Article published online: 2022-06-17

QUIZ

A Fifty Nine Year Old Female with Flank Pain and Hematuria

Anjali A Satoskar

Department of Pathology, The Ohio State University, Columbus, Ohio, United States of America.

Corresponding author: Anjali A Satoskar Email: Anjali.Satoskar@osumc.edu
Published: 25 July 2012
Ibnosina J Med BS 2012,4(4):134-136
Received: 30 May 2012
Accepted: 03 June 2012
This article is available from: http://www.ijmbs.org

Abstract
The patient was a 59 year old Caucasian female who presented with flank pain and gross hematuria. She had similar symptoms approximately 3 months ago prior to presentation and has been on antibiotics for presumed pyelonephritis. The clinical course, differential diagnosis and pathological findings are discussed in detail

Key words: Haematuria, Renal pain, Pyelonephritis,

Case History
The patient was a 59 year old Caucasian female who was being evaluated for flank pain and gross hematuria. She had similar symptoms 3 months previously and has been on antibiotics for presumed pyelonephritis. An abdominal computed tomogram (CT) scan done during this period revealed multiple small cysts in the kidney. Serum creatinine was around 2.0 mg/dl 2 years ago. Current serum creatinine was 6.0 mg/dl. No definite history of proteinuria or edema.

Laboratory results
C3: 111 mg/dl, C4: 19 mg/dl, anti-nuclear antibody (ANA) and anti-neutrophil cytoplasmic antibody (ANCA) serology was pending at the time of the biopsy. Anti-glomerular basement membrane (anti-GBM) antibody less than 0.2 U/ml, hemoglobin 8.3 g/dl. Patient's weight was 212 pounds, blood pressure was 142/72 mm Hg and body mass index was 28 kg/m^2.

Biopsy
Biopsy specimen contained predominantly renal medulla and cortico-medullary junctional tissue with up to two glomeruli. One glomerulus appeared unremarkable and one was at the edge of the tissue. The findings in the renal medulla are shown in the figures 1, 2. The renal cortex is shown in figure 3. The one glomerulus at the edge of the tissue is shown in figure 4.

Pathology Quiz

1. What does the renal medulla show?
a. Allergic interstitial nephritis.
b. Acute pyelonephritis.
c. Necrosis
d. Necrotizing inflammation.
e. Hemorrhage.

2. What does Fig. 3 show?
a. Interstitial fibrosis and tubular atrophy.
b. Acute tubular injury and interstitial edema.
c. Both.
d. None of the above

3. Fig. 4 shows:
a. Segmental sclerosis.
b. Intracapillary proliferative lesion.
c. Mesangial expansion.
d. Extracapillary proliferative lesion/crescent.
e. Unremarkable glomerulus.

4. Which lab test would be crucial in this case?
a. ANCA serology.
b. Blood culture.
c. Urine culture.
d. ANA serology and rheumatoid factor.

Pathology Report

The biopsy shows necrotizing inflammation of the vasa recta and collecting ducts in the renal medulla (Fig. 1 and 2).

