BANTAO Journal 2008; 6(2): 34-37

BJ
BANTAO Journal

Case report

Primary renal vein thrombosis: a case report

Gursu Meltem[1], Ozturk Savas [1], Yıldıray Savas[2], Uzun Sami[1], Aydın Zeki[1] and Kazancioglu Rumeyza[1]

[1]Department of Nephrology and [2]Department of Radiology, Haseki Training and Research Hospital, Istanbul, Turkey

Abstract

Renal vein thrombosis is an uncommon condition with variable etiology and clinical presentation. The most common reasons are nephrotic syndrome, malignancies especially those of the kidneys. Hypercoagulability states, estrogen-containing pills, pregnancy, trauma and surgery of kidneys and its vessels, systemic inflammatory states and renal transplantation are other possible reasons.

Here we present a 39-year-old female patient who was hospitalized with the complaints of fever, right flank pain, nausea, vomiting and blood in her urine for about 15 days. Hematuria, elevated CRP and erythrocyte sedimentation rate, anemia due to iron and vitamin B12 deficiency, mildly elevated transaminase levels and mild proteinuria were remarkable in her laboratory results. Ultrasonography showed increased dimensions and echogenity of the right kidney. Doppler ultrasonography showed thrombus in the right renal vein; and CT angiography confirmed the diagnosis. She was found to be negative for hypercoagulability with normal prothrombin time, activated partial thromboplastin time, protein C and S, homocystein levels and negative antiphospholipid antibodies, factor V Leiden and prothrombin gene mutations. Antinuclear and anti-double stranded antibodies were negative. There was no clinical and radiological sign of malignancy and tumor markers were negative. We started anticoagulant therapy immediately; but during follow-up the thrombus sustained with a nonfunctioning and shrunken right kidney demonstrated with CT and scintigraphy.

This case is rare in that she has no nephrotic syndrome, no malignancy, hypercoagulability and other etiologies except the use of oral contraceptive agent. It also reminds us the importance of early diagnosis and treatment to preserve renal functions.

Keywords: primary; renal; thrombosis; vein

Correspondence and offprint request to: Meltem Gursu, Haseki Egitim ve Arastirma Hastanesi, Nefroloji Klinigi, Haseki Aksaray 34390 Istanbul, Turkey; Tel: +90 2125294400; Fax: +90 212 5294453; E-mail: meltem1401@yahoo.com

Introduction

Renal vein thrombosis (RVT) is an uncommon condition with variable presentations. The causes are also variable; the most common ones being nephrotic syndrome and, malignancies, mainly renal cell carcinoma [1]. Other less common etiologies include primary hypercoagulability, trauma, estrogen containing preparations, pregnancy, infections, sepsis, systemic inflammatory diseases like inflammatory bowel disease [2], interventions to or sugery of renal veins, extrinsic compression and renal transplantation [3]. Idiopathic cases have been reported rarely [4].

The most common symptoms are flank pain and hematuria, which may be mistaken for renal colic or pyelonephritis [5]. Nausea and vomiting, fever, a palpable flank mass, edema often accompany other symptoms. Leukocytosis is a frequent finding [6]. Moreover; unexpected worsening of proteinuria and renal functions may be observed in nephrotic syndrome patients. When chronic, it may be totally asymptomatic as collateral veins develop and allow preservation of renal function, but still with risk of pulmonary embolism. Waldemer *et al* [1]. studied the clinical characteristics and long-term follow-up of 218 patients with RVT and reported the occurrence rate of signs and symptoms as follows: Flank pain (73%), gross hematuria (36%), nausea/vomiting (13%), anorexia (21%), fever (12%), dyspnea (2%), edema (2%), anemia (38%), palpable flank mass (9%), splenomegaly (6%), ascites (6%), peritoneal signs (4%), encephalopathy (2%), and asymptomatic (15%).

Diagnosis is based on clinical, laboratory and radiologic findings. Computerized tomography (CT) angiography is the investigation of choice. Magnetic Resonance Imaging and venography in highly selected patients are other alternatives [7].

