Case Report

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Atypical Presentation of Sarcoidosis with Retroperitoneal Fibrosis, Non-Caseating Granulomatous Perinephric Lesion and Obstructive Uropathy

Ramchandani Santosh[1]*, Syed Ali Aown[1], Joseph Guzzo[1], Lata Ramchandani, Benjamin Wilcox[1] and Glen Markowitz[2]

[1]*Lehigh Valley Hospital, Allentown, USA*

[2]*Columbia University Medical Center, USA*

***Corresponding author:** Ramachandani Santosh, Lehigh Valley Hospital, Allentown, PA, USA

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ABSTRACT

A 66 year old female was admitted to the hospital with acute renal failure, hyperkalemia, and anemia. Etiology of her acute renal failure was obscure, and all serologic studies were negative. A kidney biopsy was performed shows interstitial infiltration noncaseating granulomas. CT scan abdomen revealed bilateral kidney enlargement and retroperitoneal fibrosis (RPF). Perinephric tissue also demonstrated dense fibrosis, inflammation and noncaseating granulomas. Treatment was initiated with 80mg of prednisone and bilateral ureteral stents were inserted because of fullness of collecting system seen on CT scan. Over 2 weeks, renal function improved towards normal. Kidney and retroperitoneal findings highly suggested renal sarcoidosis.

Keywords: Sarcoidosis; Acute Kidney Injury; Retroperitoneal Fibrosis and Obstructive Uropathy

Introduction

Sarcoidosis is a multisystem disease of unknown etiology characterized by noncaseating granulomatous lesions involving primarily lymph nodes, lungs, skin and eyes [1]. Common presentation of sarcoidosis include abnormal calcium metabolism, nephrocalcinosis and lymphadenopathy [2]. We have a very rare case of renal sarcoidosis which presented with acute renal failure, normocalcemia, bradycardia, and retroperitoneal fibrosis causing obstructive uropathy.

Case

A 66-year-old Caucasian smoker female with past medical history significant for hypertension, hyperlipidemia, tobacco use disorder, and COPD was admitted to the hospital with acute renal failure, hyperkalemia and anemia. On further inquiry, the patient reported she had been feeling tired, shortness of breath for 2 weeks and had prescribed Bactrim for bronchitis infection by her PCP. Patient self-discontinued antibiotic due to abdominal pain prior to presentation. On

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arrival, her blood pressure was 120/79 mmHg, pulse of 52 bpm, respiratory rate 19, temp 97.1, SPO2 94% on 2L oxygen via nasal canula. Physical exam revealed ill-appearing female with 1+ lower extremity pitting edema. Her cardiac rhythm was regular and slow, there was no friction rub or murmur. Her lungs had bilateral wheeze but no rales. Her Initial hemoglobin was 6.7 g/dL, WBC 7.6, serum creatinine 4.37 mg/dl (baseline 0.8–1.1 mg/dl 6 months ago), BUN of 100 mg/dl, potassium of 6.3 meq/l. Urinalysis negative for protein and blood. Microscopy exam showed no RBCs, WBCs or cast. Patient was managed with IV diuretics, glucose and insulin for hyperkalemia. A CT scan of the abdomen (Image 1) demonstrated enlarged kidneys fullness of the collecting system and perinephric retroperitoneal fibrosis. Her urine culture was negative. Kidney biopsy was performed on the 4th hospital day. The perinephric tissue showed dense fibrosis, inflammation, and noncaseating granulomas (Images 2 & 3). The Renal cortex contain 13 glomeruli 5 globally sclerosed and remaining demonstrated ischemic wrinkling.

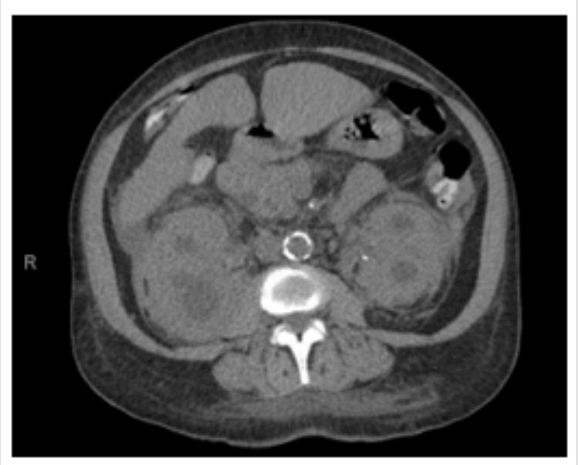

Image 1: CT scan A/P Enlarged kidney with retroperitoneal fibrosis.

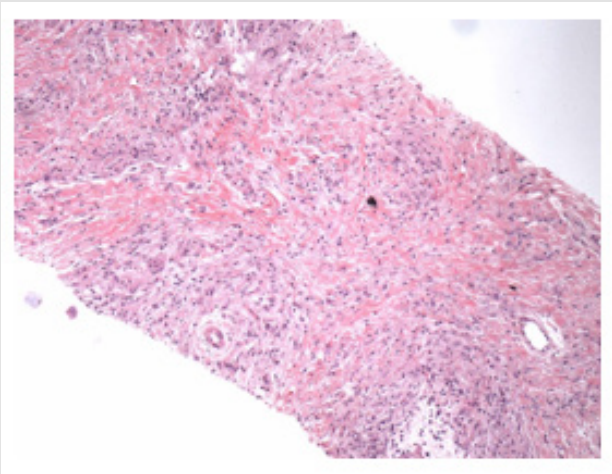

Image 2: Show the granulomatous inflammation outside the kidney (in the capsule and contiguous connective tissue) H&E stain.

