An Unusual Presentation of Xanthogranulomatous Pyelonephritis and its Management by Nephron-Sparing Surgery

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Citation

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Abstract

Xanthogranulomatous pyelonephritis has been coined as “the great imitator” as it is associated with a wide range of atypical presentations. We report yet another atypical presentation as a focal renal neoplasm in a young patient. The definitive diagnosis was derived only after histopathological examination.

INTRODUCTION

Xanthogranulomatous pyelonephritis (XGPN) is a rare form of chronic suppurative atypical renal parenchymal infection, usually caused by calculous obstructive uropathy. [1] Patients are often immunocompromised and have associated urinary infection or urolithiasis. [2] We report a case of a young male who presented with a right renal neoplasm. Provisional diagnosis was renal cell carcinoma. Histopathologic examination concluded the diagnosis of XGPN.

CASE REPORT

A 19-year-old male presented to our outpatient clinic with complaints of dull aching pain in the right flank for 2 months and occasional low-grade fever. There was no history of hematuria, lithuria, pyuria or weight loss. There was no past history of diabetes or tuberculosis. On examination, he was afebrile, his abdomen was soft and there was no palpable lump. Hemogram, blood glucose and renal function tests were normal. Urinalysis and urine culture was normal. USG revealed a heterogeneous mass in the right kidney. CT scan (Fig.1) revealed a well circumscribed heterogeneous mass of 4.0 3.0 3.4cm involving the mid-polar region of the right kidney. The remaining part of the kidney demonstrated good function. Left kidney, both ureters and urinary bladder were normal. There was no lymphadenopathy. FNAC done at a primary care centre prior to referral was reported inconclusive. A presumptive diagnosis of renal neoplasm was made. In view of the well circumscribed lesion and the preserved function of the remaining right kidney, a right upper partial nephrectomy (Fig. 2) was performed. Histopathology was reported as xanthogranulomatous pyelonephritis (Fig. 3). The patient is followed up regularly with good function of the remaining right kidney (Fig. 4).

DISCUSSION

Xanthogranulomatous pyelonephritis (XGPN) is a debilitating chronic inflammatory disorder of the kidney characterised by an infectious phlegmon arising in the renal parenchyma. [2] It is rare, representing 0.6% of all renal infections. [1] Urinary tract anomalies, chronic infections, renal ischemia, immunodeficiency and abnormal lipid metabolism contribute to its pathogenesis. Affected patients are usually in the sixth and seventh decades and it is commoner in females and diabetics. [2] Unilateral diffuse involvement is more common than focal variants. [3] The focal variant is more common in females and children. Our patient, in contrast, was a healthy young male in the second decade. He had no comorbidities. He had no evidence of calculous disease or chronic obstructive uropathy. Clinically and radiologically, XGPN often mimicks renal cell carcinoma, abscess or chronic inflammatory diseases. The clinical profile in our case was quite non-specific. The preoperative diagnosis of XGPN is challenging. CT may reliably diagnose XGPN and also reflect on renal function. Characteristic CT features include poorly enhancing mass, thickened Gerota's fascia and the classic “bear paw sign”. CT scan in our case suggested a neoplastic etiology. The uninvolved part of the right kidney showed good function in contrast to the reported association of non-functional kidneys with this condition. [1] This contributed to the

diagnostic dilemma in our case and we planned for partial nephrectomy. Histopathology demonstrated sheets of lipid-laden histiocytes with surrounding inflammatory cells and necrotic areas and concluded the diagnosis of XGPN. Despite improvements in imaging, the diagnosis of XGPN is arrived at in most cases after histopathological examination of the resected specimen. [3]

XGPN has been associated with a wide range of atypical presentations. Focal XGPN is reportedly rare, usually present in fourth and fifth decades, in females with suppressed immunity and is associated with a positive urine culture in 60% of cases. [4] Our patient was an adult male of second decade, was fully immunocompetent and had no evidence of chronic infection. Imaging studies were in favour of a renal neoplasm and final diagnosis was only by histopathological examination.

In a young male with inconclusive clinical features and imaging suggestive of focal renal neoplasm, XGPN should be kept in mind. Partial nephrectomy is a good treatment option in these cases with no long term morbidity.

