Primary Lymphoma Of Urinary Bladder: Report Of An Unusual Case And Literature Review

A Sharma, A Singh

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Abstract

Primary lymphoma of the bladder is a rare entity. We report a case with a long term follow up in which a initial biopsy diagnosis of a highly malignant tumour, probably sarcoma was made. The role of cystitis of long duration in pathogenesis, involvement of ureteric orifice and brief clinical, pathological and therapeutic aspects of the disease are also reviewed. We would like to point out the involvement of ureteric orifice which is considered rare but was present in oir case. The initial diagnosis of sarcoma and our long term follow up also needs to be looked at.

INTRODUCTION

Primary lymphoma of the urinary bladder is rare (less than 70 cases have been reported in literature) $\binom{1}{2,2,3,4}$. This was first described by Eve and then by Chaffey in 1885 (1, 4).

Primary bladder involvement by Non Hodgkins Lymphoma is usually seen in elderly women who often present with a history of chronic cystitis (1, 4). Obstruction of the ureter with hydronephrosis is uncommon since the tumour typically does not involve the ureteric orifice/s. (1, 4) Involvement of the entire bladder wall is also a rare phenomenon.

CASE REPORT

A 63 year old woman presented to the surgical clinic on 14th February 1989 with pain in the suprapubic region of 7 years duration and painless haematuria with difficulty in passing urine for a year. In the last seven years she had had multiple episodes of cystitis with positive urine culture.

Physical examination revealed a firm, non tender, smooth, immobile mass of 10 x 7 cms in the suprapubic region. No other abnormality was found in the neck, chest or abdomen. The lump was not arising from the uterus or adnexa.

Haematological and biochemical investigations were normal except blood urea which was raised to 8.5 mmol/l. Urine culture was positive for coliform bacilli and urine cytology showed dyskaryotic transitional cells in an inflammatory background. Ultrasound demonstrated the presence of numerous mass lesions arising from the entire mucosal

surface of the bladder with occlusion of the left ureteric orifice, resulting in gross hydroureter and hydronephrosis on that side. The right kidney also showed a mild degree of hydronephrosis. There was no evidence of other organ involvement or retroperitoneal masses. The findings were reported as strongly suggestive of a bladder carcinoma. Intravenous urography confirmed the above findings.

Cystoscopic examination showed multiple pedunculated sub-mucosal tumour masses involving the entire bladder surface. There was no ulceration of the overlying mucosa. Transurethral biopsy of the bladder tumour was done and the histology was reported as 'a highly malignant tumour possibly a sarcoma'. In light of the diagnosis, a cystectomy and hysterectomy was done with an ileal conduit. This was followed by a radical course of radiotherapy.

CT scan done after 15 days and 9 months after operation were normal. The patient has continued to remain disease free and is on regular follow up. When she was seen last on 31st October 2000 (11 years follow up) there was no evidence of tumour recurrence or metastasis.

PATHOLOGY

The entire mucosal surface of the bladder was involved by multiple polypoid masses varying in size from 1-5 cm in diameter. The overlying mucosa was thin and haemorrhagic. Cut sections showed firm, homogenous areas with focal haemorrhage. Microscopic examination showed a Non Hodgkins lymphoma involving the full thickness of the bladder wall.

Figure 1

Figure 1: Pathologic specimen



DISCUSSION

Although secondary involvement of the genitourinary tract is occasionally seen ingeneralised lymphomas, primary lymphoma of the urinary bladder is extremely rare (1,2,4). The tumour comprises 0.2% of all bladder neoplasms (1,5). The majority of the affected patients, like the present one are elderly females in the fifth to seventh decades.

The most common symptom is periodic haematuria of variable intensity associated with frequency and dysuria occurring in 77% of patients in contrast to only 12% of patients with other bladder malignancies (1,4) Associated urinary tract infection occurs in about 50% of patients. (6)

Development of primary lymphoma of the bladder is controversial as it is documented that no lymphoid tissue is normally present in the bladder.

However, it is now agreed that lymphoid tissue can develop in bladders affected by chronic cystitis and this can give rise to the development of lymphoma (4).

Nevertheless a number of cases have been reported with absence of chronic cystitis and here it is postulated that the tumour may originate in the lymphoid tissue derived from the embryonic cloaca (5).

The knowledge of radiographic findings in bladder lymphoma is limited. A radiological diagnosis is possible but a definite distinction cannot be made between Transitional Cell Carcinoma and Lymphoma.

Obstruction with hydronephrosis is uncommon since the tumour typically does not involve the ureteric orifice (4) whereas in this case there was left ureteric occlusion with hydroureter and hydronephrosis.

Cystoscopic appearance of a polypoid or sessile sub-mucosal mass projecting in the lumen is quite presumptive of the diagnosis but a mistaken diagnosis ofunusual saracoma or undifferentiated carcinoma may be made and a biopsy is required for the definitive diagnosis. (1,4) Heamatoxylin and Eosin sections may be unable to differentiate lymphoma from sarcoma or undifferentiated carcinoma and immunohistochemical staining techniques need to be used. (4,6). The overlying mucosa is intact although occasionally thinning or superficial ulceration may be present (1,4).

Treatment includes total or partial cystectomy, external beam radiation, chemotherapy or a combination of these. Prognosis with adequate treatment is excellent. $(4,_7)$ Aggressive local therapy produces long term survival as the tumour remains localized for considerable periods of times and metastasis is a late event. (4,7).

Due to high radiosensitivity of most lymphomas it has become the treatment of choice although surgery too gives good results. Radiotherapy offers advantages over surgery as it is not associated with urinary diversion, has less mortality and is useful in disseminated disease. (4, 7)

CORRESPONDENCE TO

Dr Abhiram Sharma, 16 St Abbs Close, Victoria Docks, Hull, HU9 1TX UK

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Author Information

A Sharma, MBBS. M.S

Clinical Observer, Department of Surgery, Prince Charles Hospital

A B Singh, FRCS

Associate Specialist in General Surgery, Department of Surgery, Prince Charles Hospital