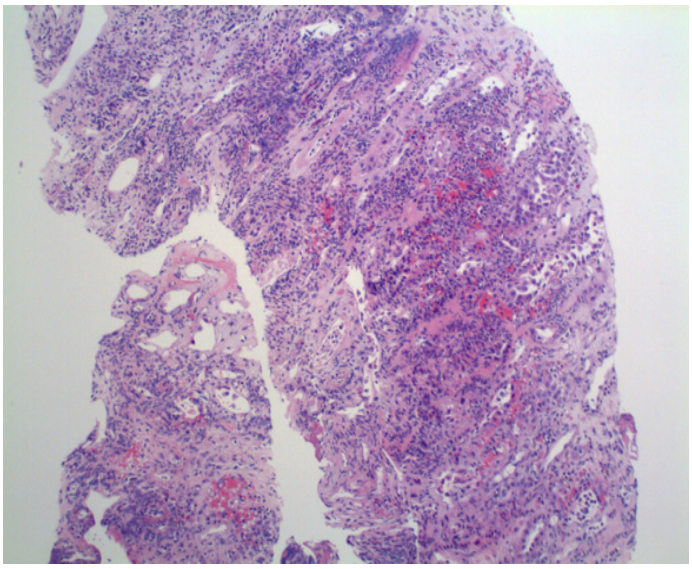

Figure 1

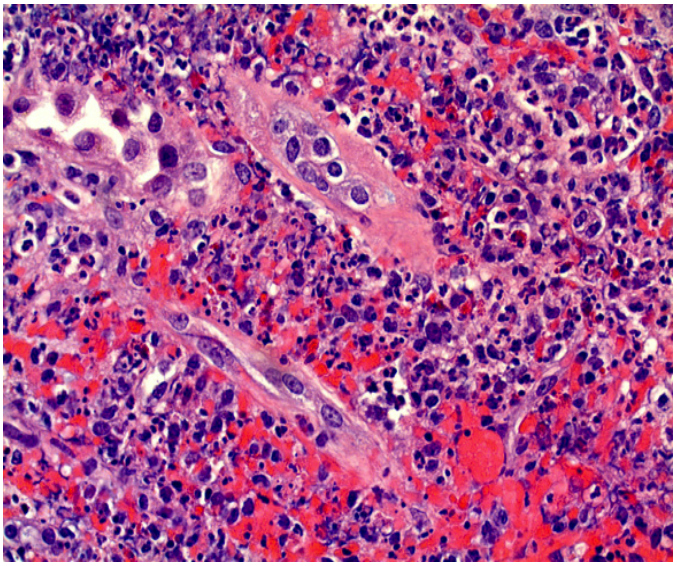

Figure 2

The inflammatory cell infiltrate contains many neutrophils along with mononuclear cells. There is fibrinoid necrosis with cellular and nuclear karyorrhexis and hemorrhage. The renal cortex in the biopsy was small and consisted of mainly the cortico-medullary junctional tissue with two to three glomeruli. There were four glomeruli in the frozen portion of tissue submitted for immunofluorescence studies. Only one glomerulus shows a distinct cellular crescent (Fig. 4). The renal cortex shows acute tubular injury and interstitial edema (Fig. 3). This is secondary to the active inflammatory process going on in the kidney. Direct immunofluorescence and electron microscopic examination did not show immune-type deposits. The ANCA serology was pending

