Primary Retroperitoneal Transitional Cell Carcinoma – A Rare Clinical Entity

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INTRODUCTION

Transitional Cell Carcinoma (TCC) most commonly arises from the urinary bladder and less frequently from the upper urinary tract. We report a case of primary retroperitoneal TCC in a male patient. The entire urinary tract did not have any evidence of tumour. As far as we are concerned, only four such cases have been reported in the literature previously[1] and all of them were female.

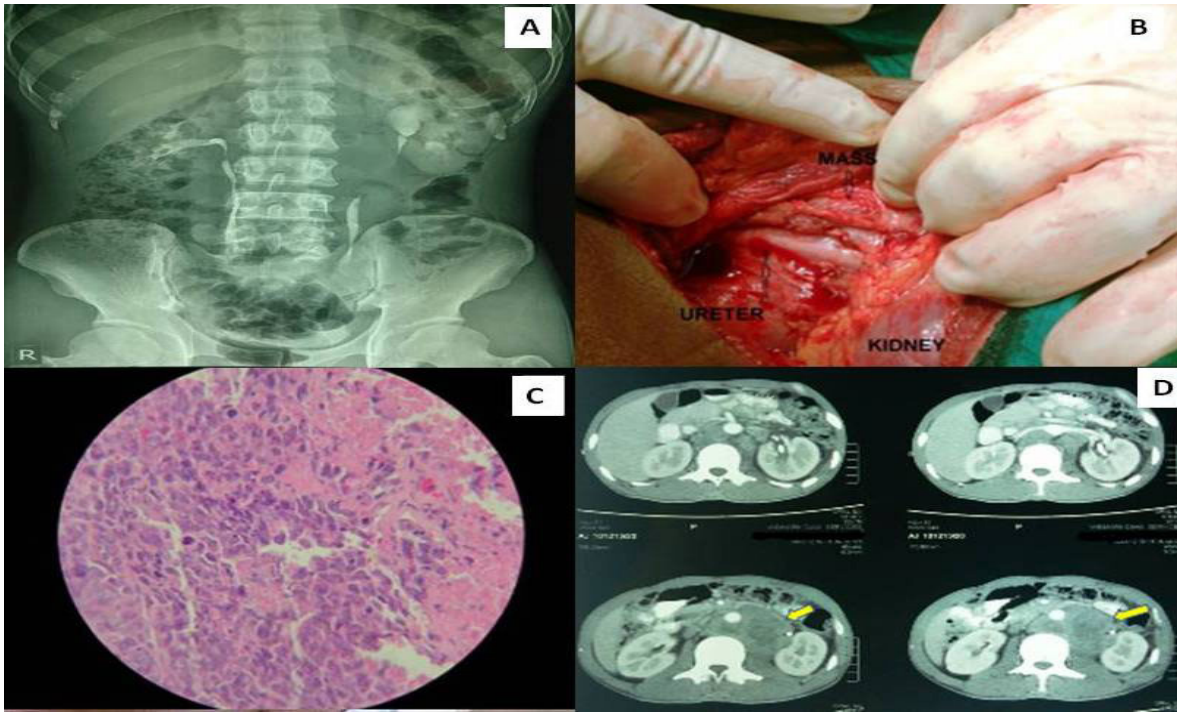

Figure 1. (A) Intravenous urography revealing hydronephrotic left kidney suggestive of pelvi-ureteric junction obstruction. **(B)** Intraoperative image showing a large retroperitoneal mass with lateral deviation of ureter. **(C)** Hematoxylin & Eosin stained picture of incisional biopsy of retroperitoneal mass showing transitional cell carcinoma under 400X magnification. **(D)** Computed Tomography scan of abdomen showing mass encasing the great vessels.

