CASE REPORT



Xanthogranulomatous pyelonephritis (XGPN) mimicking a "renal cell carcinoma with renal vein thrombus and paracaval lymphadenopathy" [v1; ref status: indexed, http://f1000r.es/2bv]

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Abstract

We present a case of Xanthogranulomatous pyelonephritis mimicking as a renal cell carcinoma. This was an elderly lady who presented with pyonephrosis due to urolithiasis. On evaluation she was found to have a space occupying mass in the right kidney. Further investigations revealed an enhancing tumor with renal vein thrombus and paracaval lymphadenopathy. Subsequent histopathology showed evidence of XGPN with no malignancy. This case report highlights the fact there are a number of imaging and clinical overlaps in the diagnosis, assessment and management of this entity.

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2	Stefanos Kachrilas , Bart's Health NHS Trust UK			
3	Daron Smith, University College London Hospitals NHS Trust UK			
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Case presentation

A 67 year old Hindu female presented to us in May 2010 with history of right flank pain, fever and vomiting. She had raised total leukocyte count: 16600/µL and deranged renal function (serum creatinine: 3.1mg/dL). A non-contrast CT (NCCT) scan revealed moderate hydronephrosis, right upper ureteric calculus and a well circumscribed lesion on the medial aspect of the kidney. A percutaneous nephrostomy was performed on account of the deranged renal function. Subsequently, the patient underwent a percutaneous nephrolithotomy (PCNL).

At one month from presentation and after the serum creatinine improved to 1.47mg/dL, a contrast CT revealed an enhancing mass (enhancement from 33 to 118 Hounsfield units) on the medial aspect of the kidney (Figure 1; a contrast CT not done at initial presentation due to deranged renal function) with evidence of renal vein thrombosis and multiple paracaval lymph nodes. A provisional diagnosis of renal cell carcinoma with

renal vein thrombus was made. The clinical stage was T3aN2M0. A laparoscopic radical nephrectomy was done. The gross specimen revealed evidence of renal vein thrombus and Xanthogranulomatous pyelonephritis (XGPN) (Figure 2). On H & E (Hematoxylin & Eosin) microscopic examination, it was composed of foamy macrophages admixed with inflammatory infiltrate (Figure 3). There was no evidence of malignancy. The patient recovered well and was discharged in stable condition after 4 days with a serum creatinine of 1.16mg/dL.

Discussion

XGPN is an uncommon, severe, chronic suppurative renal parenchymal infection characteristically leading to renal destruction. The majority of cases are unilateral and result in a nonfunctioning, massively enlarged kidney associated with obstructive uropathy secondary to urolithiasis. XGPN has been described as a great imitator or a masquerading tumor in adults and pediatric age groups^{1,2}. The etiological factor in this case was the renal calculus with chronic infection.

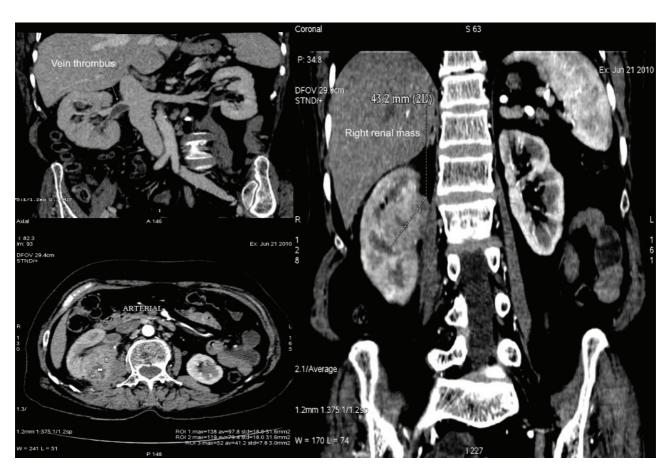


Figure 1. Well defined soft tissue density mass of right kidney measuring $49 \times 35 \times 43$ mm enhancing from 33 HU to 118 HU with non-enhancing areas of necrosis.



Figure 2. Gross specimen showing thrombus in renal vein.

The imaging findings in this case showed a significantly enhancing mass, lymph nodes and a renal vein thrombus. The mass was seen closely abutting the psoas as well. The CT findings mimicked a case of T3N2Mx renal cell carcinoma. Localised XGPN is amenable to partial nephrectomy if diagnosed preoperatively. XGPN has been found to be associated with renal cell carcinoma, papillary transitional cell carcinoma and squamous cell carcinoma and hence nephrectomy should be performed when malignancy cannot be excluded. This case highlights the need to keep XGPN as a differential diagnosis of a renal mass especially in presence of urolithiasis.

Consent

Written informed consent for publication of clinical details and clinical images was obtained from the patient.

