CLINICAL VIGNETTE

Xanthogranulomatous pyelonephritis

Avi Baskin, MSIII, and Maria Romanova, M.D.

Case Presentation

A 53-year-old man with a past medical history of nephrolithiasis, hypertension, benign prostatic hypertrophy, anemia, and Hepatitis C presented to the emergency department for evaluation after a CT urogram ordered by his primary care provider revealed an enlarged right kidney with dilated collecting system.

In the emergency department, the patient confirmed a constant stabbing, right lower quadrant pain along with night sweats and fevers. He confirmed losing 20 pounds in the past month. The pain was similar to his prior episodes of nephrolithiasis. Review of systems was negative for lower urinary tract symptoms, dysuria, or hematuria.

Vitals were notable for fever to 100.1 degrees Fahrenheit and a blood pressure of 147/93. Physical examination revealed tenderness at the right costovertebral angle and in the right lower quadrant. A firm, nodular contoured mass was felt on the right flank. Complete blood count revealed a normocytic anemia with a hemoglobin of 7.9 g/dL. Iron studies were indicative of anemia of chronic disease. ESR was markedly elevated at 115. Urinalysis was positive for numerous leukocytes and spot protein. Urine cultures grew over 100,000 colonies of *Citrobacter koseri*. HIV antibodies and TB quantiferon gold both came back negative.

The CT urogram showed a markedly enlarged right kidney (18.9 cm), severely dilated renal collecting system, a small lower pole renal calculus, perinephric fat stranding, and small loculated fluid collections in the hepatorenal space. There was also evidence of decreased renal function with no excretion of contrast on delayed imaging (Figure 1).

The patient was started on broad-spectrum antibiotics for his apparent urinary tract infection. His fever and right lower quadrant pain resolved in the first few days of hospital stay. Additionally, a double-J stent was placed in the right ureter with the goal of relieving hydronephrosis. Immediate drainage of purulent material through the stent into the bladder was observed during the procedure. Subsequent complete kidney imaging with vascular flow confirmed improvement of collecting system dilatation. However, relative renal function of the right kidney was only 14%.

The patient was discharged home and followed as an outpatient. His anemia remained stable and required no repeat transfusions. His double-J stent was removed from the right ureter and repeat renal functional study showed no right renal function observed on radionuclide renal scan. Given the history of nephrolithiasis, a non-functioning right kidney, and both clinical and radiological suspicion for xanthogranulomatous pyelonephritis (XGP), open nephrectomy was performed 6 months after initial presentation.

Histological examination revealed diffuse destruction of glomeruli by mixed inflammatory infiltrate of lymphocytes and lipid-laden macrophages, a dilated collecting system, areas of fibrosis, and a single stone obstructing the ureteral orifice consistent with XGP (Figure 2).

The patient recovered well from the nephrectomy and maintained stable renal function and hemoglobin level.

Discussion

XGP is a variant of chronic pyelonephritis in which there is massive destruction of the kidney by granulomatous tissue.[1] The most common age of XGP occurrence is from 45 to 55, but reported cases ranged from 2 to 84 years.[2] Women are more likely to be affected than men. Overall, XGP accounts for only 0.6% cases of chronic pyelonephritis.[2]

While the exact pathogenesis of XGP is unknown, there is typically an underlying urinary tract obstruction that becomes complicated by infection. Infection, such as C*itrobacter koseri* in our example, leads to ischemia, damage of parenchymal tissue, and granuloma formation with subsequent accumulation of lipid deposits within macrophages.[3] In most cases, XGP occurs unilaterally.[1] Tuberculosis and HIV infection may present in similar manner and should be ruled out.

Symptoms of flank or abdominal pain, fever, and weight loss are common in patients with XGP.[2] Generally, symptoms are similar to those of chronic pyelonephritis.[3] Laboratory findings show leukocytosis (41% of cases) and anemia (63%). Urinalysis reveals pyuria in most cases (57%) and positive culture in the majority of patients. One third of patients may have negative cultures due to recent antibiotic use. ESR is usually elevated (94%).[2]

CT scans generally provide the clue to diagnosis. A large nonfunctioning kidney with a contracted renal pelvis as well as enlargement of the calices and inflammation is strongly suggestive of XGP.[2] Ultrasound is non-specific and would only show an enlarged kidney with large central echogenicity representing a calculus.