CASE DESCRIPTION

A 30 year-old male patient belonging to Asian-Indian race presented with intermittent left flank pain for 2 months with normal abdominal examination findings. The patient was a farmer by occupation and did not have any history of radiation exposure or any known family history of malignancy. Ultrasonography along with intravenous urography (IVU) (**Figure 1A**) revealed hydronephrotic left kidney suggestive of pelvi-ureteric junction obstruction (PUJO) and diuretic renogram showed obstructive drainage from the left kidney. However, on careful retrospective evaluation of IVU plates there was evidence of upper ureteric compression with mild lateral deviation of left ureter which we missed preoperatively. With preoperative diagnosis of left PUJO, the patient was taken up for pyeloplasty and on table retrograde pyelogram (pyelogram) exhibited patent ureter with jet sign at pelvis which collabo-

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rated with our diagnosis. On exploration, a large mass was found in the retroperitoneum which was encasing the great vessels (**Figure 1B**). The ureter was arching over the mass with no evidence of primary PUJO. Incisional biopsy from the mass was taken and double-J stent was inserted into left ureter via ureterotomy. Histopathological examination showed multiple pieces of greyish tissue on gross examination, and microscopically it was suggestive of high grade urothelial carcinoma with area of necrosis and perineural invasion (**Figure 1C**). Considering that the mass could be a secondary from an unknown primary site within the urinary tract, Computed Tomography (CT) urogram, cystoscopy, urine for malignant cell cytology, bilateral ureteroscopy and retrograde pyelography were performed, but no evidence of urothelial carcinoma was found in the urinary tract. CT of abdomen showed a large mass encircling the aorta and compressing the inferior vena cava with areas of necrosis (**Figure 1D**). With the final diagnosis of primary retroperitoneal TCC, chemotherapy with Gemcitabine and Cisplatin was started after consultation with medical oncologists. Patient did not respond to the chemotherapy and his general condition deteriorated. Moreover, a swelling was noticed in left side of neck after first cycle of chemotherapy. Fine needle aspiration cytology of that mass was suggestive of metastatic deposits of malignant epithelial neoplasm. Finally, 5 months after diagnosis he succumbed to death.

DISCUSSION

Adult retroperitoneal tumours are rare and constitute only 0.2-0.5% of all malignant tumours [(2)]. They may arise from fat, areolar connective tissue, fascia, muscle, vascular tissue, nervous tissue, lymphatic tissue and urogenital ridge remnants. The two most common malignant primary tumours of the retroperitoneum are lymphosarcoma and liposarcoma [(3)]. Amongst the retroperitoneal tumours, primary TCCs are extremely rare and carry a worst prognosis. Till date, only 4 cases have been reported and all of which were cystic pelvic mass occurring in female patients [(1)]. Like ours, three out of those four patients had unresectable mass. This is the first case reported in male as per our best knowledge. Retroperitoneal tumours grow unrestricted because of loose surrounding tissue. They are generally asymptomatic until they grow to very large sizes. The symptoms are often due to obstruction or compression of the adjacent structures. As they grow to a very large size before being detected, only 25% of these tumours can be completely excised [(4)]. Primary retroperitoneal TCC is an extremely uncommon entity. They are thought to originate from the remnants of urogenital ridge[(5)]. Braasch et al.[(6)] reviewed 101 cases of retroperitoneal tumour of which only 3 cases were found to arise from urogenital ridge. However, Handfied-Jones et al.[(7)] were of the opinion that majority of retroperitoneal tumours arise from urogenital apparatus. Hansmann et al.[(8)] also supported their view of similar origin. Koyanagi et al.[(9)] were first to report primary TCC of retroperitoneum arising from cloacal cyst. Later Gupta et al.[(4)] and Basu et al.[(2)] also reported primary retroperitoneal TCC. All these above 3 cases died within 2 years of diagnosis. Radiotherapy alone was used in 2 cases, while surgery was the primary modality of treatment in one of them followed by adjuvant chemotherapy. Recently, Ichinobe et al. (1) reported a case of primary retroperitoneal TCC with complete remission of the disease achieved with radiotherapy and concurrent oral chemotherapy (tegafur-uracil). The explanation they gave for their success was that the chemotherapeutic agent acted as a radiosensitisor. In our case, the patient died after 5 months with development of a metastatic neck deposit. To conclude, primary retroperitoneal TCC is an exceptionally infrequent neoplasm. Due to retroperitoneal location they grow to very large size before being detected. Modality of therapy has not been yet defined due to their rarity. Prognosis is poor as most of the patient die within months.

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