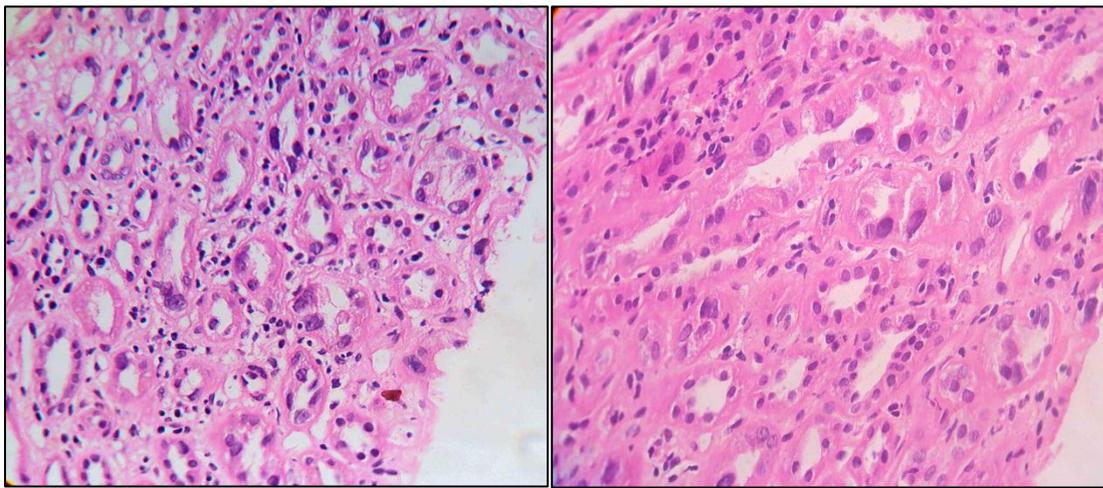
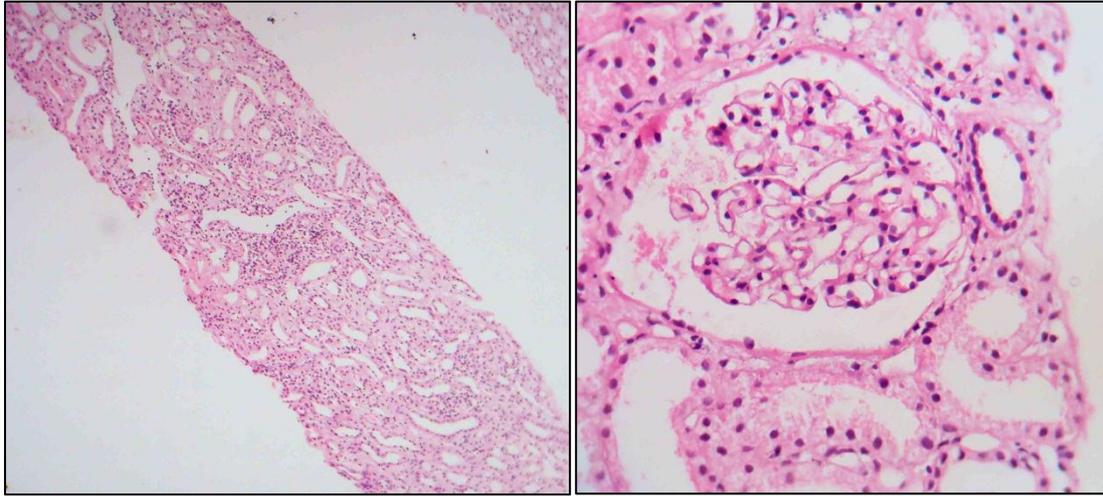


Image Quiz 1

Diagnosis: Karyomegalic Tubulointerstitial nephritis



Microscopic Examination

Low magnification shows features of patchy interstitial inflammation. Glomeruli were essentially unremarkable. On closer examination there were bizarre changes in the tubular epithelial cell nuclei including nucleomegaly (karyomegaly), alteration of chromatin texture, hyperchromasia, nuclear pleomorphism and occasional multinucleation. Changes were not very different from what we call viral cytopathic effects. Blood vessels (not shown) were relatively unremarkable.

