

A ureteral mass presenting as deep vein thrombosis: A very rare presentation

Dushiant Sharma¹, Umesh Sharma¹, Rajeev Sood²

From ¹Assistant Professor, ²Professor and Head, Department of Urology & Renal Transplant, Dr RML Hospital & PGIMER, New Delhi, India.

Correspondence to: Dr. Dushiant Sharma, Department of Urology & Renal Transplant, Dr RML Hospital, New Delhi-110001, India. E mail: dushiant@gmail.com

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ABSTRACT

Deep Vein Thrombosis (DVT) is a dreaded condition, caused by an interplay of a triad of factors consisting of hypercoagulability, vascular endothelial dysfunction, and stasis. Stasis can lead to the development of DVT due to mass external compression over pelvic veins. Here, we present the case of a 45-year-old female who presented with sudden onset left lower limb swelling who was diagnosed to have left iliac venous thrombosis caused by external compression due to an advanced ureteric malignancy underlining the need for a thorough evaluation in such patients to reveal a more sinister pathology.

Keywords: Deep vein thrombosis, Iliofemoral thrombosis, Upper tract urothelial cancers, Ureteric malignancy.

Every year, roughly 10 million new cases of Venous Thromboembolism (VTE) occurs [1]. Many possible causes and factors predispose to Deep vein thrombosis (DVT) and VTE. All of these have been known to affect one or more factors in Virchows Triad [2] consisting of hypercoagulability, vascular endothelial dysfunction, and stasis. Stasis is responsible for the development of DVT due to direct compression. Iliofemoral thrombosis in malignancy patients can be caused due to vein compression by pelvic malignancy or enlarged malignant lymph nodes and usually presents as unilateral lower limb swelling. This can rarely be the only presenting symptom of a malignancy. Therefore, in patients with sudden onset of unilateral lower limb swelling without any perceived or diagnosed medical condition, as seen in our case, a detailed evaluation may lead to early identification, appropriate management and possibly cure of pelvic or abdominal malignancies. Early detailed evaluation in our case has led to the early diagnosis and treatment of ureteric malignancy.

CASE REPORT

A 45-year-old post-menopausal female presented to her primary physician with complaints of the left foot swelling for 3 weeks which gradually progressed to involve the entire left lower limb. She did not give any antecedent history of trauma, ulceration on lower limb, hematuria, lower urinary tract symptoms, bowel complaints, vaginal bleeding, and loss of weight, loss of appetite and previous history of DVT. There was no history of previous surgeries and coexisting morbidities.

On general examination, her vitals were stable. There was no pallor, icterus, cyanosis, clubbing, and lymphadenopathy. Systemic examination was within normal limits. Her left lower

limb was reddish in color, tense and tender to touch. Per-vaginal examination revealed induration within the left fornix.

A venous Doppler revealed a partial thrombus of the left proximal external iliac vein and left common iliac vein. Thrombophilic screen (prothrombin time-International Normalized Ratio, activated partial thromboplastin time, Protein C, Protein S, D, Factor V Leiden Mutation) was within normal limits.

The patient was started on warfarin and put on total bed rest by her primary physician. Due to the absence of any predisposing factors, further evaluation was considered and an ultrasound of the abdomen was done. It revealed a grossly dilated left renal pelvis with thinned out parenchyma and a 1.3x1.1cm lesion near the left vesicoureteric junction (VUJ). Contrast-Enhanced

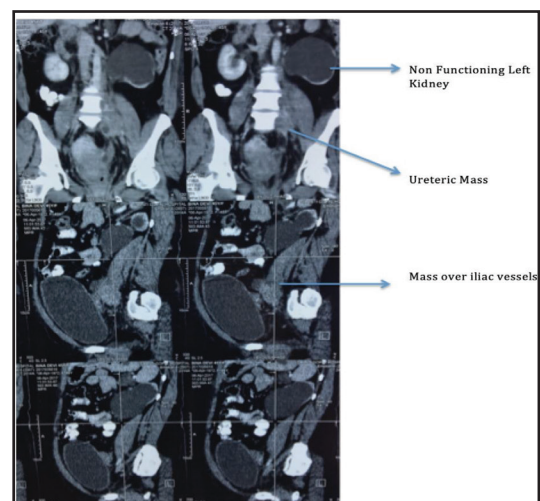


Figure 1: Preoperative CECT of the abdomen and pelvis (coronal and sagittal sections)