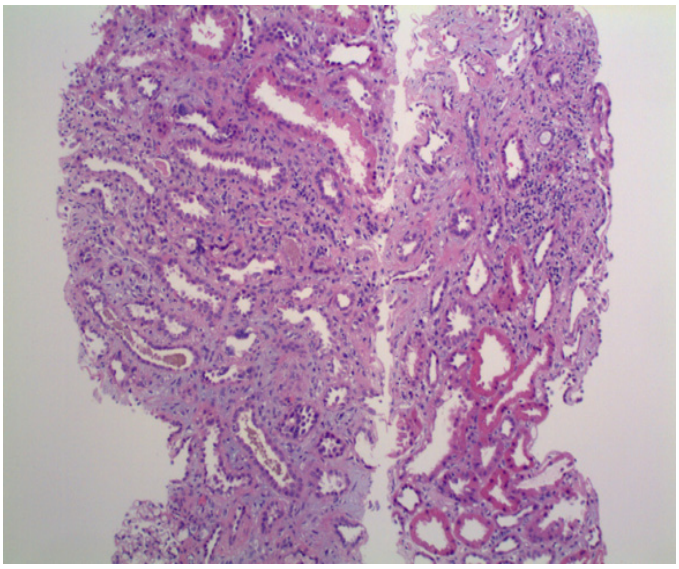

Figure 3

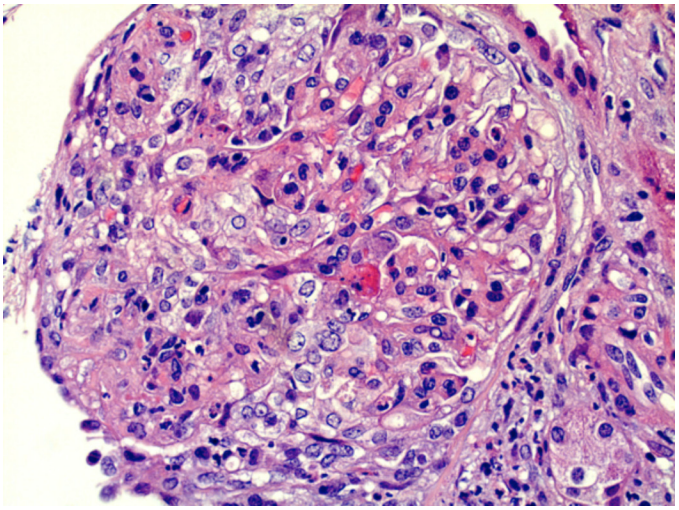

Figure 4

at the time of the biopsy. This case was signed out as :
Active necrotizing inflammation and hemorrhage in the renal medulla with a single glomerulus showing crescent, raising the possibility of pauci-immune crescentic and necrotizing glomerulonephritis and vasculitis.

Differential Diagnosis

1. ANCA associated pauci-immune glomerulonephritis
2. Acute interstitial nephritis
3. Pyelonephritis

Discussion

ANCA associated pauci-immune glomerulonephritis can occur as a systemic disease usually with renal and lung involvement (Wegner's granulomatosis, microscopic polyangiitis, Churg-Strauss disease), or renal limited vasculitis and some drug-induced ANCA associated vasculitis. The classic finding on a renal biopsy is a focal necrotizing and crescentic glomerulonephritis. It is thought to be a glomerulocentric disease. What is unusual about this case is the extensive necrotizing inflammation in the medulla. It is difficult to estimate the prevalence of medullary involvement in cases of ANCA vasculitis, the reason being that the renal medulla is inconsistently sampled in routine biopsies. Diagnosis of renal diseases is usually made on the findings in the renal cortex. The microenvironment in the renal medulla is different from the cortex because of the hypertonic interstitium and slightly different blood supply of the inner medulla. Two of the case series published on this topic are given below (1, 2). ANCA vasculitis can manifest as necrotizing capillaritis/arteriolitis in the medulla as well as papillary necrosis. Other diseases that can be associated with papillary necrosis include severe diabetic nephropathy, analgesic nephropathy, and sickle cell disease. The clinical scenario in this patient does not fit all these aforementioned conditions. Another differential diagnosis in this case includes acute pyelonephritis which usually involves the renal medulla (ascending infection). But necrotizing inflammation and hemorrhage would be uncommon in pyelonephritis. Also, in acute pyelonephritis, polymorphonuclear cells inside tubules (tubular microabscesses) are more common which is not apparent in this case. The single glomerulus with crescent formation was also supportive of a diagnosis of pauci-immune glomerulonephritis. Acute drug-induced interstitial nephritis is another consideration in the differential, but the necrotizing inflammation centered around the vasa recta rather than a diffuse infiltrate, with glomerular crescent is more supportive of a vasculitic process. In this case, ANCA serology came back positive for pANCA with 1:160 titer for anti-myeloperoxidase by ELISA.

Conclusions

ANCA associated pauci-immune crescentic glomerulonephritis is not just a glomerular disease, and can develop medullary lesions such as necrotizing capillaritis, arteriolitis and papillary necrosis.

References

1. Bonsib SM, Goeken JA, et al. Necrotizing medullary lesions in patients with ANCA associated renal disease. Mod Pathol 7(2):181-5.
2. Jennette JC, Wilkman AS, et al. Anti-neutrophil cytoplasmic autoantibody-associated glomerulonephritis and vasculitis. Am J Pathol 135(5):921-30.

Answers:
1. d
2. c
3. d
4. a