DOI: 10.7860/JCDR/2021/50550.15594

Case Report

Surgery Section

Xanthogranulomatous Pyelonephritis- A Diagnostic Dilemma

KRISHNENDU MAITI[1], PARTHA PRATIM SINHA ROY[2], DILIP KUMAR PAL[3]

ABSTRACT

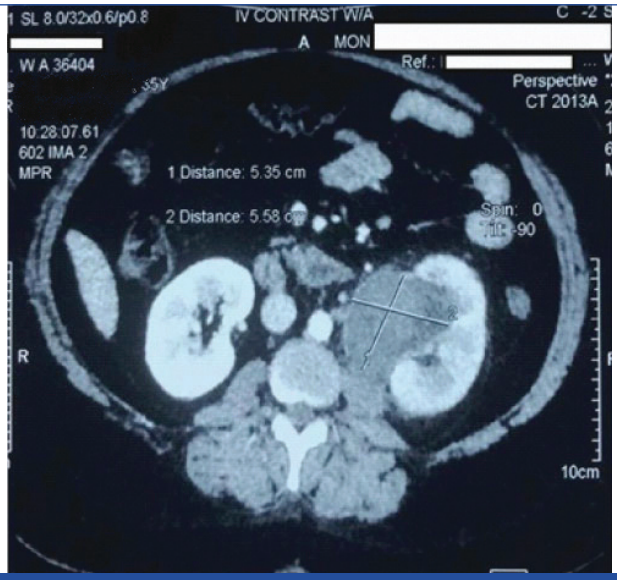
Xanthogranulomatous Pyelonephritis (XGPN) is a chronic inflammatory disease usually associated with renal stones, mimicking several malignant and benign pathology causing diagnostic dilemma. A rare case of XGPN of renal pelvis has been presented here. A 34-year-old female presented with left renal colicky pain, with gross haematuria without any palpable renal lump. Computed Tomography (CT) scan showed a 4.5×4.3×4.0 cm sized heterogeneous solid cystic non enhancing Space Occupying Lesion (SOL) in left renal pelvis. It was provisionally diagnosed as transitional cell carcinoma of left renal pelvis and patient underwent laparoscopic nephroureterectomy. Histopathology revealed it as XGPN. This highlights the fact that there are several overlaps of clinical and radiological findings of these entities making its diagnosis a challenging task.

Keywords: Inflammatory diseases, Malignancy mimickers, Renal lesions

CASE REPORT

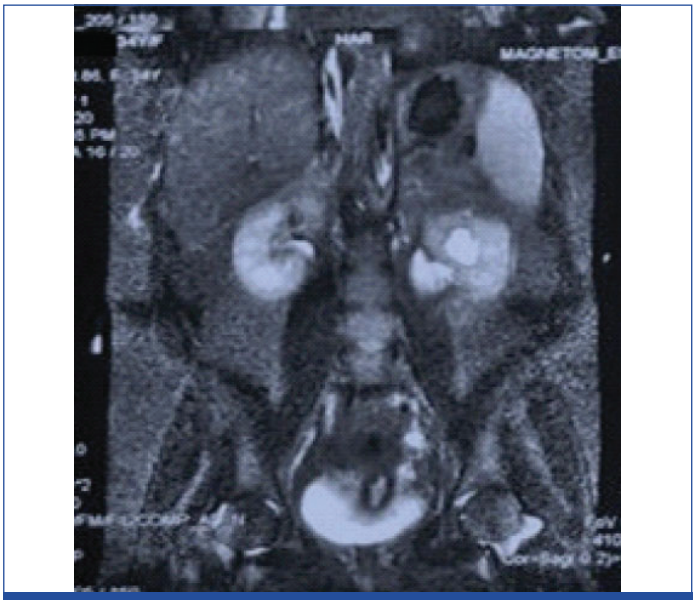
A 34-year-old female patient presented to the Urology Outpatient department with complaints of intermittent left renal colic and gross haematuria, for the past four months. The patient denied any history of fever or previous episodes of haematuria or any history of hypertension or diabetes. On clinical examination, she was anaemic, afebrile, blood pressure of 128/68 mm of Hg. There was no tenderness or any palpable lump per abdomen. Investigations showed her haemoglobin was 9 gm/dL and total leukocyte count of 6.3×10^9/L with a normal differential count. Biochemical investigations revealed blood urea of 14 mg/dL, and a serum creatinine of 0.8 mg/dL. The serum electrolyte was normal. Urine culture was sterile. Ultrasound of the whole abdomen revealed left kidney of normal size and echotexture. However, a hypoechoic lesion with exophytic component was seen arising from pelvis measuring 34.3×32.4 mm, suggestive of a mass lesion.