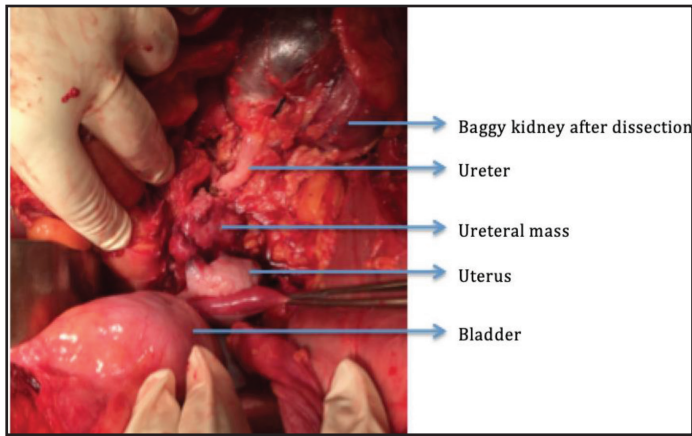


Figure 2: Baggy kidney with Ureteric mass. Bladder retracted in the lower part of photo

Computed Tomography (CECT) of the abdomen and pelvis (Fig. 1) was done which showed poorly functioning left kidney with gross hydronephrosis, thinned out parenchyma, a large 6.6x4.5cm heterogeneously enhancing solid cystic mass in the left ureter at the level of the iliac vessel crossing and 3.5x2.1cm heterogeneously enhancing solid cystic lesion in the left distal ureter extending upto VUJ. Her hemoglobin, serum creatinine, liver function tests, urinalysis were within normal limits. Urine for malignant cytology was negative. Cystoscopy showed a bulge just lateral to the left ureteric orifice and no other visible mass. The left ureteric orifice could not be cannulated.

Considering the diagnosis of a ureteric mass, a left nephroureterectomy with bladder cuff excision was planned. Warfarin was stopped 5 days before surgery and the patient was put on bridging therapy with low molecular weight heparin. A cardiology consult was taken and it was decided to insert an Inferior Vena Cava (IVC) filter 24 hours before surgery as prophylaxis to decrease the risk of intraoperative and postoperative pulmonary embolism. Intraoperatively, DVT pump was not applied. A radical nephroureterectomy along with hysterectomy and the left salpingo-oophorectomy (Fig. 2 and 3) were done with gynecological assistance in view of local invasion of the cervix. Mid-ureteric mass was dissected off the iliac vessels with cardiothoracic and vascular surgery assistance and final specimen consisted of the left kidney, ureter, uterus, left ovary along with lymph node dissection. Total intraoperative blood loss was 600ml. Postoperatively, the patient was kept in the ICU before shifting to the ward. Warfarin was re-started on day 3. The patient had a relatively uneventful post-operative period.

Histopathological examination revealed a high-grade Urothelial Carcinoma of the ureter with focal squamous differentiation with invasion into cervix and uterus. Lymph nodes were free of tumor. The final stage was pT4 N0 Mx. The patient was given 3 cycles of adjuvant chemotherapy till the time of abstract submission (Gemcitabine+Cisplatin) and was doing well at final follow-up with a resolution of DVT. Follow-up CECT abdomen+pelvis is planned but the patient is yet to follow-up despite repeated reminders.

