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Isolated Renal Sarcoidosis: A Rare form of the Disease

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Abstract: ***Introduction***: Sarcoidosis is a multi-system disease characterized by frequent involvement of the lungs. Other organ manifestations without accompanying lung findings are extremely rare. Kidneys may be rarely affected. We present a case, who presented with hypercalcemia, had no lung involvement, and was diagnosed with isolated renal sarcoidosis by renal biopsy. ***Case***: A 76-year-old female patient, who applied with complaints of fatigue and dry mouth, was examined after the examinations revealed increased creatinine and hypercalcemia. Serum creatinine was 4.46 mg/dl, her calcium, parathormone, 1,25-dihydroxy-vitamin D, and serum ACE levels were elevated. She had been examined for hypercalcemia for 5 years, and a diagnosis could not be made. A renal biopsy was performed due to creatinine progression, hypercalcemia and high 1,25-dihydroxy-vitamin D levels. The biopsy was consistent with sarcoidosis-associated interstitial nephritis. Corticosteroids were administered and she responded well. ***Conclusion***: It is crucial to keep extrapulmonary sarcoidosis in mind in patients with hypercalcemia and elevated serum ACE, even in the absence of lung involvement. Although rare, extrapulmonary forms of sarcoidosis can also be seen isolated.

Keywords: Sarcoidosis, Hypercalcemia, Sarcoidosis-Associated İnterstitial Nephritis
Isolated Renal Sarcoidosis: A rare form of the disease.

Case Report

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INTRODUCTION

Sarcoidosis is a multi-system disease of unknown cause, characterized by the formation of non-caseating granulomatous lesions in various organs, frequently involving the lower respiratory tract [1]. Lungs are most commonly involved, and other organ involvement without accompanying lung findings is extremely rare. Kidneys may be rarely affected. Although autopsy findings reveal interstitial granulomatous lesions in up to 13% of the renal parenchyma in sarcoidosis patients, renal involvement is a very infrequent manifestation of the disease. Isolated renal impairment as initial presentation of sarcoidosis is very uncommon [2, 3].

Renal manifestations of sarcoidosis include abnormal calcium metabolism, nephrocalcinosis, and nephrolithiasis [4]. Some studies have described clinical, laboratory, or histological features in renal sarcoidosis, and demonstrated that granulomatous tubulointerstitial nephritis is associated with acute renal failure and may lead to hemodialysis [5]. In previous clinical series on sarcoidosis, the rate of renal failure ranged from 0.7% to 4.3% [6-8].

The cornerstone of treatment is corticosteroid therapy. Its effectiveness has been proven in the advanced stage sarcoidosis tubulointerstitial nephritis. In some cases, immunosuppressive treatment may be required [9]. There are few studies, which address specific issues regarding renal involvement in sarcoidosis [10]. Little is known about the prognostic factors of this condition and its relationship with other systemic manifestations and calcium metabolism. (10) We present a case, who presented with hypercalcemia, had no lung involvement, and was diagnosed with isolated renal sarcoidosis by renal biopsy.

CASE

A 76-year-old female patient, who applied with complaints of fatigue and dry mouth, was found to have increased creatinine, and hypercalcemia, and was referred to the nephrology outpatient clinic. Her medical history revealed hypertension and diabetes mellitus, glaucoma and chronic renal failure that did not require hemodialysis for 6 years. On physical examination, breath sounds were normal and pretibial edema was minimally positive. Other system findings were normal. In the laboratory examination performed on admission, serum creatinine was 4.46 mg/dl, basal creatinine value

was 2.1 mg/dl, serum calcium was 11.60 mg/dl (8.8-10.6), parathormone was 5 ng/L(15-65), 1,25 dihydroxy vitamin D >150.00 ng/ml and serum angiotensin-converting enzyme (ACE) was 175 U/L (13.3-63.9). Twenty-four-hour urine analysis revealed 732 mg proteinuria and 243 mg albuminuria.

