Retroperitoneal Fibrosis: Report of 3 Cases and Review of Literature

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Retroperitoneal fibrosis (RPF) is a rare condition with poorly understood etiology. Patients present with non-specific symptoms and diagnosis is usually made during evaluation for urinary tract obstruction or venous/arterial insufficiency. We evaluated and managed three cases in last four years and followed them up. This is the summary of our cases and review of literature highlighting the disease.

Keywords: ormond's disease, periureteritis, retroperitonitis, retroperitoneal fibrosis.

Retroperitoneal fibrosis (RPF), also known as Ormond's disease or periureteritis, is a rare condition characterized by the presence of inflammatory and fibrous retroperitoneal tissue that often encases the ureters or abdominal organs.[1] We had the opportunity to manage three cases over a period of four years which have been summarized below.

Case 1

A 46-year-old lady presented with diffuse right loin pain since one year which was associated with anorexia and weight loss. She was a smoker and had menopause 2 years back with history of taking multiple medications including proton pump inhibitor for acid peptic disease, non-steroidal anti-inflammatory drugs (NSAIDs) for left trigger thumb and topical ointment containing clobetasol and salicylic acid for active psoriasis vulgaris.

On clinical examination, bilateral renal angles were mildly tender. There were scaly skin plaques of psoriasis on bilateral lower limbs. Routine blood tests revealed raised erythrocyte sedimentation rate (ESR) of 98 mm in the first hour. Other tests including compete blood count, blood borne virus screening, liver function test,

renal function test, urinalysis and echocardiography were normal.

Her abdominal ultrasound (USG) and subsequent contrast enhanced computerized tomography (CECT) revealed long segment enhancing retroperitoneal soft tissue mass/mantle along the superior mesenteric artery (SMA) which was encasing it and extending towards the lower abdominal aorta upto the common iliac bifurcation. There was compression of right ureter at third lumbar vertebrae resulting in mild-moderate right sided hydroureteronephrosis **(Figure. 1)**.

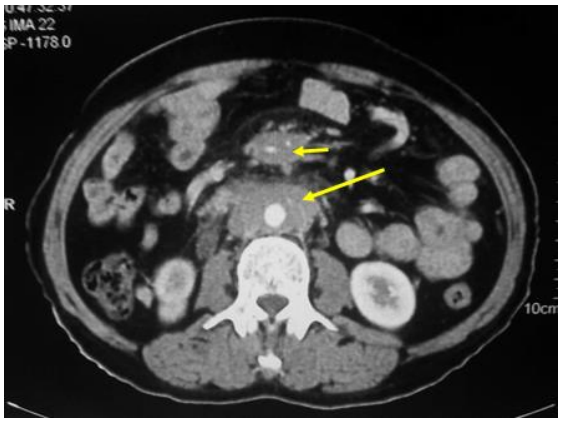

Figure 1. Contrast enhanced computed tomography abdomen Showing retroperitoneal soft tissue mass encasing the lower abdomen aorta (long arrow) and superior mesenteric vessels (short arrow)

CT guided biopsy was tried to rule out malignancy but was not possible and hence laparotomy and incisional biopsy was done. Intraoperatively, the retroperitoneal tissue was plastered with aorta and superior mesenteric artery (SMA), from which biopsy was taken. Histological examination of the specimen revealed chronic inflammation with marked fibrosis and final diagnosis of retroperitoneal fibrosis was made.

Right ureter was stented using long duration silicon double J stent and patient improved symptomatically. She was discharged on oral medications which included Tamoxifen and Prednisolone for 3 months. On 3 months follow-up, the dose of prednisolone was tapered and gradually stopped. Double J stent was removed after total one year and she currently has no symptoms. Her right hydroureteronephrosis has resolved and ESR dropped to normal level.

Case 2

A 53-year-old smoker and diabetic male presented with dull aching low back pain since 4 months associated with scrotal swelling. He gave history of weight loss about 8 kilograms in the past 6 months. On clinical examination, he was found to have grade II bilateral varicocele. His right lower limb was colder and femoral pulse feeble in comparison to left.

Routine laboratory tests revealed ESR of 77 mm in 1st hour, CRP 101 mg/l, random blood sugar (RBS) 170 mg/dl and serum creatinine 1.24 mg/dl. CECT abdomen revealed thick conglomerated slightly enhancing retroperitoneal soft tissue lesion in pre and para-aortic areas encasing the abdominal aorta from the level of main renal artery and caudally seen extending and encasing the bilateral common iliac arteries, left common iliac vein, inferior mesenteric artery and the mid-distal ureter. There was also features of complete occlusion of left common iliac artery and moderate bilateral hydronephrosis with

poorly excreting left kidney.

