An Unusual Case of Xanthogranulomatous Pyelonephritis
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Citation

Abstract
Xanthogranulomatous pyelonephritis (XGPN) is an uncommon form of chronic infectious tubulo-interstitial nephritis. It commonly presents in the age group of 50-70 years and with primary female predominance. It is always accompanied with systemic manifestations and usually presents in some or the other form of complications. In our case, a young, 20-year-old male was affected with this pathology, without any obvious systemic manifestations of the disease or any of its complications. We would like to present such an unusual case at our institute.

CASE HISTORY
A 20-year-old Hindu male patient came to the outpatient department with dull aching pain in the right loin, on and off since six months. The pain was not accompanied with fever or any urinary disturbances. There was no history of trauma or bowel disturbances or other major illnesses or any surgery done in the past. There was no history of any addictions, either.

On investigating, the patient’s CBC showed: Hb 12.3g%, tc 9800/cumm and ESR 47; creatinine was 1.1 mg/dl. Urine routine showed occult blood, few pus cells and proteinuria. An ultrasound of abdomen and pelvis showed a large hydroureteric right kidney with internal echoes and thinned-out parenchyma with a single central 1.3cm calculus, with the opposite kidney being normal. The rest of the viscera were normal. CT scan, plain & with contrast, showed global involvement of the right kidney with a large mass and a central large calculus with destroyed renal parenchyma, replaced with a low-attenuated mass, and a highly thickened Gerota’s fascia with dense adhesions within. The affected kidney was non-functioning. DTPA renal scan showed 2% function in the affected kidney with a normally functioning left kidney.

A decision to perform open right nephrectomy was taken keeping in mind a differential diagnosis of renal cell carcinoma (RCC) or XGPN. At surgery, all CT-scan findings were confirmed and the specimen was sent for histopathology study. A drain was kept in situ which was removed after 5 days. The sutures were removed after 10 days. The patient’s recovery was uneventful. Now, at 2-year follow-up, the patient is asymptomatic.

On gross pathology, the kidney measured 10 x 7.4 x 5.6cm with indurated perinephric fat and a 1.3cm calculus at the dilated renal pelvis. Soft yellow nodules replaced the corticomedullary junction and the calyces were filled with pus and debris. The renal parenchyma was destroyed: Stage 2 XGPN.

Figure 1
Figure 1: Large kidney with yellow nodules replacing the cortico-medullary junction and calyces filled with pus and debris.

XGPN on microscopy
DISCUSSION

XGPN is a serious debilitating illness characterized by an infectious renal phlegmon. This disease process shares many characteristics with a true neoplasm in terms of its radiographic appearance and ability to involve adjacent structures or organs. Treatment of XGPN is generally extirpative and can pose a formidable challenge to the surgeon. Schlagenhaufer first described XGPN in 1916. It is defined as a chronic inflammatory disorder of the kidney characterized by a mass originating in renal parenchyma. The condition has a common association with Proteus or Escherichia coli infection. Pseudomonas species have also been implicated. The kidney is usually non-functional (1). XGPN is almost always unilateral and only one patient with bilateral disease has been described so far. The kidney is either involved globally or focally. Changes of XGPN have been described in kidneys destroyed as a result of pyonephrosis; in renal cell carcinoma; in transitional cell carcinoma; and rarely in renal cysts. These focal pathological changes are detectable only by using histologic analysis and usually they do not appear on images. It occurs in 1% of all renal infections. Although it is rare in the pediatric population, its incidence in pediatric nephrectomy is approximately 16%. XGPN is 4 times more common in females than in men and is usually noted in the fifth and sixth decades of life.

The exact etiology of XGPN is unknown, but it is generally accepted that the disease process requires long-term renal obstruction and infection. Stones (frequently of staghorn proportions) may occur in 75% of patients with XGP but are not required to make the diagnosis. XGPN is often observed in patients with diabetes or in patients who are immunocompromised. Abnormal lipid metabolism has also been hypothesized as an etiological factor in patients with XGP.

Pathophysiology: XGPN displays neoplasm-like properties capable of local tissue invasion and destruction and has been referred to as a pseudotumor. Adjacent organs, including the spleen, pancreas and duodenum may be involved. For this reason, Malek and Elder proposed the following stages of XGP involvement:

XGPN has been divided in 3 stages: Stage 1: the lesion is confined to the kidney; stage 2: the pathologic process extends to Gerota’s fascia; stage 3: the process spreads to the paranephric space and to other retroperitoneal structures.

Treatment options: Medical therapy alone is inadequate to treat XGP. Antibiotics may be appropriate as the temporizing measure for patients requiring medical work-up prior to nephrectomy. The choice of antibiotic should be geared towards the identity and sensitivity of the organism. Proteus and E. coli are usually sensitive to a variety of antibiotics, including first-generation cephalosporins and trimethoprim-sulfmethoxazole.

Surgical therapy: Nephrectomy is the standard procedure for XGPN. These are particularly challenging cases, especially if there is local organ involvement as the disease mimics renal cell carcinoma macroscopically. The goal is to remove all the involved granulomatous tissue. If this is not accomplished, the remaining infected tissue may lead to formation of cutaneous fistulae. Laparoscopic nephrectomy is a feasible option nowadays. Small pediatric cases show better outcome with laparoscopy. The increased use of hand-assisted laparoscopy (HALS) may allow for an acceptable compromise between technical feasibility and patient morbidity. Laparoscopic approach may be undertaken in all cases, but the patient and the patient’s family can expect a conversion rate approaching 50%. The basic principles of nephrectomy apply to the extirpation of renal XGPN. Partial nephrectomy is performed where the disease is focally present. If needed, a radical nephrectomy may be performed, since the mass may represent a neoplasm. All involved tissue must be removed. Liberal irrigation with antibiotic fluid is done and a suction drain is kept in situ.

Immediate postoperative care mirrors that of a standard...
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nephrectomy: Watch for signs of sepsis; institute aggressive pulmonary care; and encourage early ambulation. Follow-up: Annual imaging of the contralateral urinary tract is important. Aggressive treatment of all urinary tract infections is a must. If possible, isolate and treat all predisposing factors. Evaluation of lower urinary tract pathology and voiding dysfunction may be important in these patients.

Complications: Bleeding should be aggressively controlled. Splenic injuries are to be avoided during left-side nephrectomies. Pancreatic injuries should be avoided. On the right side, hepatic and duodenal injuries should be avoided. The colon is vulnerable to damage on either side. Vascular injuries, mainly on the left side, involving vena cava, require great care and maybe CVTS standby. Post-operative fistulae and abscesses should be avoided.

The overall prognosis for XGPN is good as it is a benign entity. Death from this pathology is exceedingly rare, although its morbidity is substantial. Nephron-sparing surgery is recommended in many centers as a hope to save the affected kidney, but only when XGPN is involving the kidney focally. Further study is needed to arrive at the exact pathophysiological mechanism and etiology of XGPN.

Our case showed a relatively asymptomatic young male, without any systemic involvement or any other complications, thus making this case unusual. Now, even after a 3-year follow-up, the patient is asymptomatic without any contralateral affection and healthy.

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References
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