Bilateral xanthogranulomatous pyelonephritis: Morphologically rotund, functionally lame!

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A 54-year-old diabetic male presented with pain in both flanks, recurrent fever and malaise for the past few months. Abdominal examination revealed tender vague masses in the both lumbar regions. Laboratory investigations showed hemoglobin of 7.7 g/dl and serum creatinine 2.8 mg/dl. Urinalysis demonstrated pus cells and *Escherichia coli* on culture. Ultrasound abdomen revealed bilateral enlarged hydronephrotic kidneys with papery-thin renal parenchyma. Noncontrast computed tomography (CT) revealed bilateral large staghorn calculi causing obstructive nephromegaly and hydronephrosis with low attenuation contents within the pelvicalyceal system. There was associated perinephric, juxtahilar and parapelvic fat proliferation causing bulging of bilateral renal fasciae, suggestive of bilateral diffuse xanthogranulomatous pyelonephritis (XGP) [Figure 1].

First described by Schlagenhaufer in 1916, XGP is a rare, serious, chronic inflammatory disorder characterized by destruction and replacement of renal parenchyma by lipid-laden macrophages (foam cells).[1] It is most commonly associated with *Proteus* or *E. coli* infection often seen in the setting of nephrolithiasis, diabetes and/or immunocompromised status. XGP constitutes <1% of chronic pyelonephritis and is thrice as common in women as in men. The disease can be either diffuse (85%) or focal (15%) in distribution. Most cases are unilateral and the affected kidney is usually nonfunctional. Bilateral diffuse involvement is rare and carries a grave prognosis. The typical imaging feature includes staghorn calculi, large kidneys with poor or absent renal function. Demonstrable macroscopic fat is infrequent on CT. Stage I disease remains confined to renal parenchyma; while in stage II and III, there is contiguous extension of the inflammatory process into the perinephric fat and adjacent retroperitoneal structures, respectively. Timely diagnosis and prompt therapy play a crucial role in limiting the morbidity

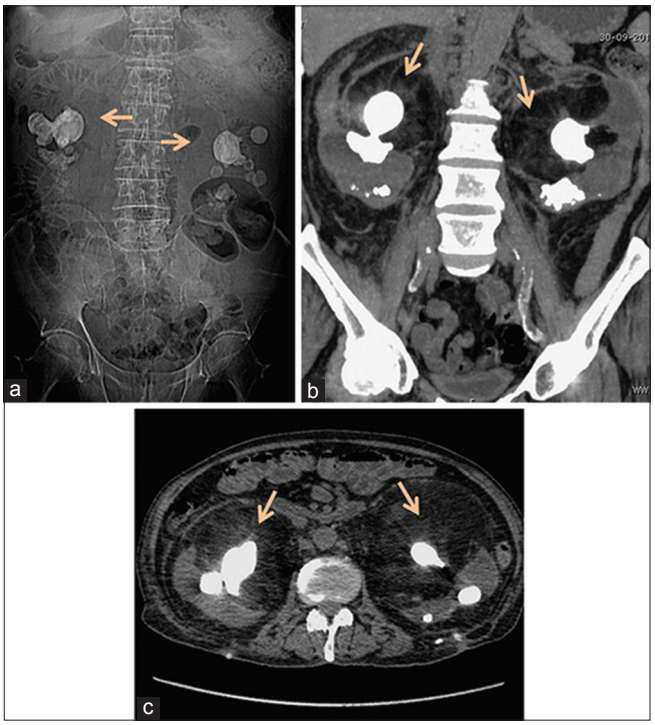

Figure 1: (a) Computed tomography (CT) scout image showing bilateral staghorn calculi (arrows) with surrounding halo of lucency (b and c) Coronal MIP and axial noncontrast CT images showing bilateral staghorn calculi (arrows) resulting in nephromegaly and hydronephrosis with marked fat proliferation and adjacent inflammatory stranding

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Access this article online	
Quick Response Code:	Website: www.indianjnephrol.org
	DOI: 10.4103/0971-4065.149960

and mortality. By the time XGP has become established, no conservative or medical therapies exist. Surgical nephrectomy is usually curative. The presence of inflammatory reaction in adjacent tissues often requires a large operative field and an anterolateral transperitoneal approach.[2]

Differential diagnoses are renal tuberculosis and renal abscesses in diffuse form; renal cell carcinoma and angiomyolipoma with minimal fat in focal form of XGP.

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How to cite this article: Bansal K, Sureka B, Jain V, Arora A. Bilateral xanthogranulomatous pyelonephritis: Morphologically rotund, functionally lame!. Indian J Nephrol 2015;25:255-6.

Source of Support: Nil, **Conflict of Interest:** None declared.