscopy procedure with Trans Urethral Resection of Bladder Tumour (TURBT) was performed. Postoperative microscopic examination and immunohistochemistry showed medium to large lymphocytes with hyperchromatic nuclei and strong expression of CD3 in the tumour cells and negativity for CD20 and cytokeratin. A diagnosis of T-cell non Hodgkin Lymphoma was entertained.

Conclusion: Presentation of bilateral hydronephrosis with primary T cell lymphoma has not been reported previously. This sort of presentation can mimic other bladder malignancy and present a diagnostic challenge. An ancillary technique, Immunohistochemistry in our case, helped us arrive at the correct diagnosis and subsequent appropriate management.

Keywords: Lymphoma; Urinary bladder; T cell; Bilateral hydronephrosis; Immunohistochemistry

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Introduction

Primary T cell non Hodgkin lymphoma of urinary bladder is a very rare tumour; lymphoma account for 0.2% of all bladder cancer [1]. Most reported lymphomas are of low grade B cell type of non

Hodgkin lymphoma (NHL) and women are affected more frequently than men [2-4]. Unlike the Hodgkin lymphoma, NHL may appear as nodal or extra nodal disease; most common sites of extra nodal primary

NHL are stomach, intestine, skin.⁵ Urinary bladder as primary extra nodal site of NHL is extremely rare particularly of T cell type.

Presentation may vary from asymptomatic frank haematuria, urinary tract infection, dysuria, increased frequency, loin pain from hydronephrosis [4-7]. Diagnosis is done based on immunohistochemical study in absence of any evidence of nodal or extra bladder involvement [4,7]. We encountered a case of primary T cell NHL in a 42 year male presented as haematuria with loin pain, the diagnosis of NHL with immunohistochemistry in absence of extra bladder involvement (by PET scan) help in planning for further management.

Case presentation

42 years Middle Eastern male presented to us with complained of intermittent painful gross haematuria associated with bilateral loin pain and frequent episodes of nausea and vomiting for about 3-4 week. Apart from essential hypertension diagnosed 3 year ago, patient did not have any significant medical present or past medical or surgical history. Apart from mild pallor, general examination was unremarkable.

Systemic examination did not reveal any gross abnormality (no organomegaly or lymphadenopathy). Routine haematological analyses including coagulation were normal except for mild hypochromic microcytic anaemia (Hb-9.8 gm/dl); biochemical and electrolyte evaluations were normal except for mildly elevated Blood Urea Nitrogen (BUN) 23.5 mmol/dl and creatinine 186 mmol/dl.

Ultrasonographic imaging of abdomen and pelvis was done as initial radiological evaluation which revealed bilateral hydronephrosis with hydroureter and thickened bladder wall. A detailed radiological evaluation with contrast enhanced Computed Tomography (CT) revealed large circumferential irregular growth involving the entire bladder and both ureteric orifices with internal calcification, peri-vesical infiltrations and bilateral gross hydro-ureteronephrosis without any lymph node or hepato-splenomegaly, CT chest was unremarkable. We proceeded with contrast enhanced CT despite mildly elevated creatinine as this was a obstructive uropathy and we had planned for relief of obstruction and as our hospital policy we do take patient with mildly elevated creatinine for contrast study with proper hydration and renal protective strategy.

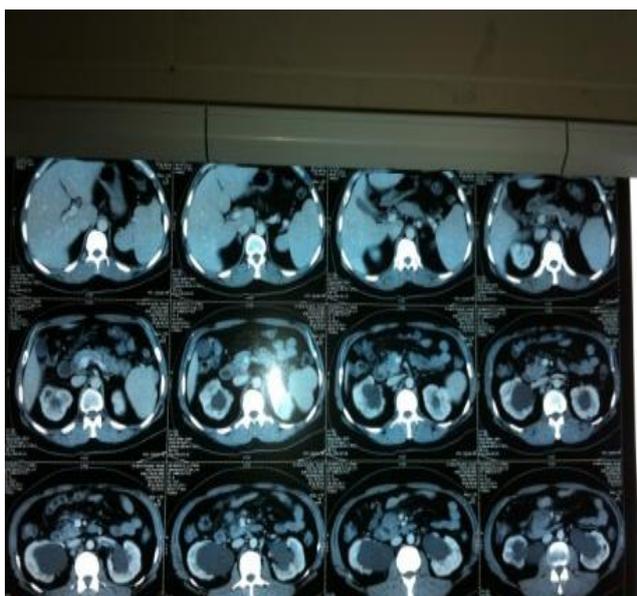


Figure 1(Left) & 2(Right) CT scan showing bilateral hydronephrosis and thickened bladder wall

Transurethral Cystoscopic examination and placement of bilateral double J stent (DJ) with biopsy was planned. Cystoscopic examination revealed irregular thickened bladder mass involving both the ureteric orifice; Trans urethra resection of the bladder mass was performed followed by placement DJ stent. Histopathology examination showed diffuse, dense infiltration of lamina propria by medium to large atypical lymphocytes with pleomorphic hyperchromic nuclei. Immunohistochemistry showed strong expression of CD3 in the tumour cells and negativity for CD20 and cytokeratin. Further investigation including chest X ray, CT chest, Bone Scan and bone marrow biopsy were completely normal. A diagnosis

of Primary T-cell non Hodgkin Lymphoma was entertained. Patient has been referred to oncology centre for chemotherapy. Patient had received 4 cycle of chemotherapy with CHOP regime; (cyclophosphamide, doxorubicin, vincristine and prednisolone). The DJ stent were removed after 2 month. The patient is in regular follow up in urology and oncology follow up and is clinically free of disease 6 month after the biopsy. This indexed case has been highlighted for its rarity and unusual histological appearance. An ancillary technique, Immunohistochemistry in our case, helped us arrive at the correct diagnosis and subsequent appropriate management.