The treatment options include anticoagulation, thrombolytic therapy and surgery [8]. The prognosis is variable; the main prognostic factors are the acuteness of the disease, involvement of one or two veins, underlying etiology (membranous glomerulopathy having a better prognosis) and presence of initial renal insufficiency (a bad prognostic factor) [9].

Here we present a case of renal vein thrombosis with a delayed diagnosis and no response to anticoagulation.

Case report

A 39-year-old woman presented with right flank pain, nausea, vomiting and blood in urine that started 15 days ago. Three months earlier she was admitted to another hospital with right pleural pain, cough, fever and hemoptysis; where lobar pneumonia and parapneumonic effusion were diagnosed. Her symptoms completely resolved without any sequel with antibiottherapy. Fifteen days ago a sudden onset right flank pain, nausea and vomiting began; followed by hematuria. When she presented to the emergency unit, nephrolithiasis was suspected; but urinary ultrasonography (US) and abdominal CT did not verify this diagnosis; ultrasonography showed increased echogenity of the kidneys while CT was reported to be totally normal. Her past medical history consisted of a uterine myomectomy performed six months ago. She gave birth to a healthy girl 6 years ago. She had no abortus or stillbirths. She smoked 10 pack-years.

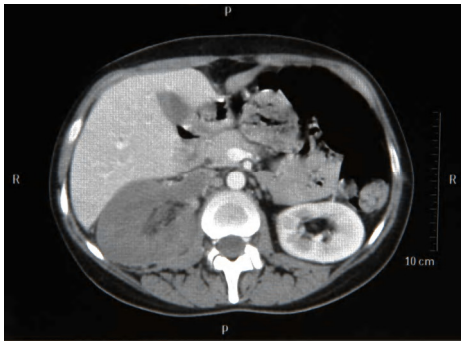
On admission her blood pressure was 100/70mmHg, pulse was 88/minute and rhythmic, the temperature was 37.2°C. Skin was pale without jaundice, cyanosis and edema. Examination of the respiratory and cardiovascular systems revealed no pathological findings. Abdominal examination revealed right upper quadrant tenderness without distension and rebound. There was tenderness of the right costovertebral junction.

Table 1. Laboratory parameters of the patient

Parameter	Value	SI unit
White blood cells	14.5	$x10^3/mm^3$
Hemoglobin	8.4	gr/dl
Mean corpuscular volume	61.5	femtoliter
Erythrocyte sedimentation rate	124	mm/hour
C- reactive protein	149	mg/dl
Urea	23	mg/dl
Creatinine	1.03	mg/dl
Sodium	134	mmol/L
Potasium	4.83	mmol/L
AST	74	U/L
ALT	54	U/L
GGT	316	U/L
ALP	280	U/L
LDH	864	mg/dl
INR	0.94	
aPTT	22	Seconds
Fibrinogen	1214	mg/dl

Laboratory investigations were as follows: Urine sediment was active with many erythrocytes and leukocytes at each high power field. Blood tests included white blood cells $14.5x10^3$/microliters, hemoglobin 8.4 g/dl with mean corpuscular volume of 61.5 fl, platelets $661x10^3$/microliter, erythrocyte sedimentation rate 124 mm/hour, C-reactive protein 149 mg/L, urea 23 mg/dl, creatinine 1,03 mg/dl, sodium 134 mmol/L, potassium 4,83 mmol/L, AST 74 U/L, ALT 54 U/L, GGT 316 U/L, alkaline phosphatase 280 U/L and lactate dehydrogenase 864 mg/dl. Coagulation parameters included international normalized ratio of 0.94, partial thromboplastin time of 22.7 seconds and fibrinogen level of 1214 mg/dl. Iron (16 µg/dl) and vitamin B12 levels (142 pg/ml) were below the normal limits. Creatinin clearance rate was 52 ml/min and there was a microalbuminuria of 283 mg/day. Her hypercoagulopathy panel was negative for antiphospholipid antibodies, hyperhomocysteinemia, factor V Leiden mutation and prothrombin gene mutation. Protein C and S levels were within normal limits. Antinuclear antibodies and Anti double stranded DNA antibodies were negative.