USG guided fine needle aspiration (FNA) of the mass was done which showed features of benign fibrous lesion negative for malignancy. Diagnosis of retroperitoneal fibrosis causing bilateral obstructive uropathy was made and bilateral retrograde pyelogram and double J stenting was done. Patient was discharged on oral prednisolone (40mg daily) but due to uncontrolled diabetes, the dose was readjusted to 5mg daily. DJ stent was later removed after 3 months.

Case 3

A 66-year-old lady presented with left loin pain since 7 months and associated with burning sensation during micturition and fever with chills and rigor for last 7 days. She was non-smoker, taking metformin for diabetes and amlodipine for hypertension. On clinical examination, left renal angle was tender. Routine lab tests revealed leukocytosis upto 17300/cumm, ESR upto 80 mm in 1st hour and serum creatinine upto 2.21 mg/dl. Urinalysis was suggestive of urinary tract infection.

USG abdomen revealed moderate left hydronephrosis. Non-contrast computerized tomography (NCCT) could only be done due to increased creatinine which revealed abrupt narrowing at the distal ureter associated with perinephric fat stranding and thickening of renal fascia **(Figure 2)**. With the suspicion of ureteral mass causing obstructive uropathy, retrograde pyelogram followed by ureterorenoscopic biopsy of the distal ureteric thickening was done under antibiotic coverage and double J stent was placed to relieve the obstruction. The histology of the specimen was non-specific inflammation without any features of malignancy and hence no further intervention was taken. She gradually improved with creatinine drop upto 1.8 mg/dl. Patient was discharged with DJ stent in situ and anti-inflammatory medication (NSAIDs).

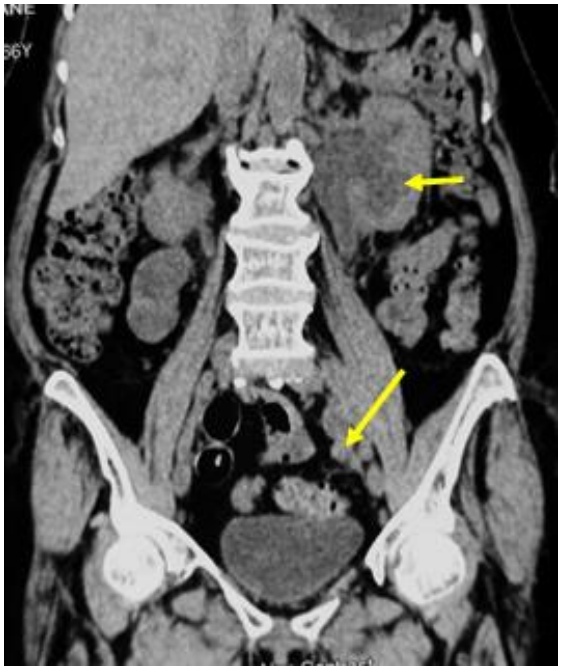

Figure 2. Plain CT KUB showing left lower ureteric mass about 4.5cm (long arrow) Causing proximal hydroureteronephrosis (short arrow)

On three months follow-up, DJ stent was removed during which period she was asymptomatic. However, after 2 months of the stent removal, patient was readmitted for left obstructive uropathy with creatinine reaching upto 3.2 mg/dl and moderate hydronephrosis. Exploration, excision of the mass encasing the ureter followed by re-implantation of the ureter was planned.

On exploration, there was dense fibrosis

and thickening at the left distal periureteric region causing kinking of the left distal ureter with proximal hydroureter. Soft tissue mass was palpable at the region of mesorectum with large lymph nodes in the left obturator and external iliac region. Left ureterolysis, distal ureteric segment excisional biopsy, neoureterovesicostomy was done and periureteric tissue was sent for biopsy which confirmed retroperitoneal fibrosis. Patient was discharged on tamoxifen and prednisolone on tapering dose with removal of DJ stent at 3 months. On follow-up at one year, she is in remission with creatinine of 2 mg/dl and free of symptoms.