Further details of the case:

There was no history of significant drug/therapeutic ingestion including exposure to mycotoxins or other herbs. The patient was in good health until the onset of renal disease. There was no history of recurrent respiratory tract infections or specific viral infections. His liver functions were normal throughout the disease course. Serum IgG4 levels were normal. I did stain the biopsy for BK virus and CMV; both were negative.

Discussion:

Karyomegalic nephropathy was first identified in 1974 by Burry [1]. Mihatsch *et al.* [2] subsequently described the disorder in 1979 and proposed a new disease syndrome. Reports of few additional cases with similar findings have been cited in the literature.[3,5-8]. Classically patients present with a history of recurrent upper respiratory infections and progressive renal failure. Extra-renal manifestations are uncommon, although karyomegalic cells have been identified in numerous tissues including brain astrocytes, intestinal smooth muscle, Schwann cells of peripheral nerves, and bile duct epithelium. Transient elevations in liver enzymes may be seen. Histologically, the presence of interstitial nephritis in conjunction with atypical epithelial cell and nuclear polymorphism, predominantly of tubular cells, is characteristic. The absence of other environmental factors associated with interstitial nephritis (non-steroidal anti-inflammatory agents and heavy metals) and other glomerular disorders, are suggestive of karyomegalic nephropathy.

Key Point:

While evaluating biopsy of a patient with unexplained renal failure, careful examination of tubular epithelial cells for features of unexplained karyomegaly should raise the suspicion of this disease.

References:

1. Burry AF. Extreme dysplasia in renal epithelium of a young woman dying from hepatocarcinoma. J Pathol 1974; 113: 147-150

<http://www.ncbi.nlm.nih.gov/pubmed/4372331>

2. Mihatsch MJ et. al. Systemic karyomegaly associated with chronic interstitial nephritis. A new disease entity? Clin Nephrol. 1979 Aug; 12(2):54-62.

<http://www.ncbi.nlm.nih.gov/pubmed/527271>

3. Bhandari S et. al. Karyomegalic nephropathy: an uncommon cause of progressive renal failure Nephrol Dial Transplant (2002) 17: 1914-1920

<http://ndt.oxfordjournals.org/content/17/11/1914.full.pdf+html>

4. John Molne: Lecture presentation ; Karyomegalic tubulointerstitial nephritis : Case 3

<http://www.nephropathology-esp.org/meeting-lecture/slide-seminar-karyomegalic-tubulointerstitial-nephritis>

5. Spoenclin M et. al. Karyomegalic interstitial nephritis: Further support for a distinct entity and evidence for a genetic defect. American Journal of Kidney Diseases 1995;25(2): 242-252.

[http://www.ajkd.org/article/0272-6386\(95\)90005-5/abstract](http://www.ajkd.org/article/0272-6386(95)90005-5/abstract)

6. Monga G, Banfi G, Salvatore M, Amatruda O, Bozzola C, Mazzucco G. Karyomegalic interstitial nephritis: report of 3 new cases and review of the literature. Clin Nephrol. 2006 May;65(5):349-55.

<http://www.ncbi.nlm.nih.gov/pubmed/16724656>

7. Baba F, Nanovic L, Jaffery JB, Friedl A. Karyomegalic tubulointerstitial nephritis - a case report. Pathol Res Pract. 2006;202(7):555-9.

<http://www.ncbi.nlm.nih.gov/pubmed/16678356>

8. Verine J, Reade R, Janin A, Droz D. Karyomegalic interstitial nephritis: A new French case. Ann Pathol. 2010 Jun;30(3):240-2.

<http://www.ncbi.nlm.nih.gov/pubmed/20621605>

Posted on 08-02-2012

© Dr. Alok Sharma

www.nephro-pathology.com