a)



b)

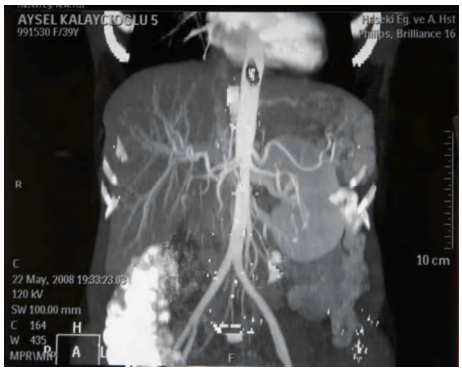


Fig. 1. CT and CT angiography of the kidneys

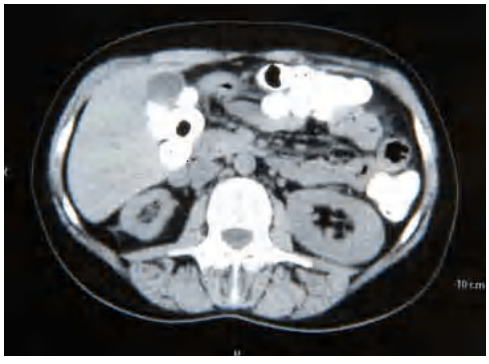


Fig. 2. CT scan two months after the onset of treatment

Urinary system US and CT was repeated that showed an enlarged right kidney (124x61mm) with increased echogenity. Doppler ultrasonograpy of renal vasculature was consistent with right renal vein thrombosis. CT angiography demonstrated that there was a thrombus in the right renal vein with extension into the inferior vena cava and the nephrogramme phase at the right kidney could not be demonstrated (Figure 1). With DTPA/DMSA

renal scintigraphy, a nonfunctioning right kidney was reported.

Low molecular weight heparin was started followed by warfarin. During follow-up, she had no symptoms, renal functions were well preserved, no hematuria recurred, but control renal Doppler ultrasonography performed two months after the onset of anticoagulant therapy revealed that the thrombus occluding the right renal vein remained with no sign of flow. Control CT showed that the right kidney was shrunken (Figure 2), with no function as assessed by scintigraphyc study. She still uses warfarin for the risk of embolism.

Discussion

Diagnosis of renal vein thrombosis requires a high index of suspicion. Since the clinical presentation is so variable, the diagnosis is often delayed. The pain it causes may easily be confused with nephrolithiasis, pleurisy; and inquiry to these diagnoses may lead to waste of time and loss of renal parenchyma.

Our case presented with right flank pain, fever and hematuria and there was a history of pneumonia. Nephrolithiasis, pyelonephritis, pulmonary embolism and renal infarct were within the list of differential diagnosis. Macroscopic hematuria and right loin pain supported the diagnosis of nephrolithiasis; but urinary US and CT revealed no stones. Pyuria, fever and high CRP levels suggested pyelonephritis; but urine culture remained sterile. The CT scan demonstrated that the right kidney was larger than normal. A unilateral large kidney may be due to renal cysts, tumors, hydronephrosis, abscess formation or edema due to vascular obstruction. Our case did not have signs consistent with tumor, hydronephrosis or abscess in the CT scans. So the possible diagnosis was renal vascular disease which was proved with Doppler ultrasonography and CT angiography.

Experimentally acute RVT is associated with immediate enlargement of the kidney with marked increase in renal vein pressure, leading to a marked decrease in renal arterial flow. In dogs the kidney enlarges over a period of one week, then fibrosis gradually ensues [10] which suggests that the practitioner must carry the suspicion to prevent delay in diagnosis and loss of renal tissue. In our patient there was a delay of at least 15 days.

The left renal vein is in communication with ureteric, gonadal, adrenal and phrenic veins; whereas the right renal vein has not this much communication. This network of venous complexes provides some protection against renal infarction [10]. These may be major reasons of our patient not responding to anticoagulant therapy; i.e. late diagnosis and thrombosis of the right renal vein.