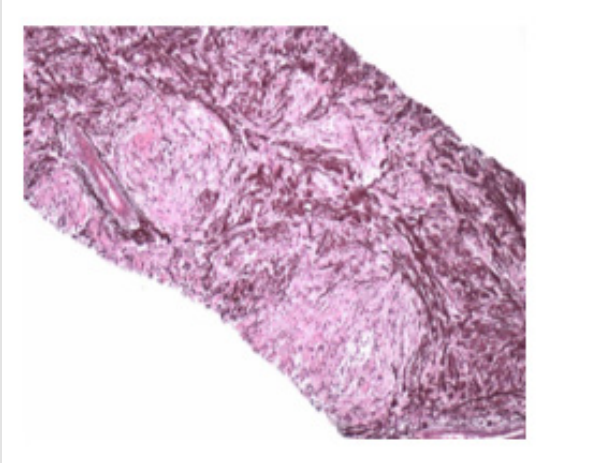

Image 3: Well-delineated non necrotizing granulomas with adjacent tubulointerstitial fibrosis.

IF was negative. The interstitium had dense infiltration of lymphocytes, and occasional plasma cells and non-caseating granulomas (Image 4). Acid-fast bacilli and fungal stain were negative. These findings were interpreted as renal sarcoidosis. 125 dihydroxy vitamin D and, angiotensin-converting enzyme were negative. High-resolution CT scan chest and abdomen shows interstitial lung disease and retroperitoneal fibrosis with no evidence of lymphadenopathy. Because of bradycardia cardiac MRI was planned to evaluate of conducting to rule out cardiac sarcoidosis. The patient was treated with prednisone 80 mg daily and bilateral ureteral stents were placed because of possible obstruction due to retroperitoneal fibrosis. After 5 days at the time of discharge the serum creatinine had decreased to 1.1mg/dl. Her wheeze, and bradycardia also resolved on the steroids.

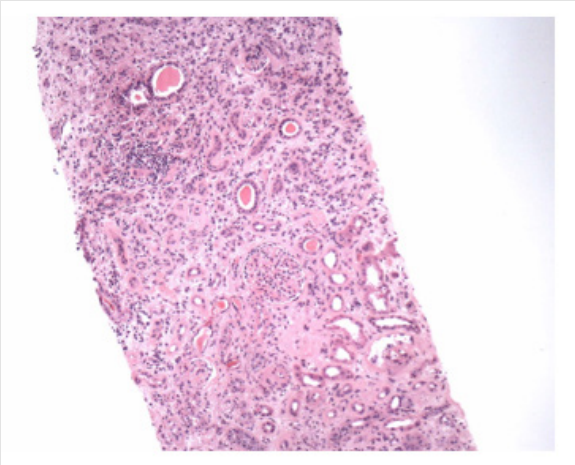

Image 4: Shows the findings with granuloma in the renal cortex, below the capsule.

Discussion

This patient had pulmonary manifested by dyspnea, wheeze, hypoxia and interstitial lung diseases on CT but no perihilar adenopathy. Renal involvement is evident in 11% sarcoid patients [3]. Hypercalcemia and nephrocalcinosis are common manifestations of sarcoidosis which leads to renal insufficiency; however, glomerulopathy, interstitial nephritis, and obstructive uropathy may also occur [3]. Retroperitoneal fibrosis is a late manifestation of sarcoidosis with pulmonary involvement noticed in some studies [4]. There was no evidence of kidney stone, hypercalcemia, proteinuria, hematuria, and nephrocalcinosis in outpatient. At autopsy 40% of patient with sarcoidosis may have renal granuloma [5]. Isolated retroperitoneal fibrosis along with ureteral obstruction is rarely seen in sarcoidosis [4]. There are

no clear guidelines on how retroperitoneal fibrosis (RPF) patients should be managed. The main goal is to preserve renal function in RPF. Currently, the suggested treatment is the combination of steroids with ureteral stents in urinary tract obstruction or renal insufficiency [6]. Our patient had a good renal recovery with steroids and ureteral stents. Tamoxifen has been recommended as an alternative to steroids [7,8]. There is limited data on the use of other immunosuppressive drugs, methotrexate [9]. Cyclophosphamide [10]. Azathioprine [11]. and mycophenolate mofetil. Surgical ureterolysis are necessary by laparoscopy in refractory cases [12,13]. Most patients with RPF have a good prognosis but relapses may occur in 15 to 50% of cases [14-17]. and some may progress to ESRD. In RPF, Mortality rate is reported 0-9% [14,15,18-20]. and ESRD of 0-8% [14,16,17]. The risk factors of relapse include a shorter duration treatment with steroids [14]. higher baseline ESR [14]. smoking [16]. and AKI [16]. In this patient the treatment was planning to continue steroid for 6 months to decrease the chance of relapse.

Conclusion

We presented a case of renal sarcoidosis and sarcoid retroperitonea fibrosis leading to renal failure. Our patient fully recovered with steroids and ureteral stents. Larger studies are needed to further explore treatment options and prognosis.

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