Sarcoidosis - Renal Manifestation

Dr. Athulya Sadeesh, Dr. Seethalekshmy, Dr. Annie Jojo
Dept of Pathology, Amrita Institute of Medical Sciences, Kochi, Kerala

Abstract
- Sarcoidosis is an illness of granulomatous inflammation with multi-organ association
- Commonly involving the lung and reticuloendothelial system.
- Involvement of the kidney in sarcoidosis is rare and may manifest as proteinuria, hematuria, hypercalcemia, or nephrolithiasis.
- Sarcoidosis presenting initially with renal manifestation is even more rare
- Here we report 3 cases of sarcoidosis presented with renal manifestation and diagnosed by renal biopsy.

CASE-1
- 42 Yr old Male,
- Diabetic (+) 2 yrs on oral hypoglycemic, Detected mild renal dysfunction when evaluated for nephrolithiasis.
- Polyarthralgia : 2 months. No HTN, proteinuria or edema.
- Systemic examination : No palpable lymph nodes, liver and spleen.
- Evaluation showed sub nephrotic proteinuria (1.5 gms/24 hrs), persistent microhemaoturia, mild renal dysfunction (creatinine: 2.6 mg/dL).
- Chest x ray was normal.
- S.Ca: 13.4 mg/dL, S.P: 4.5 mg/dL, S.PTH: 8.3 pg/ml (normal:15-65), S.alb 3.8 gm/dL, Serum alkaline phosphatase: 115 IU/l, ANA profile was negative.
- Immunofluorescence study: negative.
- He was found to have xeroptalmia and xerostomia. Schirmer's test: positive.
- Lip biopsy showed minor salivary glands: non caseating granulomas. Many giant cells showing typical asteroid bodies.
- Renal biopsy done
- Diagnosis: Renal Biopsy: Sarcoidosis (Schirmer test positivity, hypercalcemia, granulomatous inflammation of both kidney and salivary glands and a negative ANA profile).

CASE-2
- 48 yr old female
- Systemic hypertension - 2 yr, B/L iridocyclitis x 8 months, Polyarthralgia - 2 months.
- Hemoptoemegaly. Froaty urine.
- BP: 130/90, 24 hr UP: 1.4mg/dL, S.creat: 3.8mg/dL, S.Alb: 4.6mg/dL, Urine alb: trace. ESR: 25mm/1/hr. ANA, R factor and ANCA negative
- Clinical Impression: 7 IAGN.
- Renal biopsy done
- Diagnosis: Renal Biopsy: Sarcoidosis (Schirmer test positivity, hypercalcemia, granulomatous inflammation of both kidney and salivary glands and a negative ANA profile).

CASE-3
- 47 yr old male
- <i>k/c/o</i> Type 2 Diabetes mellitus and systemic hypertension.
- c/o anorexia, myalgia and acute weight loss (around 12kg within 1 month). H/o intermittent fever.
- Clinical Impression: ? Drug induced interstitial nephritis. ? Multiple myeloma

Microgranulomas composed of epithelioid cells and giant cells

Renal biopsy done
- Glomeruli appear normal in size.
- Tubules show mild epithelial vacuolation and calcific crystals in the lumina. A few tubules also reveal lymphocytic infiltration of epithelium.
- Interstitium also reveal 2 discrete non caseating microgranulomas composed of epithelioid cells and giant cells. 2 similar granulomas are noted in the renal capsule also.
- Interstitium also reveal a few calcified lamelated concretions(von kossa positive).
- Immunofluorescence study: negative
- Diagnosis: Renal Biopsy: Acute on chronic tubulointestinal nephritis with interstitial microgranulomas, mild global glomerulosclerosis and hyperensive vascular changes.

Microscopy and Immunohistochemistry
- Masson trichrome
- PAS (Negative)
- AFB (Negative)
- Jones Silver
- Von kossa stain (calcific lamelated concretions)

Discussion
- Sarcoidosis is an inflammatory disorder characterized by noncaseating epithelioid cell granulomas commonly involving the lung and reticuloendothelial system.
- Pulmonary manifestations-common.
- 10%-30% of sarcoidosis presents with extra pulmonary manifestation.
- ACE levels are seen increased in patient serum
- Renal manifestations
  - Hypercalcemia (40-50%)
  - Hypercalcuria (5-10%)
  - Nephrocalcinosis
  - Granulomatous interstitial nephritis
  - Rarely glomerulonephritis
- Reported patterns of kidney injury:
  - Intertstitial nephritis (with or without granulomatous inflammation),
  - Minimal change disease,
  - Focal segmental glomerulosclerosis,
  - Membranous glomerulopathy,
  - IgA nephropathy,
  - Membranoproliferative glomerulonephritis crescentic glomerulonephritis (with positive or negative [ANCA] test results).

Pathogenesis

- Hypercalcemia in sarcoidosis
- [Hypercalcemia in sarcoidosis]
- [Pathogenesis diagram]

- Main stay of treatment of sarcoid with renal involvement is glucocorticoids.
- Renal 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

References
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