Renal vein thrombosis is rarely primary. Our patient had no sign of malignancy in either clinical and laboratory findings and radiologic evaluations. Tumor markers were negative. She had no hypercoagulability state. She only had non-nephrotic range proteinuria, which was thought to be secondary to renal vein thrombosis. Cases of renal vein thrombosis secondary to that of ovarian vein and than inferior vena cava have been reported [11,12]. But our patient did not have any sign of thrombosis in pelvic veins examined with CT angiography. The treatment of RVT remains controversial and depends on the acuteness and renal consequences. Surgery is reserved for complications like capsular rupture due to edema, or long-term consequences like hypertension and infection of the nonfunctioning kidney. In adults with acute RVT secondary to hypercoagulability, thrombolytic therapy may be useful in regaining the flow in the renal vein. But the long-term effects are not clear. If left renal vein is occluded a conservative approach may be preferred due to extensive collateral circulation at that side. Systemic anticoagulation is effective in the acute period in preventing extension to the inferior vena cava and pulmonary embolism. The duration of anticoagulation is not certain. If it is due to hypercoagulable states treatment must go on indefinitely. Eventual recanalization may lead to delayed improvements in renal functions. For our patient it seems that anticoagulation is not effective and the right kidney is nonfunctional. But we continue the treatment to prevent thrombus extension and pulmonary embolism.

Conclusions

RVT is a disease, which requires a high index of suspicion to diagnose. The diagnosis of RVT is not a single one but necessitates investigation of a probable underlying disease, although it may be rarely primary. It is always important to pay attention to flank pain and hematuria not to loose time and renal parenchyma.

Conflict of interest statement. None declared.

References

1. Waldemar E, Wysokonski WF, Gosk-BierskAI *et al*. Clinical characteristics and long-term follow-up of patients with renal vein thrombosis. *Am J Kidney Dis.* 2008; 51(2): 224-232.
2. Tabibian JH, Lada SJ, Tabibian N *et al*. Combined inferior vena cava & renal vein thromboses: case and synopsis of thromboembolism in inflammatory bowel disease. *Medscape J Med.* 2008; 9: 10(1): 6.
3. Harris R, Ismail N *et al*. Extrarenal complications of nephrotic syndrome. *Am J Kidney Dis* 1994; 23: 477-497.
4. Hmidi M, Hani MA, Hafsia G, Nouira Y, Sallami S, Horchani A**:** Idiopathic renal vein thrombosis. About two cases. *Tunis Med.* 2006, 84(6): 374-376.
5. Kumar S, Singh SK, Mavuduru RS *et al.* Acute pyelonephritis with renal vein and inferior vena cava thrombosis in a case of hyperhomocysteinemia. *Int Urol Nephrol.* 2008 Apr 1.
6. Zucchelli P *et al*. Renal vein thrombosis. *Nephrol Dial Transplant* 1992; 7(suppl 1): 105-108.
7. Asghar M, Ahmed K, Shah SS, Siiddque MK, Dasgupta P, Khan MS. Renal vein thrombosis. *Eur J Vasc Endovasc Surg* 2007; 42: 217-223.
8. Witz M, Korzets Z *et al.* Renal vein occlusion: diagnosis and treatment. *Isr Med Assoc J.* 2007; 9(5): 402-405.
9. Laville M, Aguilera D, Maillet PJ, Labeeuw M, Madonna O, Zech P. The prognosis of renal vein thrombosis: a re-evaluation of 27 cases. *Nephrol Dial Transplant* 1988; 3(3): 247-256.
10. John Feehally, Jürgen Floege, Richard J Johnson *et al. Comprehensive Clinical Nephrology 3rd edition*. Mosby Elsevier, Philadelphia, 2007:741-742.

11. Hassen-Khodja R, Gillet JY, Batt M *et al*. Thrombophlebitis of the ovarian vein with free-floating thrombus in the inferior vena cava. *Ann Vasc Surg* 1993; 7(6): 582-586.

12. Reichelt HG *et al*. Neoplastic thrombosis from a uterine cancer via the ovarian vein. Venous collateralization via the vasa vasorum in occlusions of the renal vein and the inferior vena cava. *Radiologe* 1986; 26(8): 385-7.