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Case Report
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Primary Plasmacytoma Involving Kidney, Ureter & Urinary Bladder

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Abstract

Primary plasmacytoma of urinary tract is extremely rare clinical entity, presenting diagnostic challenges due to its paucity of evidence. We are presenting a case of primary plasmacytoma of kidney, ureter & urinary bladder in a 74-year-old female patient. Computed tomography (CT) revealed that the mass was in the pelvis of the right kidney invading the renal vein, ureter & urinary bladder along with enlarged right paracaval lymph nodes. After optimization, radical right nephroureterectomy done with excision of cuff of bladder mucosa and the retroperitoneal lymph nodes was resected. Histopathological examination revealed plasmacytoma involving renal parenchyma, ureter & bladder with lymph nodes, which was confirmed by immunohistochemistry studies. Following this unexpected diagnosis, various examinations were performed, but there was no evidence of systemic plasma cell disease. Although the general prognosis and outcome of extra medullary plasmacytoma (EMP) is good, regular follow-up is recommended due to the possibility of relapse or progression to plasma cell neoplasm (PCN). This case is presented for reference, as it is imperative to keep plasmacytic tumours in mind and to include them in the differential diagnosis of anaplastic tumours, even in unusual location, such as the urinary system.

Introduction:

Extramedullary plasmacytoma (EMP) is malignant neoplasm that develops due to uncontrolled proliferation of plasma cells and monoclonal plasmacytic infiltration.[1] They may arise from soft tissue (extramedullary) or within the skeleton. The majority of EMPs are detected in the head and neck regions 80-90%, upper respiratory airways, digestive tract, pleura, lung, lymph nodes, skin, subcutaneous and soft tissue, testicle and liver with more than 80% situated above the diaphragm.[2,3] Occurrence of EMP in the kidney is extremely rare & has been rarely described in the literature.[6] EMP typically affect patients during middle age (median 55-60 years) and are more common in males (Male: Female Hd 3:1) and constitutes about 3% of all plasma cell tumours.[4] Risk factors for plasmacytomas remain unknown; however, prior radiation exposure has been suggested.[5] No clear therapeutic guidelines are available due to its rarity.

Case Report:

A 74-year-old female was presented to the urology OPD of Evercare Hospital Dhaka with the complaints of lower urinary tract symptoms (LUTS) including urinary frequency, urgency and occasional hematuria for last 6 months. She had a history of Recurrent UTI. She was also a known case of right renal calculus and bilateral renal cyst & CKD. She had Haemoglobin E Trait & her family history was non-contributory. CT Urogram revealed mildly enhancing irregular soft

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tissue mass in the right renal pelvis measuring about 4.3 x 4.2 cm and invading the renal hilum. Right ureter was dilated with wall thickening and right paracaval lymph nodes were enlarged, largest one measuring about 22x10 mm. Urinary bladder was smaller in size with irregular thickening of the wall having a maximum thickness of 13mm. Findings were in favor of Transitional Cell Carcinoma (TCC) involving right renal pelvis, ureter and urinary bladder. Following preparation, Right Nephroureterectomy with excision of cuff of bladder was done along with paracaval lymph node dissection. Her post operative period was uneventful. The patient was discharged on 8th POD with OPD follow up advice. Histological examination of the resected specimen (Right Kidney, Ureter and Cuff of Urinary Bladder) revealed a focus of sheets of Plasma cells (Plasmacytoma) including atypical plasma cells in the renal pelvis, ureter and bladder wall. Following this unexpected diagnosis, Immunohistochemical analysis was done which demonstrated that the tumour cells were positive for CD138, MUM-1, KAPPA and LAMBDA. There was no light chain restriction. Feature was compatible with reactive plasmacytosis. Patient was further investigated for plasmacytoma of other sites. Bence jones protein was absent in urine and serum protein electrophoresis showed polyclonal rise of gamma globulin. Bone marrow biopsy revealed Hypercellular marrow consistent with secondary reactive changes. She is under surveillance now.