A Contrast Enhanced Computed Tomography (CECT) showed a 4.5×4.3×4.0 cm sized heterogeneous solid cystic non enhancing SOL in left renal pelvis. The pelvicalyceal system was focally dilated in upper pole of left kidney [Table/Fig-1]. There was no evidence of any calculus or hydronephrosis. Both kidneys had normal excretion. Both ureters were normal in course, calibre and excretion pattern.



[Table/Fig-1]: Contrast Enhanced Computed Tomography of kidneys ureter bladder showing renal pelvis Space Occupying Lesion (SOL).

A contrast enhanced Magnetic Resonance Imaging (MRI) revealed a dilated left renal pelvis having medium sized soft tissue intensity mass lesion being heterogeneously hyper-intense on T2, Short Tau Inversion Recovery (STIR), isointense on T1, diffusion restriction and mild heterogeneous contrast enhancement, measuring about 4.6×4.3×4.0 cm, leading to a radiological diagnosis of transitional cell carcinoma of renal pelvis with no enlarged loco regional lymph nodes [Table/Fig-2]. The other differential diagnoses were renal cell carcinoma with sarcomatoid features, leiomyosarcoma, lymphoma, malakoplakia, pyelonephritis and tuberculosis.

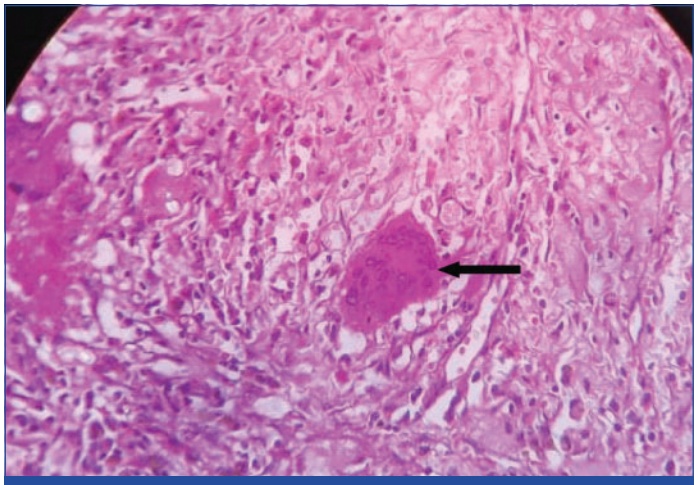


[Table/Fig-2]: Coronal section of MRI T1W image showing left renal Space Occupying Lesion (SOL).

With clinical features consistent of malignancy and radiological investigations suggesting the diagnosis of transitional cell carcinoma, the patient had undergone laparoscopic left radical nephro-ureterectomy with bladder cuff excision. During resection extensive adhesion was encountered.

The specimen grossly consisted of enlarged yellowish lobulated kidney with adherent perinephric fat measuring 15×18×5 cm. The ureter was 10 cm in length and 5 mm in diameter. On cut section of the kidney, multiple whitish cystic areas with solid intervening parts were seen with dilated pelvis and loss of cortico-medullary distinction. No calculus was seen in kidney or ureter. Microscopically

it revealed, a diffuse inflammatory infiltrate composed of foamy histiocytes, multinucleated giant cells, lymphocytes, plasma cells and polymorphonuclear leukocytes, confirming the diagnosis of diffuse variety of XGPN [Table/Fig-3].



