# Focal Xanthogranulomatous Pyelonephritis (a Pseudotumour) associated with Retroperitoneal Fibrosis: Case report

Shruti Thakur, Anupam Jhobta, R G Sood, Arun Chauhan, Charu S Thakur

## ABSTRACT

Xanthogranulomatous Pyelonephritis (XGP) is a rare form of renal granulomatous inflammatory disease. Two forms of XGP are well described; a diffuse form (85%) and a focal form (15%). Preoperative diagnosis of focal XGP is uncommon as it radiologically and clinically mimicks renal cell carcinoma. Therefore, the focal form is also referred to as the 'tumefactive form'. The definitive diagnosis of focal XGP is made only after histo-pathologic examination of the resected specimen in which characteristic lipid laden macrophages are seen. We present an interesting case of a 65-year-old male who had left lumbar pain. A preoperative diagnosis of renal cell carcinoma was made on CT. On histologic analysis, a final diagnosis of focal XGP was revealed. Although rare, focal XGP should be considered as the main differential for renal malignancy.

**Keywords-** nephrectomy, retroperitoneal fibrosis, Xanthogranulomatous pyelonephritis

Corresponding author mail: tshruti878@yahoo.in Conflict of interest: none

## INTRODUCTION

Xanthogranulomatous Pyelonephritis (XGP) is an atypical form of chronic suppurative renal inflammation. The inflammatory process is usually diffuse and can extend outside the kidney. It has pathognomonic diagnostic imaging features. However, the rare focal form remains a diagnostic dilemma and simulates renal neoplasm. No definite radiologic or clinical features exist for focal XGP. In appropriate clinical context, keeping focal XGP in mind, the need for radical nephrectomy may be obviated.

## CASE REPORT

A 65-year-old male presented with left flank pain and nocturia since 10 days. He had no history of fever, anorexia, weight loss, hematuria or dysuria. The blood tests revealed no anaemia or leucocytosis (Hb-

13.7g/dl, Total WBC Count-7.29 X 10^9/l). The liver function tests (SGOT/AST-12U/L, SGPT/ALT- 36U/L) and renal function tests (blood urea- 47.9mg/dl, serum creatinine-1.3mg/dl) were normal. Urine microscopic examination and culture were normal. PSA was 0.56 ng/l and X- ray KUB was normal. He was a hypertensive and diabetic for the past 15 years. He had a history of right DJ stenting for calculus disease. Abdominal sonography showed a hypoechoic mass in left kidney. CT was done for definitive assessment. On plain CT, a well defined solid mass was seen in the midpolar region of left kidney. No calculus or hemorrhage was seen. There was fat stranding along its anterolateral border (Figure 1).

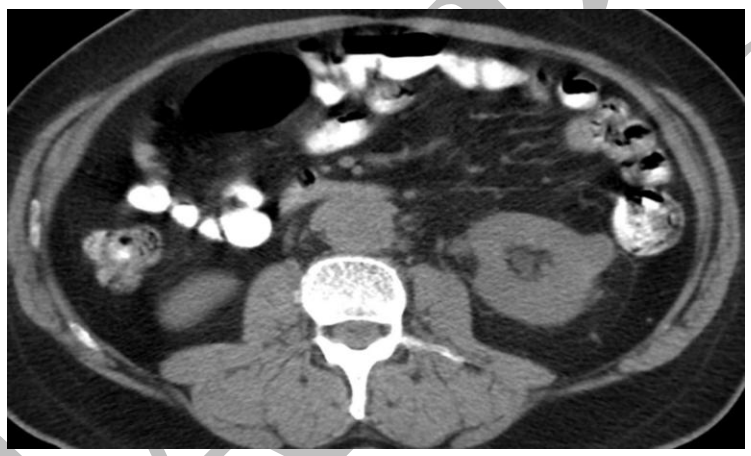

**Figure 1** Plain CT axial image shows a mass in midpolar region of the left kidney. Slight fat stranding is seen along its anterolateral aspect. No haemorrhage or calculus is seen in relation to this mass.