Discussion: Extramedullary plasmacytoma (EMP) is a solitary tumour composed of proliferated cells with plasmacytic differentiation in an extramedullary site. EMP are uncommon tumour with worldwide annual incidence of 3 per 100,000 population. Solitary EMP account for less than 3-10% of all plasma cell neoplasm.[7] Primary plasmacytoma characterized by strictly localized growth and no systemic manifestation of plasma cell malignancy.[9] The most common plasma cell malignancy is multiple myeloma (MM) which demonstrates broader systemic involvement including bone marrow involvement and paraproteinemia.[10] Plasmacytomas are clonal proliferation of plasma cells that are cytologically and immunophenotypically indistinguishable from MM. The majority of EMPs involve the head and neck region, particularly the upper respiratory tract.[2] Others involve diverse anatomical sites, including the gastrointestinal tract, central nervous system, thyroid, breasts, parotid gland, lymph nodes, skin, lungs, pleura, muscle, liver, spleen and pancreas.[11,12] Primary EMP involving urinary tract is rare clinical condition with just two dozen cases reported in the literature. Renal plasmacytomas could mimic renal cell carcinomas (RCC) of the kidney or even transitional cell carcinomas (TCC) of the kidney.[3]

The diagnosis of an EMP is complex and requires radiological, haematological, biochemical and histological investigation. Primary renal plasmacytoma are not distinguishable from other renal tumours in pre-operative imaging tests. In this patient, EMP was diagnosed based on presence of diffuse monoclonal plasma cell infiltration at a single site in the immunohistochemical staining for the k and A light chains and following the exclusion of a diagnosis of MM.[2,9] The diagnosis of localized plasmacytoma can be made only after detailed clinical, biochemical, haematological & radiological investigations. Radiological findings in renal plasmacytoma are indistinguishable from those of renal cell carcinoma and tissue biopsy showing monoclonal plasma cell histology without any evidence of Multiple Myeloma is the only way of diagnosis.[13] In this case, histopathology revealed plasmacytoma, thus immunohistochemical staining was carried out which showed monoclonal immunoglobulin. Following IHC markers were applied: CD138, MUM-1, CD56, CD45, GATA-3, PAX 8, CK, KAPPA, LAMDA and the plasma cells expressed CD138, MUM-1, KAPPA & LAMBDA. No light chain restriction is found. Features are compatible with plasmacytosis. Bone marrow biopsy, Urinary Bence Jones Protein, Serum protein electrophoresis ruled out systemic disease and confirmed the diagnosis of primary renal plasmacytoma.

As a result of the lack of typical clinical symptoms and evidence from specific laboratory tests, a diagnosis may be delayed. In the present study, the patient was initially misdiagnosed with right upper tract TCC with paracaval lymphadenopathy. Later, post-operative histopathology confirmed the final diagnosis.

To the best of our knowledge, there is no specific guidelines for the treatment of renal plasmacytoma. Treatment options for renal plasmacytomas include surgery, chemotherapy and radiotherapy, either alone or in combination. Combined therapy may provide the best results.[14] Local radiotherapy is the preferred therapeutic modality for EMP owing to its documented radiosensitivity. EMP has a relatively good prognosis,

but local recurrence and metastasis develop in 30% and 40% of patients, respectively. The five-year survival rate of EMP is excellent at 90% and previous data suggest that local progression does not necessarily indicate a worse prognosis.[15]

The periodic evaluation of patients with EMP is necessary due to the possibility of relapse and progression to MM.[16] Physical examinations coupled with laboratory tests, including CBC, renal function tests, analyses of blood calcium, serum albumin and immunoglobulin levels, serum protein electrophoresis, free light chain assays and radiographic studies of the skeleton are required for follow-up.

Renal involvement is frequent in the setting of Multiple Myeloma (MM) mainly due to myeloma cast nephropathy. Diagnosis of EMP depends upon the demonstration of extramedullary plasma cell tumour with no evidence of systemic signs and symptoms associated with MM.

It is important to note that plasmacytoma may be composed of pleomorphic cells with little resemblance to normal plasma cells. Therefore, it is important to include plasmacytoma in the differential diagnosis of anaplastic tumours, even in usual locations, such as the kidney.

Conclusion:

This case represents a rare example of extramedullary plasmacytoma of renal origin. A review of the literature shows that there is currently not widely established standard treatment for EMP of the kidney. Surgery in combination with radiotherapy may be the best treatment. Although general prognosis & outcome is good, regular follow up is recommended due to the possibility of relapse or progression to plasma cell neoplasm.

Conflict of interest: None

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