Discussion

RPF is a rare condition with incidence of 1.3 per 100,000 person-years and prevalence of 1.4 per 100,000 inhabitants, commonly occurring in individuals 40 to 60 years of age with male predominance.[2-4] There are two leading theories regarding its pathophysiology.[5] One theory suggests that there is exaggerated local inflammatory reaction to aortic atherosclerosis, incited by oxidized low density lipoprotein (LDL). The other theory suggests that it is a manifestation of systemic autoimmune disease. There are other processes described as well such as large-vessel vasculitis, formation of antibodies to fibroblasts and the infiltration of IgG4-producing plasma cells (Ig-G4 related disease).[6]

Presentation is non-specific with dull aching low back pain being the most common symptom.[7] The pain is often associated with malaise, anorexia, weight loss, fever, nausea, vomiting and testicular pain.[8] Some patients may present with unilateral or bilateral flank pain radiating to the inguinal region which may be similar to renal colic. Often RPF is considered only once there is significant organ involvement, most commonly ureters & kidneys and hence most patients have ureteral obstruction and renal impairment by the time they come to diagnosis. Among our three cases, all had loin pain and last two cases presented with obstructive uropathy. Other manifestations described in literature may be new onset hypertension, hydrocele, varicocele or lower extremity involvement like edema, thrombophlebitis or deep vein thrombosis resulting from obstruction of the inferior venacava and/or iliac veins.[9] The differential diagnosis include lymphoma or sarcomas, retroperitoneal fibromatosis, inflammatory pseudotumor or infections such as tuberculosis.[9]

Laboratory findings are non-specific with raised ESR & C reactive protein (CRP). There may be anemia due to renal insufficiency. Ultrasonography is often the first procedure done which may reveal a poorly marginated, peri-aortic mass that is typically echo-free or hypoechoic and may be associated with hydronephrosis.[10] CT Scan is the examination of choice which is done to visualize the extent of fibrosis, to assess the presence of lymphadenopathy and tumor.[11] Other diagnostic tests are magnetic resonance imaging (MRI), intravenous pyelogram (IVP), retrograde or percutaneous pyelography and positron enhanced tomography (PET) using 18-

flurodeoxyglucose.

However, we diagnosed our cases on the basis of histology given that radiological features were inconclusive. There is role for biopsy when the location and nature of mass is atypical, when clinical, laboratory or radiologic findings suggest the presence of an underlying malignancy or infection and if the patient does not respond to initial therapy.[12]

Retroperitoneal fibrosis is an uncommon but treatable cause of obstructive uropathy. The goals of therapy are to relieve the obstruction caused by fibrosis, stop the progression of the fibrotic process and to prevent recurrence. Further goals are preservation of renal function and freedom from pain, steroids and stents.[13] Treatment is according to underlying etiology. If idiopathic, immunosuppressive therapy is initiated with glucocorticoids as the mainstay of therapy.[12] A variety of other medical therapies have been used with or without glucocorticoids including tamoxifen, colchicine and other immunosuppressive agents like mycophenolate mofetil, methotrexate, azathioprine, cyclophosphamide and cyclosporine.

The first and second case were managed by DJ stenting and steroid therapy. However, in the third case, we might have missed the diagnosis of RPF at the very beginning. On the basis of clinical and radiological clues, distal ureteric mass was thought to be the cause of obstructive uropathy. Because of the atypical location, background history of hysterectomy and histology ruled out malignancy, we focused only on relieving the obstructive uropathy by DJ stenting.

Surgical management is decompression of the urinary tract when patients present with renal compromise due to obstruction of the urinary tract.[14] Based upon the condition of the patient and severity of the condition, different techniques can be used. Double J stenting or percutaneous nephrostomy is done for milder form of the disease whereas ureterolysis is done for more severe form and patients suitable for anesthesia. Laparoscopic or open ureterolysis can be done followed by wrapping the ureters with omental fat, translocation of the ureters to an intraperitoneal position or transposition of the ureters laterally, with interposition of retroperitoneal fat between the ureters and the fibrous tissue.

Once treatment is started, response is monitored with USG or CT scan usually every 2-3 months or more frequently depending on severity. There should be resolution of size of the mass, decrease in pain and obstruction with decrease in ESR and CRP.

Pain and constitutional symptoms usually improve within few days of initiating treatment. Patients who respond to steroids, mortality is less than 10% (excluding cases associated with malignancy). Full resolution depends on the duration of entrapment. If patient has developed renal insufficiency with associated hypertension or anemia, it may persist despite relief of obstruction as there might have been permanent renal damage.

Conclusion

RPF is a rare but potentially treatable

disease. Many patients have ureteric obstruction and renal insufficiency during presentation. Goal of therapy is to relieve obstruction, stop progression & prevent recurrence. CT scan is the choice of investigation and prednisolone is the mainstay of therapy. Life-long monitoring is recommended.

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