XGP is characterized by a granulomatous mixed inflammatory infiltrate with fibrosis and cholesterol clefts.[2]

Macrophages, foamy cytoplasm, plasma cells, and giant cells can all be seen on histology. Of note, the cells can mimic the appearance of clear cell renal cell carcinoma

For focal XGP, patient can be treated with antibiotics. The pyelonephrosis can be more immediately managed with drainage of the infected collecting system. Drainage of the obstruction through the lower urinary tract should be avoided in septic patients as this may exacerbate their clinical state.[1]

Surgical treatment of XGP is often indicated because conservative measures such as antibiotics fail to treat the underlying pathological process. XGP has the potential to grow to involve the surrounding structures such as the liver, duodenum, colon, and great vessels. At this time, surgical treatment remains the only definitive option.[4]

Conclusion

XGP is a rare form of chronic pyelonephritis that should be on the differential diagnosis for patients with recurrent urinary tract infections who show signs of hydronephrosis on imaging.

Figures

Figure 1.

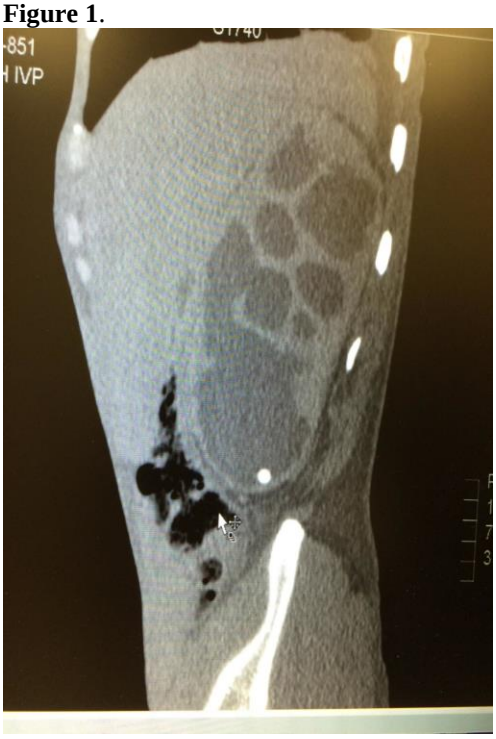
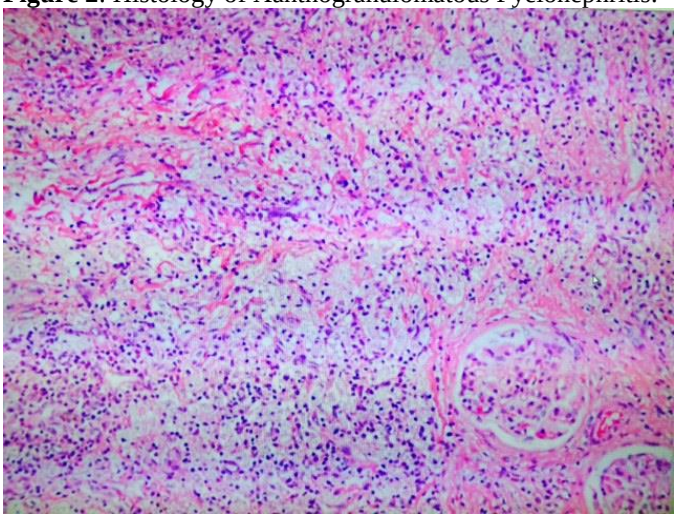


Figure 2: Histology of Xanthogranulomatous Pyelonephritis.



REFERENCES

1. **Nguyen, H.T.** Bacterial Infections of the Genitourinary Tract. In: McAninch JW and Lue, TF, editors. Smith & Tanagho's *General Urology*, 18e. New York: The McGraw-Hill Companies; 2013.
2. **Li L, Parwani AV**. Xanthogranulomatous pyelonephritis. *Arch Pathol Lab Med*. 2011 May;135(5):671-4. doi: 10.1043/2009-0769-RSR.1. Review. PubMed PMID:21526966.
3. **Lerma EV.** Chronic Tubulointerstitial Nephritis. In: Lerma EV, Berns JS, Nissenson AR, editors. *Current Diagnosis and Treatment Nephrology and Hypertension*. New York: The McGraw-Hill Companies; 2009.
4. **Addison B, Zargar H, Lilic N, Merrilees D, Rice M.** Analysis of 35 cases of Xanthogranulomatous pyelonephritis. *ANZ J Surg*. 2015 Mar;85(3):150-3. doi:10.1111/ans.12581. Epub 2014 Mar 25. PubMed PMID: 24661744.

Submitted October 21, 2015