

Ureteral involvement in Xanthogranulomatous pyelonephritis - Rare manifestation

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Xanthogranulomatous pyelonephritis (XGPN) is an uncommon form of granulomatous inflammation characterized by destruction of the renal parenchyma and replacement by solid sheets of lipid-laden macrophages [1]. The process may be focal (10-17%) or diffuse (83-90%) with extension of inflammation outside the kidney. This entity is of particular interest to the radiologist, as the focal type may mimic renal cell carcinoma, while the diffuse form has imaging features which are characteristic [2]. XGPN is usually confined to the kidney, extension and involvement of ureter though reported, is rare [3]. We report one such case of XPGN having characteristic CT findings with involvement of the ureter.



Fig. 1:- Plain radiograph KUB, showing a large staghorn calculus in right renal collecting system.

CASE REPORT:

A 45 years old woman presented with the history of pain in the right loin. There was no history of dysurea or fever. The haemogram was unremarkable. Plain radiograph KUB revealed a large staghorn calculus in the right renal area (Fig 1). On USG, the right kidney was poorly visualized; instead multiple calculi surrounded by echogenic fat were seen. The urinary bladder showed a thin walled cyst of 1.2cm in size in relation to the right ureterovesical junction (Fig. 2). CT scan confirmed the staghorn calculus in the right kidney. The right renal parenchyma and the perinephric space were infiltrated by a fat density (-30 to -55 HU) mass which showed no enhancement on post contrast scans. Also, there was no excretion of contrast by the right pelvicalyceal system. The Gerota's fascia was thickened and the fat planes with the adjacent small bowel and the IVC were lost (Fig. 3 and 4). The wall of right ureter was thickened with fat density within its lumen (Fig. 5). The thickening extended till the right ureterovesical junction and terminated as a cystic mass projecting into the lumen of urinary bladder.

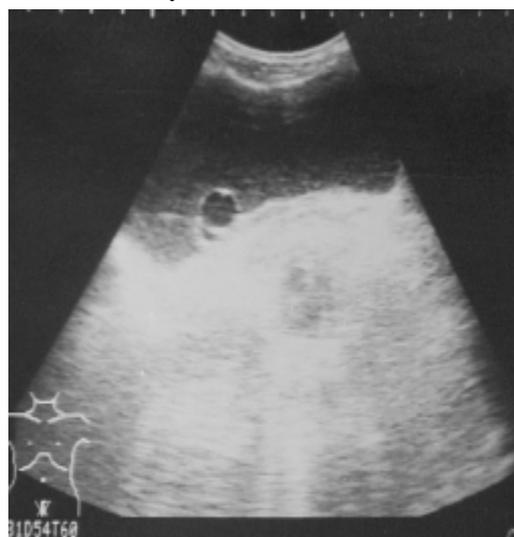


Fig. 2:- US showing a thin walled cyst in right uretero vesical junction.

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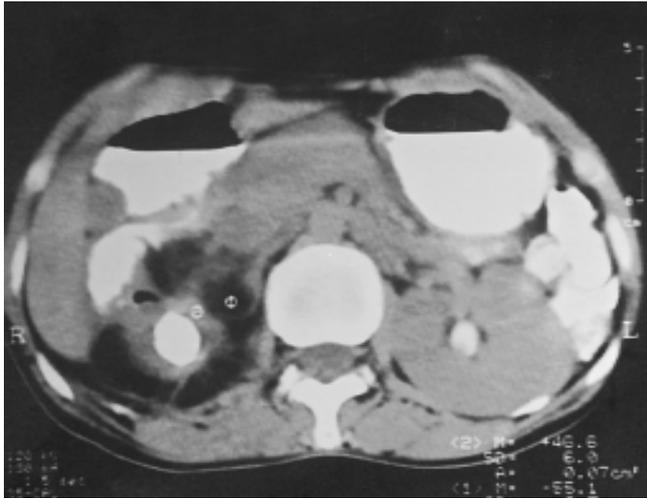


Fig. 3:- On plain CT, calculus seen in right collecting system, renal parenchyma is thinned and rest of it is replaced by a fat containing mass. Fat planes with adjacent bowel is lost.

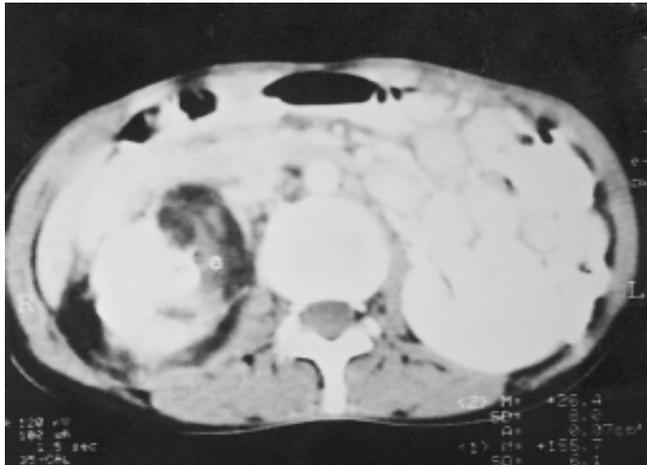


Fig. 4:- On contrast CT, there is no enhancement of renal parenchyma. The Gerota's fascia is thickened and the fat is adherent with the IVC.

