



RESEARCH ARTICLE

SARCOIDOSIS AND THE KIDNEY: CLASSICAL RENAL MANIFESTATIONS

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ABSTRACT

Sarcoidosis possibly will involve the kidneys in several ways. Incidence and prevalence of renal involvement in sarcoidosis remain uncertain. Usually, aberrations of calcium metabolism include hypercalcemia, hypercalciuria, and nephrocalcinosis, are responsible for the renal manifestations of sarcoidosis. Granulomatous infiltration of the renal interstitium produces severe derangements of renal function. Glomerulonephritis can occur with sarcoidosis, although the pathogenesis remains unclear. Besides renal insufficiency and frank renal failure, nephrotic syndrome, nephrolithiasis, hypertension, and a variety of tubular defects may complicate sarcoidosis. The sensitivity of "sarcoid nephropathy" to corticosteroids usually warrants therapeutic trial. Our case is the first in the literature that demonstrates the renal-confined sarcoidosis. A 29 year male referred to nephrologist for Chronic kidney disease pancreatitis hypertension. During his examination patient had developed renal disease and hypercalcemia. Mild splenomegaly with bilateral increased parenchymal echotexture were seen Interestingly, NO inter bowel free fluid seen, NO gross congestive features identified. NO obstructive uropathy and NO hydronephrosis were seen. Renal insufficiency associated with sarcoidosis was subsequently attributed to renal infiltration by sarcoid granulomas. Despite the presence of sarcoid granuloma in 13% to 22% of kidneys surveyed, the granulomas were usually few in number, limited in extent, and rarely suspected of producing important renal functional impairment.

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INTRODUCTION

Sarcoidosis is a disease of granulomatous inflammation with multi-organ association. The disease manifests itself typically in individuals less than 50 years of age and mainly in the third or fourth decade of life (Baughman, 2003; Thomas, 1987). Renal manifestations include abnormal calcium metabolism, nephrolithiasis, nephrocalcinosis, and acute interstitial nephritis with or without granuloma formation. The classic renal lesion is noncaseating granulomatous interstitial nephritis. Nevertheless, this lesion rarely causes clinically significant renal disease. Hypercalciuria and hypercalcemia are mainly often responsible for clinically significant renal disease. Glomerular disease, obstructive uropathy, and end-stage renal disease (ESRD) are also seen but are uncommon (Muther *et al.*, 1981; Casella, 1993). Individual with a first degree relative with the disease has a fivefold increase of developing sarcoidosis.

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Sarcoidosis affects individuals mostly in their third or fourth decade. Patient resistance also increases with certain associations of genetics and environmental factors. Indeed, combinations of genetic and environmental activators have also been examined, for example: HLA-DQB1 and water damage or high humidity in the workplace. However, it seems that a ubiquitous number of agents may initiate a similar immunologic pathway that is pathognomonic for sarcoidosis (Iannuzzi *et al.*, 2007). The extent and pathogenesis of renal involvement are two central pathways for nephron insult has been validated including granulomatous deposition and deranged calcium management. Pathways are by no means the only two routes of renal involvement, they are the most significant and the overriding themes for renal insult. Sarcoidosis is a universal disorder of unclear etiology, which results from an abnormal cell-mediated immune reaction, and is differentiated by non caseating granulomatous inflammation with epithelioid cells and multinucleated giant cells. Sarcoidosis affects the lungs, but multiple organs such as the central nervous system, liver, heart, skin, and kidney are also involved (Shen *et al.*, 1986).

Mainstay of treatment is based on corticosteroids with good success rates (Rajakariar *et al.*, 2006). Our case is the first in the literature that demonstrates the renal-confined sarcoidosis. referred to nephrologist for Chronic kidney disease pancreatitis hypertension. A 29 year male presented in the emergency department with nausea and anorexia. Pain in abdomen spasmodic type and non radical. Abdominal and renal ultrasound documented splenomegaly, enlarged kidneys (right, 13.5cm; left, 13.9cm) with regular contours and preservation of the sinus parenchyma differentiation. CT scan of abdomen showed acute edematous pancreatitis. Bilateral renal hilar vascular calcification and medullary tips hyperdensities. Patient referred to nephrologist for chronic kidney disease pancreatitis hypertension. Left cervical lymph node aspirated twice.

Moderately cellular smears show lymphoid cells in varying stages of maturation along with many epithelioid cell granulomata. Focal areas of necrosis also seen. Granulomatous lymphadenitis left cervical lymph node. During his examination patient had developed renal disease and persistent hypercalcemia. Mild splenomegaly with bilateral increased parenchymal echotexture were seen Interestingly, NO inter bowel free fluid seen, NO gross congestive features identified. NO obstructive uropathy and NO hydronephrosis were seen. Renal insufficiency associated with sarcoidosis was subsequently attributed to renal infiltration by sarcoid granulomas. The renal biopsy showed interstitial severe inflammatory infiltrate lymphocytes and plasma cells, eosinophils and epithelioid macrophages with formation of granulomas with giant cells. Despite the presence of sarcoid granuloma in 13% to 22% of kidneys surveyed, the granulomas were usually few in number, limited in extent, and rarely suspected of producing important renal functional impairment. The histopathology report indicated granulomatous tubulointerstitial nephritis compatible with the diagnosis of sarcoidosis. Treatment started corticosteroid initially



**Fig 1. Scan of the patient, showing multiple enlarged lymph nodes were seen in left upper cervical region**

Fig 1: Largest one measures 2.3 x 2.1cms. Areas of necrosis are seen, Both thyroid glands are normal in size, position, shape and texture. No focal lesion is seen. On doppler, normal vascularity is noted Isthmus measures 2.8mm and is normal, On doppler, normal vascularity is noted, No focal lesion is noted. Left cervical lymphadenopathy were seen.

## DISCUSSION

Sarcoidosis is a multisystem infection of unknown etiology; the lungs and lymph nodes are commonly involved (30 to 60% of cases). Hypercalcemia (2 to 10%) and hypercalciuria (6 to 30%) cause nephrocalcinosis and nephrolithiasis. Sarcoidosis is a disease that primarily affects the reticuloendothelial system but can affect all tissues and organ systems. This disease affects individuals worldwide and is defined pathologically by the presence of noncaseating granulomas involving the tissue. The etiology of sarcoidosis has yet to be determined but some have proposed a possible infectious etiology. Commonly sarcoid individuals present with hypercalcemia, hypercalciuria, and nephrolithiasis due to the overproduction of calcitriol from the epithelioid granulomas. We also described the rare glomerular and renovascular manifestations of sarcoidosis. Granulomatous interstitial nephritis is most commonly associated with sarcoidosis. It is a histological diagnosis and can be treated with both steroids and TNF-alpha antagonists. Kidney transplantation is safe in patients with sarcoidosis but we must keep in mind the disease can recur in the allograft. In conclusion, sarcoidosis is a complex disease and presents both a diagnostic and management challenge to the clinicians.

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