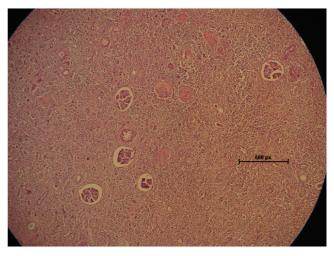


Figure 3. Microscopic examination at 100X magnification showing collection of foamy macrophages and inflammatory infiltrate diffusely infiltrating the renal parenchyma.

Author contributions

Arvind Ganpule and Jitendra Jagtap drafted the manuscript and carried out the literature search. Sanika Ganpule, Amit Bhattu and Shailesh Soni prepared the illustrations and helped to draft the manuscript. Ravindra Sabnis and Mahesh Desai revised the manuscript and did the final proofreading of the manuscript. All authors approved the final manuscript for publication.

Competing interests

No competing interests were disclosed.

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Version 1

Referee Report 04 September 2014

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Daron Smith

Institute of Urology, University College London Hospitals NHS Trust, London, UK

The authors present a case of XPN (Xanthogranulomatous Pyelonephritis) that was believed to be a renal cell carcinoma based on imaging. The propensity for XPN to "imitate" renal malignancy is well established. Indeed, I wrote a case report many years ago when, in addition to the imaging apparently suggesting a renal cell carcinoma, there was an incidental small RCC in the same kidney! (Smith RD *et al.*, 2000) It would be worth emphasizing what is unique/important about this case, and the learning message that follows. Is it the apparent vascular invasion with thrombus in the vein that the authors wish to highlight?

I have a few other suggestions. In the abstract, "tumour" should be changed to "mass". The patient did not have a renal tumour in the sense of a cancer as this word is often used. "Tumour", in its most frequently used sense of malignancy, may cause confusion to anyone reading the abstract, believing this was an renal cancer with tumour thrombus.

I would prefer to see the creatinine expressed in SI units (umol/L) and an eGFR given as well as the units in mg/dL.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Competing Interests: No competing interests were disclosed.

Referee Report 25 February 2014

doi:10.5256/f1000research.3019.r3601



Stefanos Kachrilas

Urology Department, Bart's Health NHS Trust, London, UK

 The topic of this case review is not especially novel, and the information provided is unlikely to be useful to other practitioners, as the clinical entity of XGPN is well documented in the existing literature.



- The background, history, presentation, physical examination, and diagnostic tests are appropriately presented.
- The authors have not commented on the post-operative management of the patient.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.

Competing Interests: No competing interests were disclosed.

Referee Report 15 January 2014

doi:10.5256/f1000research.3019.r3093



M Hammad Ather

Section of Urology, Aga khan University, Karachi, Pakistan

The authors present an uncommon clinical situation where XGPN mimicked a renal tumor. XGPN is indeed a rare type of renal infection characterised by granulomatous inflammation with giant cells and foamy histiocytes. It has been shown in many case reports and small case series to mimic an infiltrative malignancy. Renal vein thrombus has also been described in many previous reports.

I have few other observations that the authors may like to address. What were the findings on nephroscopy, and did the surgeon take any biopsies of the suspicious lesion? Did you noticed any xanthoma cells in the urine? The lady has a classical presentation of an XGPN (gender, obstructing middle age ... was she diabetic or immunocompromised in any way?) stone until the contrast study. Did the investigators consider a biopsy prior to planning a nephrectomy for a fair functioning kidney?

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Competing Interests: No competing interests were disclosed.

Author Response 20 Jan 2014

Jitendra Jagtap, Muljibhai Patel Urological Hospital, India

Thank you for your review Dr. M H Ather. Please find below the response to the comments:

- On nephroscopy during PCNL there were no suspicious lesions noted within the
 pelvicalyceal system so the surgeon did not take any biopsies. Absence of suspicious
 findings was double checked by reviewing the intraoperative video of this patient.
- Urine examination did not reveal the presence of any xanthoma cells.
- No, the lady was neither diabetic or immunocompromised.
- Biopsy was not considered, as it would not have altered the further management as the mass had features suggestive of malignancy on radiology namely: significant enhancement



from 33 to 118 Hounsfield units, presence of renal vein thrombus, and multiple enlarged lymph nodes obviating the possibility of a nephron sparing procedure. The current roles of renal biopsy as outlined in various guidelines include: confirmation of diagnosis of radiologically indeterminate renal masses; obtaining histology of incidentally detected renal masses in patients who are candidates for nonsurgical treatment (active surveillance, ablative therapies); and selection of the most appropriate targeted therapy for metastatic renal tumours depending upon the histology (Ljungberg B *et al.*, 2013; Novick A *et al.*, 2010; Herts BR & Baker ME, 1995; Campbell SC *et al.*, 1997; Volpe A *et al.*, 2007). Also XGPN has been found to be associated with renal cell carcinoma, papillary transitional cell carcinoma and squamous cell carcinoma, and hence nephrectomy should be performed when malignancy cannot be excluded (Tolia BM *et al.*, 1981; Schoborg TW *et al.*, 1980).

Competing Interests: No competing interests were disclosed.