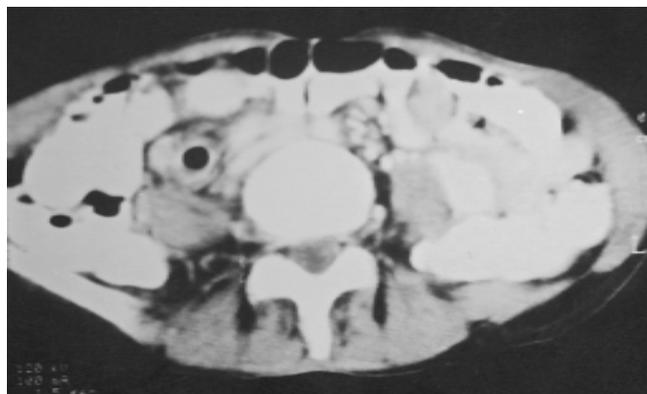


Fig. 5:- CT showing dilated thickened right ureter which is filled with fat.

Right nephrectomy with upper ureterectomy was done. A staghorn calculus filled the collecting system. The bulk of the mass was made up of fibroadipose tissue which was adherent to the duodenum, IVC and diaphragm. The ureter was also involved (Fig. 6). Histopathological examination revealed diffuse xanthogranulomatous pyelonephritis.



Fig. 6:- Gross specimen showing the yellowish fat in right kidney with involvement of the ureter.

DISCUSSION

XPGN is predominantly seen in middle aged women. Clinically the patient presents with recurrent fever, dysuria & flank pain that is unresponsive to antibiotics [4]. The disease is usually unilateral. It is the sequelae of severe, chronic obstructing parenchymal inflammation. The inflammatory process begins in the pelvis & produces gradual destruction of medulla & cortex by direct extension. In the diffuse form, the kidney is enlarged, the renal pelvis is usually dilated and contains staghorn calculus in 75% patients [1]. The surrounding parenchyma is converted to shaggy golden yellow tissue. Extension of inflammation into the perinephric space is common with invasion into the pararenal spaces, psoas muscle, small bowel, diaphragm, lung or soft tissues [5]. Involvement of ureter though reported, is rare [3, 4].

In imaging, CT is the modality of choice, though plain radiograph and US are the initial examinations [2]. Plain scout radiograph shows a staghorn calculus. On intravenous urography, an absent nephrogram or focally absent nephrogram is seen [5]. US typically shows an enlarged kidney with multiple anechoic or hypoechoic areas with a central staghorn calculus. The renal parenchyma is thinned and corticomedullary differentiation is lost. Sound transmission may be poor due to perinephric fat as was seen in our case [4].

CT scan demonstrates a large reniform mass with a central staghorn calculus. The renal parenchyma is replaced by multiple low attenuation (-15 to +15 HU) areas

representing dilated calyces and abscess cavities filled with pus and debris [6, 7]. Less common findings are a small contracted destroyed kidney with abundant perinephric fat (replacement lipomatosis) [1], as was the CT picture in our case.

XGPN has mainly to be differentiated from renal replacement lipomatosis. Both show a staghorn calculus with a non functioning kidney. But, on CT there is low attenuation material (+15 to -15 HU) filling the calyces in XGPN, whereas renal replacement lipomatosis shows attenuation of pure fat [7]. Other fat containing tumours such as angiomyolipom, lipoma or liposarcoma may be considered in differential diagnosis, but the absence of a staghorn calculus & a normally functioning kidney differentiates it from XGPN [7].

CT is also the modality of choice in depicting perinephric extension [1]. In our patient also, infiltration into small bowel & IVC was shown and confirmed on surgery. CT also depicted extension into ureter. Though CT features of ureteral involvement have not been reported but ureteral involvement has been confirmed on surgery [6, 7]. The dilated, thickened and fat containing ureter which was not well demonstrated on US, was clearly visualized on CT. CT further clarified that the cyst seen on US in relation to right ureterovesical junction was continuation of the involved ureter. CT features of ureter involvement have not been reported, by other authors but characteristic findings could be demonstrated in the present case which were confirmed on surgery and histopathology.

Some of these patients present with ureterocolic fistula. Also postoperative complications such as sinus and bowel fistula have been reported and these are usually due to failure to diagnose and properly stage the extent of disease [7]. Till date, CT shows the greatest promise in the preoperative identification and assessment of extent of XGPN especially involvement of ureters.

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