On CECT, the mass measured 2.8 x 2.5 x 2 cm which on corticomedullary phase showed heterogeneous enhancement (Figure 2).

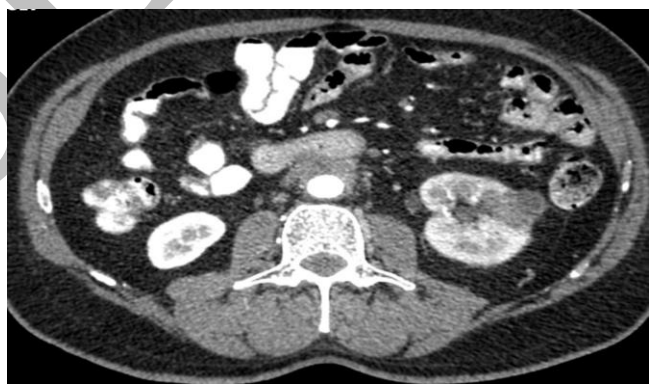

**Figure 2** Corticomedullary phase axial image shows heterogeneous enhancement of the mass. Adjacent fat shows stranding indicating the extension of the pathological process into the pararenal fat.

The enhancement persisted in nephrographic phase (Figure 3).

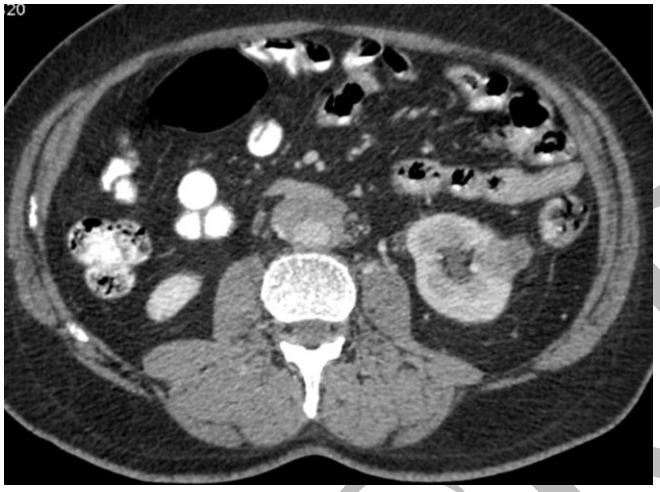

**Figure 3** Nephrographic phase coronal image shows persistence of enhancement of the left renal mass which is predominantly eccentric. Retroperitoneal fibrosis around the aorta is also seen.

There was no hydronephrosis. Imaging findings and clinical features suggested renal cell carcinoma as the diagnosis. The right kidney was normal. Also, there was plaque like area of soft tissue along the anterior and lateral aspect of infra-renal aorta. This soft tissue was isodense to the surrounding muscle on plain CT (Figure1), showed no nodular outline and little enhancement was seen on contrast administration suggestive of chronic retroperitoneal fibrosis (Figure 2). The patient further underwent CT-SPECT for the possible skeletal metastasis. An area of relative increased tracer uptake was seen in the left kidney. Abnormal focal increased tracer uptake was also seen in the mid cervical spine region (Figure 4).