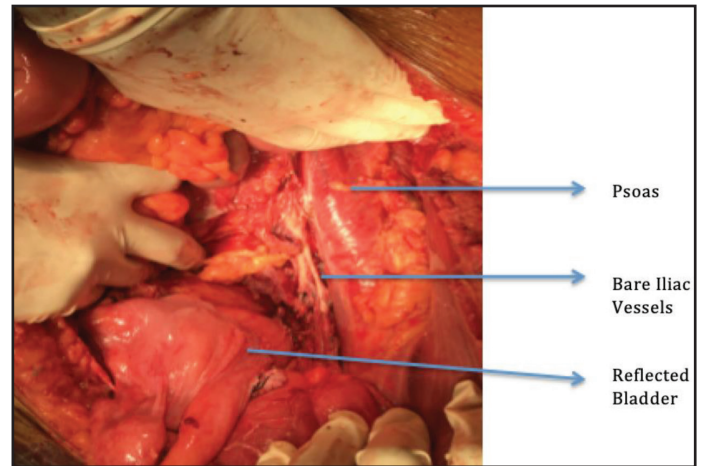


Figure 3: Tumor bed with exposed iliac vessels after removal of Left Nephroureterectomy and Left oophorectomy

DISCUSSION

Upper tract urothelial carcinomas are a rare entity comprising only 5-10% of all urothelial tumors³ but recent literature reveals an increasing incidence. Ureteral tumors occur more commonly in the lower than in the upper ureter. Overall, about 70% of ureteral tumors occur in the distal ureter, 25% in the mid-ureter, and 5% in the proximal ureter [4,5]. The most common clinical feature of urothelial carcinoma of the ureter is hematuria, either gross or microscopic, present in roughly 56-98% cases [6,7,8,] followed by flank pain in 30% cases.

Urothelial carcinoma of ureter presenting as unilateral lower limb swelling secondary to DVT is very rare. VTE risk factors have been classified as modifiable, non-modifiable and temporary [9]. Modifiable risk factors include obesity. Non-modifiable risk factors include genetic and familial disorders resulting in a hypercoagulable state. Temporary risk factors include hospitalization, malignancy, long distance travel, prolonged immobilization, hormonal therapy for various diseases. But most of the DVT is still labeled as 'idiopathic' without any known antecedent cause.

Malignant compression of iliofemoral veins in the pelvis by malignancies is one such cause in which patients may be mistakenly labeled as having idiopathic DVT with potentially disastrous outcomes. Ureteric masses causing direct ilio-femoral venous compression with resultant DVT and unilateral lower limb swelling have seldom been reported. Most of the reported cases of iliofemoral venous compression with resultant DVT have been due to compression by enlarged lymph nodes secondary to Urothelial or gynecological malignancies.

Liao et al [10] analyzed 893 cancer patients over a 10 year period. The inclusion criteria for their study were the presence of unilateral lower limb swelling, a CT showing malignancy causing external compression on iliac or femoral vein and a duplex ultrasound revealing vein thrombosis or venous flow insufficiency over a femoral vein or saphenous vein. The patients with bilateral lower limb edema suggestive of IVC involvement were excluded. Approximately, 63(8%) of these patients were diagnosed with iliofemoral venous obstruction, 21 of these 63 patients developed

iliofemoral venous thrombosis and most of the compression was by lymph nodal metastasis secondary to urothelial cancers (46).

The bladder was the most common primary site (29) followed by ureter ($n = 8$), renal pelvis ($n = 5$), prostate ($n = 2$), or penis ($n = 2$) but none of these were due to direct malignant compression by ureteric mass. Other causes of iliofemoral venous compression included the most commonly described syndrome called the Iliac Vein Compression Syndrome or the May-Thurner Syndrome involving compression of Left Iliac Vein by Right Iliac Artery [11].

Many other rare causes of iliofemoral compression and thrombosis have been reported in the literature including uterine fibroids [12], psoas abscess [13], retroperitoneal fibrosis [14], penile prosthesis reservoir [15] and synovial cyst of hip [16]. Other urological entities have also been reported to cause iliofemoral venous thrombosis.

Through our case report, we would like to emphasize the need for a thorough evaluation of patients presenting with unilateral lower limb swelling due to DVT without any predisposing factors. A simple ultrasound of the abdomen or CECT might shed light on an underlying malignancy. A proper diagnosis will guide therapy and will lead to early management of malignancy resulting in a better prognosis.

CONCLUSION

All cases with sudden onset unilateral lower limb swelling need a thorough evaluation for a possibility of life-threatening underlying pathology.

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