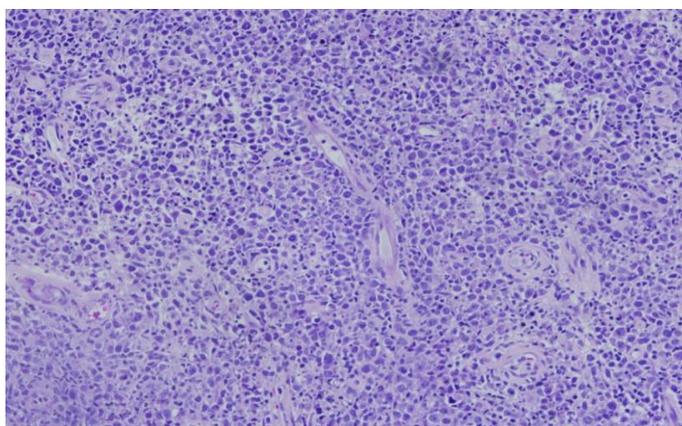


Figure 3 Microscopy of the tumour cell

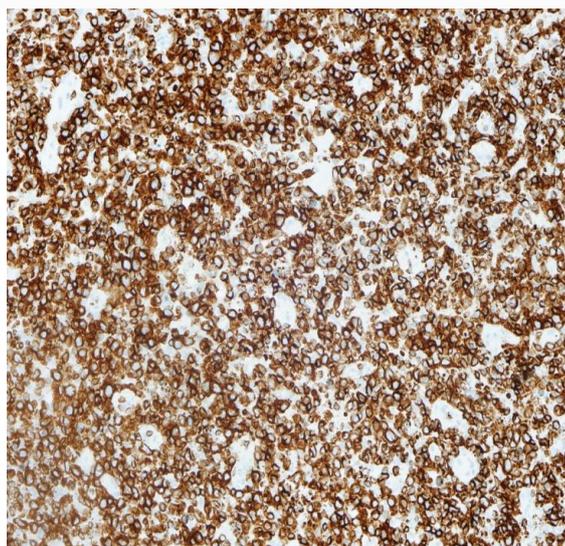


Figure 4 Immunohistochemistry of the tumour cell showing CD3 positivity

Discussion

Primary bladder lymphoma is a very rare tumour, clinical diagnosis is based on absence of evidence of extra bladder involvement in any tissue, lung, lymph node, intestine, skin or bone marrow. Most common subtypes of primary lymphoma in urinary bladder are Mucosa Associated Lymphoid Tissue (MAST) Lymphoma and large B cell type of NHL [7-9]. History of chronic cystitis has been associated in 20% cases of primary lymphoma [2, 9]. There are only few case reports of primary T cell lymphoma of urinary bladder (5 cases from English journal including the present case); contrary to the B cell lymphoma, primary T cell lymphoma might be more common in male, 4 out of 5 cases were male [7, 9-12].

Hughes et al reported in a 27 male with primary T cell lymphoma of bladder that presented with haematuria and left iliac fossa pain; surgical excision followed by chemotherapy resulted in complete cure [12]. Choie et al reported another case of calcified primary T cell lymphoma of bladder in a 30 yr female who presented as haematuria and dysuria for which no treatment could offered and patient died without any definitive treatment [11]. In another case report by Murad et al; a 52 yr male presented as haematuria and suprapubic pain for which primary chemotherapy was given but there was no post chemotherapy follow up [10]. Wang L et al encounter a case of primary T cell lymphoma in a 45 yr male who presented as haematuria and loin pain; treated with transurethral resection followed by chemotherapy resulting in complete cure for 1 year.

Haematuria was common in all the cases; our case differs from the previous cases in presentation, our case presented as bilateral hydro-uretero nephrosis in addition to haematuria which is very uncommon presentation of primary lymphoma. None of the case report so far has reported bilateral hydronephrosis in primary T cell bladder lymphoma.

The diagnosis of T cell lymphoma is not easy on routine histopathology, we diagnosed T cell lymphoma based on presence of atypical lymphocytes with hyperchromic nuclei and immunohistochemistry where these lymphocytes express strong positivity for CD 3

and negativity for CD20 and cytokeratin.

Other rare primary bladder tumors include perivascular epithelioid cell tumor [13], pseudosarcomatous myofibroblastic lesion [14], primary small cell bladder carcinoma [15] which not only poses diagnostic challenges but also the therapeutic intervention because of absence of sufficient literatures and evidences for such rare tumours. Histology, immunochemistry and genetic studies are helpful to make a diagnosis. The treatment of rare bladder tumour remained undefined [13-15].

Treatment of the primary T cell bladder lymphoma is not yet defined being the rarity of the tumour, surgery, chemotherapy, radiotherapy or combination of therapies are all treatment option. Chemotherapy is often preferred in the primary bladder lymphoma as these lymphomas are sensitive to chemotherapy and chemotherapy also treat any undetectable early systemic disease [7, 16]. Surgical approach can be beneficial to relief unpleasant urinary symptoms such as obstructive uropathy.

Conclusion

Primary bladder lymphoma is rare disease 0.2% of all bladder neoplasm, most cases are of B cell type; our case was unique in the sense that it was T cell lymphoma which is extremely rare. Presentation of bilateral hydronephrosis with primary T cell lymphoma has not been reported previously. This sort of presentation can mimic other bladder malignancy and present a diagnostic challenge.

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