Diagnostic Challenges of Xanthogranulomatous Pyelonephritis

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Learning Objectives

1. Recognize the clinical and radiologic features of xanthogranulomatous pyelonephritis (XGPN).
2. Develop a differential diagnosis of common mimics of XGPN.

Case Presentation

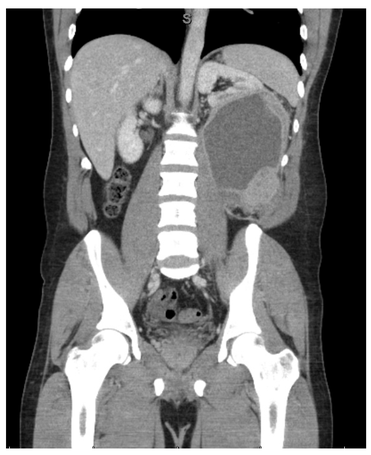
History

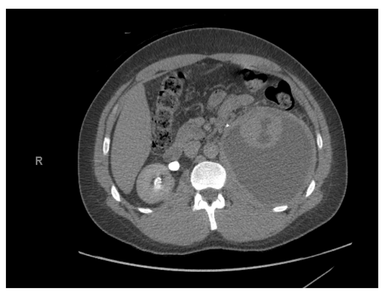
- 44-year-old male with a history of nephrolithiasis presented to an outside emergency department complaining of progressive left-sided flank pain for several months
- ROS: Positive for anorexia, nausea, weight loss; Negative for night sweats, fevers

Physical Exam/Labs

- P 111, T 36.7 C, R 18, BP 122/67, O2 93% on RA
- Pertinent positives: Left CVA tenderness, abdomen is soft and tender to deep palpation on left lower quadrant where there is firm mass , well-healing chevron incision on abdomen & left lateral lumbar spinal column
- Labs: WBC 82, Hgb 8.2, Plt 512, Cr 1.25, CRP 22.3, ESR 138, Ferritin 2,106

Imaging

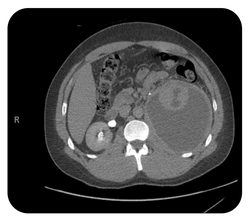




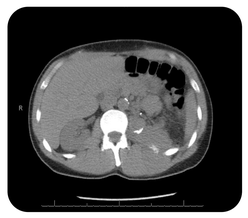
CT abd/pelvis with contrast showing large mass extending from inferior pole of left kidney

Differential Diagnosis/Hospital Course

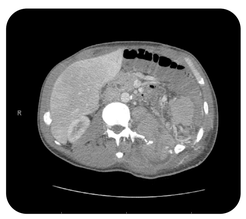
Broad differential considered:
Infectious, rheumatologic/autoimmune, malignancy



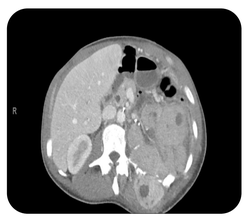
July-October 2017
- Presented with abdominal pain, biopsy with XGPN
- Left nephrectomy for definitive management



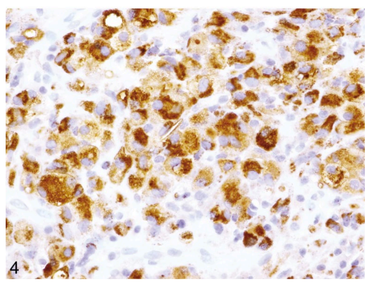
January 2018
- Recurrent abdominal pain, CT with recurrent mass
- Repeat biopsy with XGPN
- BM biopsy for persistent leukocytosis without malignancy



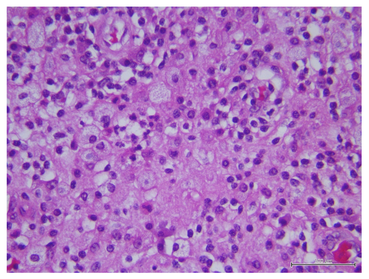
March 2018
- CT with rapid enlargement of mass, invasion through posterior abdominal wall, paraspinal muscles, ribs
- Debulking procedure



April 2018
- Repeat biopsy revealed +S100 with MDM2 amplification consistent with liposarcoma
- Sarcoma tumor board recommended neoadjuvant chemoradiation



Replacement of renal parenchyma with CD68+ foamy histiocytes



Histologic examination reveals foamy histiocytes with a background of acute & chronic inflammatory infiltrates

Discussion

- Prior to arriving at the correct diagnosis, patient underwent three renal biopsies, nephrectomy, a month of antibiotics, and had four consulting services involved.
- Recognizing XGPN and its mimics, and maintaining a broad differential diagnosis are critical to effective diagnosis and treatment.

XGPN Recognition and Differential Diagnosis

Clinical Features of XGPN:
- Common in 4th or 5th decade of life, female > male
- History of recurrent UTIs and nephrolithiasis
- Non-specific symptoms of flank pain, fever, malaise, weight loss, anorexia
- UA with hematuria, bacteruria (commonly Proteus and E.coli species implicated)
- Elevated inflammatory markers, anemia, leukocytosis
- Complications include fistulas and abscess formation

Radiologic Features of XGPN:
- CT scan: unilateral replacement of renal tissue with numerous low density areas with peripheral enhancement
- "Bear's paw sign"
- Ultrasound: enlarged kidney with multiple fluid-filled masses

Differential Diagnosis:
- Infection: renal/perinephric abscess, other forms of pyelonephritis
- Malignancy (Clear renal cell carcinoma, liposarcoma, leiomyosarcoma, etc.)
- Megalocytic interstitial nephritis, Malakoplakia

Treatment:
- Antimicrobials to treat infection, usually insufficient
- Definitive treatment is nephrectomy

References

1) http://www.pathologyoutlines.com/caseofweek/case323image6.jpg
2) https://radiopaedia.org/articles/xanthogranulomatous-pyelonephritis
3) http://www.auanet.org/education/auauniversity/education-products-and-resources/pathology-for-urologists/kidney/inflammatory/necrotic-renal-lesions/xanthogranulomatous-pyelonephritis