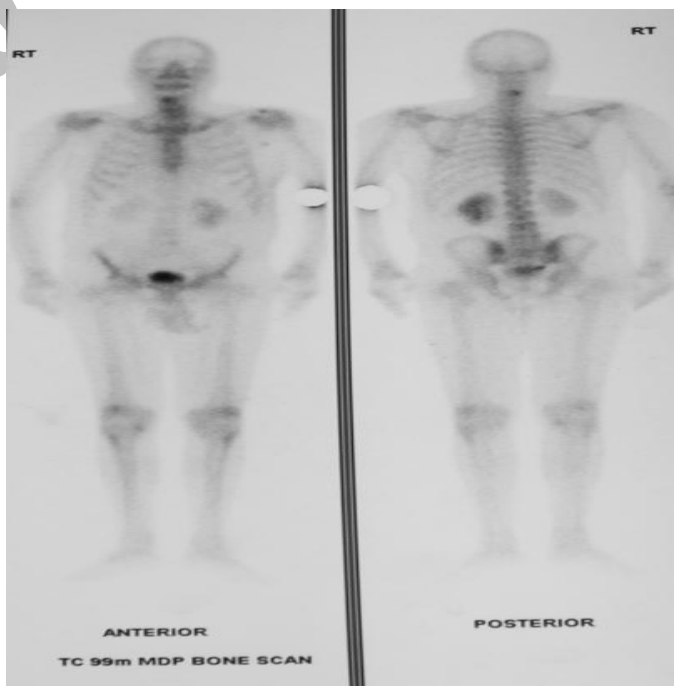


**Figure 4** SPECT –CT shows relative increased uptake of the tracer in the left kidney and in mid-cervical region.

It was localized to the osteophyte in C6 vertebral body on right side and favoured age related degenerative changes. There was no evidence of skeletal metastasis. The patient was then taken up for partial nephrectomy. The histopathologic examination of the surgical specimen showed lipid laden macrophages revealing focal XGP as the definite diagnosis. In the absence of suspicion of focal XGP as the preoperative diagnosis, the patient escaped radical surgery only because of the localized form of the mass.

**DISCUSSION**

Xanthogranulomatous Pyelonephritis is an uncommon variant of chronic renal granulomatous inflammation characterized by extensive inflammation and destruction of renal parenchyma and its replacement by lipid laden macrophages known as the foam or xanthoma cells. It is usually unilateral but bilateral cases have also been reported [1]. There is female preponderance (3:1) with presenting age group of 50 to 60 years [2]. The exact etiology is unknown but is believed to result from atypical, incomplete immune response to subacute bacterial infection. Diabetes mellitus is a risk factor and seen in 10% of the patients [3]. The patient usually presents with flank pain, fever, fatigue, anorexia, weight loss, dysuria and a tender flank mass[4]. The laboratory findings include anemia, leucocytosis, and increased ESR. Urine culture shows E. coli, P. mirabilis, Staph. aureus, Kleibsella and Pseudomonas as the causative agents [5].

Two forms of XGP are well known- a diffuse form which comprises 85% of total cases, is associated with diminution of renal functions and is seen most frequently in adults. Second is the focal form which is seen in 15% and is more common in children and women [6].

Abdominal sonography is the first imaging modality. Diffuse XGP shows generalized renal enlargement with loss of normal renal architecture. There is pelvicalyceal dilatation along with parenchymal destruction seen as hypoechoic areas and renal calculi or staghorn calculus seen as hyperechoic foci with clean posterior acoustic shadowing. However, focal XGP shows no features which are specific to diffuse XGP. It is seen as non-specific mass which may be hyperechoic, hypoechoic or isoechoic to renal cortex [5].

CT is the imaging modality of choice for confident diagnosis and for extrarenal extension of the disease. Diffuse XGP may be seen as non functioning enlarged kidney, thick enhancing septae in the hypodense area in renal parenchyma, central calculus with contracted pelvis, perinephric fat stranding and thickening of Gerota's fascia. Staghorn calculi are seen in 80% of the cases [7]. Dilated calyces and abscess cavities with pus and debris in diffuse form may be described as 'bear paw sign'. The disease progression is characterized by psoas abscess and colonic or cutaneous fistula formation. In atypical form there is renal atrophy rather than enlargement [3]. Focal XGP is usually depicted as well defined or ill defined localized mass with soft tissue or fluid attenuation and shows heterogeneous enhancement [5].

Adjacent structure or organ may be involved in this inflammatory process which is confused with renal tumour infiltrating into these structures. So, focal XGP is often erroneously interpreted preoperatively as renal neoplasm.

On SPECT-CT, this lesion shows comparatively increased tracer uptake as seen in our case. Frank increase in uptake is not seen as the inflammatory process in XGP is low grade.