Figure 1

Figure 1: CT image

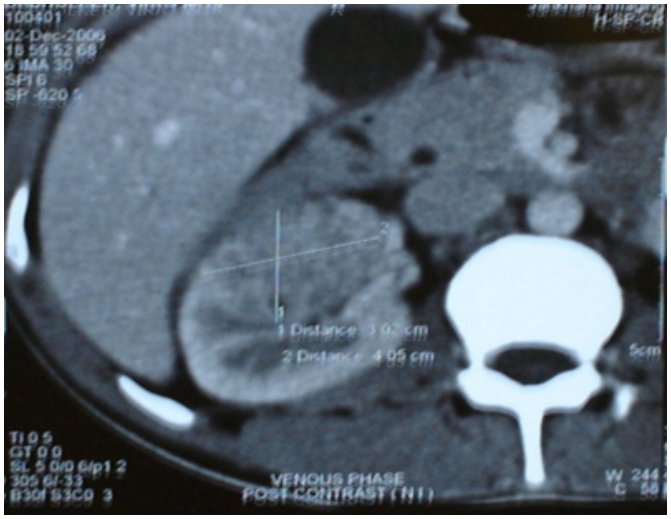


Figure 2

Figure 2: Intraop image

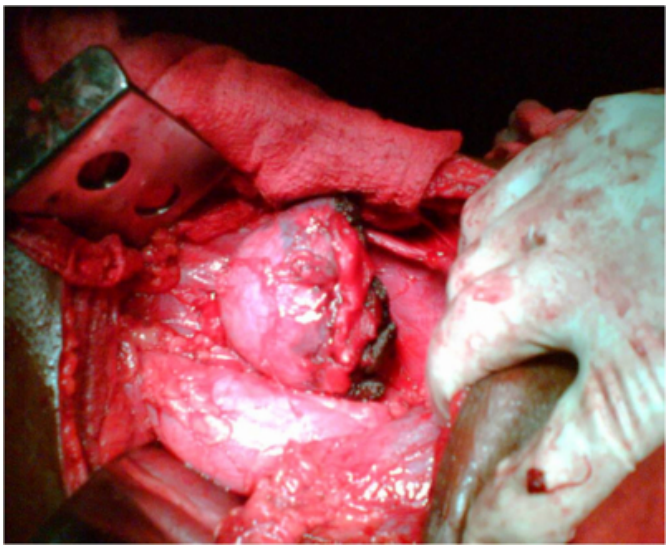


Figure 3

Figure 3: Histopathology Slide

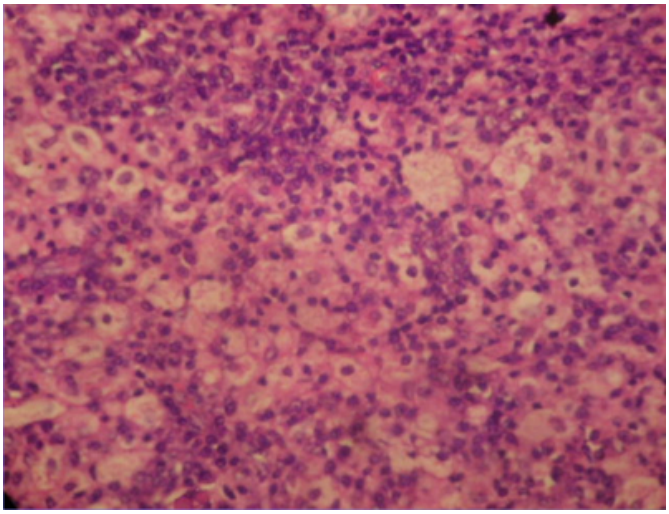
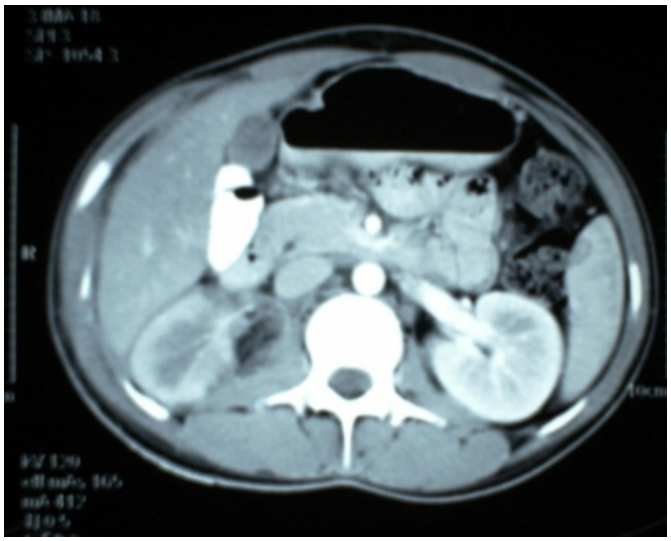


Figure 4

Figure 4: Follow-up CT Scan



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