[Table/Fig-3]: Photomicrograph showing sheets of foamy histiocytes admixed with lymphocytes, plasma cells and multinucleated giant cell (arrow) (H&E, 400x) (40X). Giant cell (←).

Postoperative period was uneventful. Patient was discharged on fourth postoperative day and resumed her normal activity within a week. She was asymptomatic on 6th month follow-up with normal renal biochemical parameters.

DISCUSSION

The XGPN is a less commonly reported chronic inflammatory disorder with 1.4 cases per 100,000 population [1]. Relevant case studies from last decade showing co-existence of XGPN with other renal pathology as well as conditions mimicking it are listed in [Table/Fig-4] [2-5]. It was first described by Schlagenhaufer in 1916. It accounts between 0.6-1% of all pyelonephritis cases [6]. It can affect all ages, but more likely middle-aged females and the elderly.

There are three forms of XGPN- diffuse, focal, and segmental [7]. The XGPN is characterised by renal parenchyma destruction by inflammatory cells, abscesses, and lipid laden macrophages and replacement by granulomatous tissue containing histiocytes and foamy cells which are centred around renal pelvis and calyces [8]. The exact pathology is still unknown. Nephrolithiasis, most commonly staghorn-type calculus, can be associated with XGPN [1]. Some of the predisposing factors are Urinary Tract Infection (UTI) especially with *Proteus mirabilis* and *Escherichia coli*, uretero-pelvic duplication, severe vesico-ureteral reflux, chronic interstitial nephritis, diabetes mellitus, rheumatoid arthritis, cirrhosis, obesity, metabolic syndrome, and immunocompromised state [2]. Altered lipid metabolism and transport, arterio-venous occlusions, lymphatic obstruction, haemorrhage, and necrosis of pericalyceal fat are other factors contributing to XGPN [9]. The condition is usually unilateral and results in a non functioning of kidney [10].

The literature describes how XGPN can mimic the features of several benign and malignant renal pathology including renal cell carcinoma and its variants [8]. The gross features of diffuse XGPN include enlargement of kidney with hydronephrosis, obstructive pelvic calculus, malignancy of ureter or congenital obstruction. Sometimes single or multiple yellow to orange nodules may be present mimicking tumour nodules. Other findings such as central necrosis with abscess formation, involvement of perinephric fat and diffuse cortical scarring with effacement of the normal renal architecture, and cortical atrophy may be seen. In severe cases, gross destruction of tissue occurs extending into the perinephric tissues and adrenal glands [11].

Histopathology is pathognomonic with diffuse inflammatory cell infiltrate. There is an admixture of lipid laden foamy macrophages, neutrophils, lymphocytes, plasma cells, and giant cells which was also evident in present case [9]. In addition to above cholesterol crystals, renal tubular atrophy, fibrosis, dilatation of tubules, micro abscesses, focal squamous metaplasia of the urothelium, lymphoid aggregates with germinal centre formation, and spindle cell proliferation can be observed [12].

Clinical symptoms of XGPN include fever, flank pain, palpable mass, malaise, anorexia, and weight loss [9]. Dysuria, frequency, pyuria, or haematuria may be experienced. Abscess formation (paranephric and psoas), fistula formation (reno cutaneous and reno colonic), and sepsis are known complications of XGPN [13]. The XGPN is a diagnostic dilemma preoperatively as the clinical and radiologic findings imitate both benign and malignant lesions [12]. In the index case, the XGPN simulated transitional carcinoma of renal pelvis.

The differential diagnosis of XGPN include renal cell carcinoma with sarcomatoid features, leiomyosarcoma, Wilm's tumour, lymphoma, malakoplakia, megalocytic interstitial nephritis, pyelonephritis, tuberculosis, and perinephric abscess [14]. Renal cell carcinoma, transitional cell carcinoma of the renal pelvis, and squamous cell carcinoma simulating XGPN has also been seen [1,2,15]. But XGPN mimicking as transitional cell carcinoma of renal pelvis has rarely been cited in literature. One such study performed by Ordones FV et al., showed how a patient presented with all clinical and radiological features of XGPN underwent nephrectomy and histopathological examination revealed transitional cell carcinoma of renal pelvis [1].