Our case had retroperitoneal fibrosis in addition to focal XGP which may be purely co-incidental or may point towards common etiology. Both of these conditions are result of chronic inflammation as a response to a variety of stimulating factors. Retroperitoneal fibrosis is a rare disorder with a reported incidence of 1 per 200,000. It usually presents between 40-60 years of age and males are 2 to 3 times affected than females [8]. It is characterized by retroperitoneal chronic inflammation and marked fibrosis. If often entraps ureters and other abdominal organs. 2 forms are identified: two-thirds are idiopathic and rests are secondary to neoplasms, autoimmune diseases, infections, trauma, radiotherapy and certain medications like ergot derivatives, hydralazine, analgesics, and beta-blockers. The pathogenesis is unclear but two mechanisms have been proposed. One, it is an exaggerated local inflammatory reaction to aortic atherosclerosis and second, it is a manifestation of systemic autoimmune disease [9].

The definitive treatment of focal XGP is surgery although case reports with medical

treatment have also been described. Radical nephrectomy is reserved for diffuse XGP. In focal form, more conservative approach like partial nephrectomy is preferred as the relapse in the affected kidney is unusual.

The constellations of features which may be helpful in preoperative diagnosis of diffuse XGP are its unilaterality, deranged renal function tests, clinical history and laboratory findings. However, focal XGP has no such distinguishing features. Very often it is diagnosed and treated as renal neoplasm. Percutaneous needle aspiration is helpful in diagnosis only if it retrieves foam cells. In the absence of urinary tract infection or obstruction and with no signs of inflammation (as was in our case), the diagnosis completely rests on pathological analysis of resected surgical specimen [10]. Awareness of the possibility of this condition helps is early recognition and possible treatment.

## CONCLUSION

Although surgery is considered the only effective treatment of focal XGP, a high index of suspicion of XGP prevents the radical nephrectomy for this pseudotumoral lesion.

## REFERENCES

1. K. Ramboer, R. Oyen, S. Verellen, S. Vermeersch, A. L. Baert and R. Verberckmoes. Focal xanthogranulomatous pyelonephritis mimicking a renal tumor: CT- and MR-findings and evolution under therapy.Nephrol Dial Transplant 1997; 12: 1028–1030
2. R. Loffroy, B. Guiu, J. Watfa, F. Michel, J.P. Cercueil, D. Krause. Xanthogranulomatous pyelonephritis in adults: clinical and radiological findings in diffuse and focal forms. Clinical Radiology 2007; 62: 884-890
3. William D. Craig, Brent J. Wagner, Mark D.Travis. Pyelonephritis: Radiologic-Pathologic Review RadioGraphics 2008; 28:255–276
4. Stanford M. Goldman, David S. Hartman, Elliot K. Fishman, Joseph P. Finizio, Olga M.B. Gatewood, Stanley S. Siegelman. CT of Xanthogranulomatous Pyelonephritis: Radiologic-Pathologic Correlation. AJR 1984; 141: 963-969

5. Jongchul Kim. Ultrasonographic Features of Focal Xanthogranulomatous Pyelonephritis J Ultrasound Med 2004; 23: 409–416
6. Amit Suri,Vivek Gondotra, Jyotsana Suri. Xanthogranulomatous Pyelonephritis. JKScience; 2010;12:44-45
7. G.Verswijvel, R.Oyen, H.Van Poppel, T.Roskams. Xanthogranulomatous pyelonephritis: MRI findings in the diffuse and the focal type. Eur. Radiol.2000; 10: 586-589
8. Carmel G. Cronin, Derek G. Lohan, Michael A. Blake, Clare Roche, Peter McCarthy and Joseph M. Murphy.Retroperitoneal Fibrosis: A Review of Clinical Features and Imaging Findings. American Journal of Roentgenology. 2008;191:423-431
9. Amaka Ezimora, Marquetta L. Faulkner, Oluwafisayo Adebiyi, Abimbola Ogungbemile, Salas-Vega Marianna, and Chike Nzerue. Retroperitoneal Fibrosis: A Rare Cause of Acute Renal Failure. Case Reports in Nephrology.Volume 2012 (2012), Article ID 645407, 5 pages doi:10.1155/2012/645407
10. Shinde S, Kandpal D K, Chowdhary S K. Focal xanthogranulomatous pyelonephritis presenting as renal tumor. Indian J Nephrol 2013; 23:76-7.