It was noted that the patient had been examined for hypercalcemia for 5 years, and no diagnosis could be made with parathyroid scintigraphy, fluorodeoxyglucose-positron emission tomography scan, cranial, thoracic and abdominal computed tomography (CT), cranial magnetic resonance imaging (MRI), magnetic resonance cholangiopancreatography, gastroscopy, colonoscopy, tumor markers and autoimmune tests.

Considering sarcoidosis in the differential diagnosis, the patient was evaluated with a thoracic CT scan. No hilar and mediastinal lymph nodes were detected, and the lung parenchyma was normal. She did not report any skin and joint symptoms. Microbiological tests excluded bacterial, fungal, and mycobacterial infections. Renal biopsy was performed due to creatinine progression, hypercalcemia and elevation of 1,25-dihydroxy-vitamin D.

Light microscopy of the renal biopsy showed global sclerosis in 14/23 glomerules, acute interstitial nephritis, nephrocalcinosis and chronic changes in the renal parenchyma. Immunofluorescence examination revealed negative IgG, IgA, IgM, C1q, C3, Kappa, Lambda, and fibrinogen. Histopathological findings, clinical, and laboratory data were evaluated in favor of sarcoidosis.

The patient was evaluated for skin, liver, cardiac, and ocular findings, and peripheral lymph nodes, Extrapulmonary involvement of sarcoidosis could not be detected.

Corticosteroid treatment with methylprednisolone. 0.5 mg/kg by mouth was started. One month later, serum ACE level decreased from 175 U/L to 21 U/L, serum calcium decreased from 11.60 md/dl to 8.10 mg/dl, and creatinine decreased from 4.46 mg/dl to 2.72 mg/dl.

DISCUSSION

The isolated renal form of sarcoidosis is a very rare condition and few cases have been reported in the literature. Our patient was found to have renal sarcoidosis, with no other organ or system involvement. She was noted to have hypercalcemia for 5 years, and no etiology could have been established. The diagnosis of sarcoidosis may be underestimated in the absence of pulmonary manifestations. This may lead to delays in diagnosing and treating the patient.

The diagnosis of sarcoidosis is usually based on clinical and radiological findings, supported by histopathological data. The disease is characterized by the presence of non-caseating granulomas. Infectious and other causes of granulomatous diseases should be ruled out to confirm the diagnosis. The patient's clinical history, family history, and environmental and occupational exposures should also be carefully evaluated. Alternative causes of granulomatous inflammation, especially tuberculosis should be excluded. Berylliosis and hypersensitivity pneumonia can mimic sarcoidosis radiographically and histologically. Our case was noted to have no predisposing exposures. Our case was diagnosed as sarcoidosis by kidney biopsy. Although sarcoidosis is known to be a granulomatous disease, detecting granulomas in the kidney is not required for diagnosis. One of the most common tests used in the diagnosis and follow-up of sarcoidosis is serum ACE level [11]. It may be increased in up to 60% of patients with active sarcoidosis, but this finding is not very specific and may also be due to other diseases. Furthermore, calcium metabolism should be carefully monitored in patients diagnosed with sarcoidosis, as hypercalcemia and hypercalciuria are frequently seen [1].

Liver function tests, especially elevated alkaline phosphatase levels, may indicate liver involvement [1]. Ophthalmologic evaluation is required for eye involvement. Our patient was consulted with Ophthalmology and ocular involvement was not detected.

The gold standard for diagnosing sarcoidosis is the histological detection of non-caseating granulomas [12]. These granulomas can usually be detected in mediastinal lymph nodes, skin, or tissues obtained through transbronchial lung biopsy. Biopsy specimens taken by mediastinoscopy or endobronchial ultrasonography (EBUS) can also be helpful in diagnosis.

In conclusion, it is important to remember that extrapulmonary sarcoidosis can present without pulmonary involvement. In evaluating hypercalcemia, elevated serum ACE levels should give rise to suspicion of sarcoidosis. Examination of the skin and peripheral lymph nodes, a basic ophthalmic examination to screen for ocular involvement, liver function tests, follow-up of serum creatinine for renal involvement, and cardiac evaluation are recommended. Lack of pulmonary manifestations should not rule out the disease in case of other suspicious findings.

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