The benefits of laparoscopic management for XGPN as compared to open surgical technique have been described in several case series. It includes shorter hospital stays, fewer hospital re-admission and more rapid return to work which was also evident in the present case. But even among the experienced surgeons about 30% conversion to open surgery from laparoscopy has been reported due to technical difficulty and failure to progress [16,17].

Fallatah A et al., reported co-existing renal cell carcinoma with XGPN and transitional cell carcionoma with XGPN [18]. According to Shah HN et al., XGPN can mimic tuberculosis [19]. As per Rahaman MK et al., renal cell carcinoma can mimic XGPN [8]. All these point towards the fact that the overlapping clinical and radiological features of

Study	Clinical features	Radiographic finding	Management	Diagnosis	Prognosis
Ganpule A et al., 2013 [2]	Flank pain, fever, vomiting	CECT scan shows enhancing renal mass with renal vein thrombosis and paracaval lymph nodes.	Laparoscopic right nephrectomy	XGPN	Discharged on day 4. No follow-up.
Kanodia KV et al., 2015 [3]	Flank pain, tenderness right flank	Gross hydronephrosis, parenchymal thinning, irregular thick enhancing wall of pelvicalyceal system favouring malignancy no excretion for four hours.	Right nephrectomy	XGPN and squamous cell carcinoma	Discharged. No follow-up.
EL Abiad Y et al., 2016 [4]	Right flank pain, unintentional weight loss	CECT shows upper polar heterogenous renal mass.	Open right radical nephrectomy	XGPN	Discharged on day 3. Two month follow-up shows no recurrence or complications.
Chang T and Tseng J, 2021 [5]	Right flank pain, right lower limb swelling	Deformed right kidney, heterogeneously enhanced soft tissue, staghorn in renal pelvis, hydronephrosis, mild hydroureter, perirenal abscess.	CT guided abscess drainage followed by right nephrectomy	XGPN with squamous cell carcinoma	Uncomplicated till four month follow-up.

[Table/Fig-4]: Relevant case studies from last decade showing co-existence of xanthgranulomatous pyelonephritis with other renal pathology and conditions mimicking it [2-5].
CECT: Contrast enhanced computed tomography; XGPN: Xanthgranulomatous pyelonephritis

XGPN with several other renal pathology makes it a challenging diagnosis for the clinicians.

CONCLUSION(S)

The XGPN is one of the less commonly reported variant of chronic pyelonephritis usually associated with renal stones with progressive loss of renal parenchyma, resulting in non functional kidney. CT can aid in the diagnosis but radiological findings often mimic benign or malignant lesions, thus posing a diagnostic challenge.

REFERENCES

[1] Ordones FV, Das K, Prowse S, Cohen P, Brook NR. High-grade transitional cell carcinoma masquerading as a xanthogranulomatous pyelonephritis and perinephric abscess. Radiol Case Rep. 2017;12:281-84.
[2] Ganpule A, Jagtap J, Ganpule S, Bhattu A, Soni S, Sabnis R, et al. Xanthogranulomatous Pyelonephritis (XGPN) mimicking a "renal cell carcinoma with renal vein thrombus and paracaval lymphadenopathy". F1000Research. 2013;2:263.
[3] Kanodia KV, Vanikar AV, Patel RD, Nigam LK, Trivedi HL. Rare co-existence of squamous cell carcinoma with infiltration of renal vein and xanthogranulomatous pyelonephritis. J Clin Diagn Res. 2015;9(12):ED15-16.
[4] EL Abiad Y, Dehayni Y, Qarro A, Balla B, Ammani A, Alami M. Xantogranulomatous pyelonephritis: The missed diagnosis. Int J Sur Case Rep. 2016;18:21-23.
[5] Chang T, Tseng J. Rare squamous cell carcinoma of the kidney with concurrent xanthogranulomatous pyelonephritis: A case report and review of the literature. Open Medicine. 2021;16(1):128-33.
[6] Siddappa S, Ramprasad K, Muddegowda MK. Xanthogranulomatous pyelonephritis: A retrospective review of 16 cases. Korean J Urol. 2011;52:421-24.
[7] Tsai KH, Lai MY, Shen SH, Yang AH, Su NW, Ng YY, et al. Bilateral xanthogranulomatous pyelonephritis. J Chin Med Assoc. 2008;71:310-14.
[8] Rahaman MK, Rana S, Jairajpuri ZS, Khetrapal S. Renal cell carcinoma and xanthogranulomatous pyelonephritis: A diagnostic challenge. Arch Med Health Sci. 2018;6;247-50.
[9] Kundu R, Baliyan A, Dhingra H, Bhalla V, Punia RS. Clinicopathological spectrum of xanthogranulomatous pyelonephritis. Indian J Nephrol. 2019;29(2):111-15.
[10] Das DP, Pal DK. Co-existing malakoplakia and xanthogranulomatous pyelonephritis of kidney: Two different spectrum of same disease process. Urol Ann. 2016;8:252-54.
[11] Li L, Parwani AV. Xanthogranulomatous pyelonephritis. Arch Pathol Lab Med. 2011;135(5):671-74.
[12] AlDarrab RM, AlAkrash HS, AlKhateeb SS, AlBqami NM. A case report of a xanthogranulomatous pyelonephritis case mimicking the recurrence of renal cell carcinoma after partial nephrectomy. Urol Ann. 2015;7:524-26.
[13] Puthenveetil RT, Baishya D, Barua S, Sarma D. Unusual case of nephrocutaneous fistula-Our experience. Asian J Urol. 2016;3:56-58.
[14] Dwivedi US, Goyal NK, Saxena V, Acharya RL, Trivedi S, Singh PB, et al. Xanthogranulomatous pyelonephritis: Our experience with review of published reports. ANZ J Surg. 2006;76:1007-09.
[15] Singh U, Jena R, Sureka S. Upper tract transitional cell carcinoma clinically mimicking inflammatory renal pathology: A report of three cases. Indian Journal of Urology. 2021;37(2):169.
[16] Kapoor R, Vijjan V, Singh K, Goyal R, Mandhani A, Dubey D, et al. Is laparoscopic nephrectomy the preferred approach in xanthogranulomatous pyelonephritis? Urology. 2006;68:952-55.
[17] Duarte RJ, Mitre AI, Chambô JL, Arap MA, Srougi M. Laparoscopic nephrectomy outside gerota fascia for management of inflammatory kidney. J Endourol. 2008;22:681-86.
[18] Fallatah A, Tarakji M, Amuesi J. Xanthogranulomatous pyelonephritis: A retrospective study of 10 cases and review of the literature. Saudi J Kidney Dis Transpl. 2001;12:520-24.
[19] Shah HN, Jain P, Chibber PJ. Renal tuberculosis simulating xanthogranulomatous pyelonephritis with contagious hepatic involvement. Int J Urol. 2006;13:67-68.

PARTICULARS OF CONTRIBUTORS:
1. Associate Professor, Department of Urology, IPGMER and SSKM Hospital, Kolkata, West Bengal, India.
2. Post Doctoral Trainee, Department of Urology, IPGMER and SSKM Hospital, Kolkata, West Bengal, India.
3. Professor and Head, Department of Urology, IPGMER and SSKM Hospital, Kolkata, West Bengal, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Dilip Kumar Pal,
IPGMER and SSKM Hospital, Kolkata-700072, West Bengal, India.
E-mail: urologyipgmer@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Jun 03, 2021
- Manual Googling: Oct 03, 2021
- iThenticate Software: Oct 23, 2021 (20%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **May 27, 2021**
Date of Peer Review: **Jul 20, 2021**
Date of Acceptance: **Oct 06, 2021**
Date of Publishing: **Nov 01, 2021**