Reminder of important clinical lesson

Chronic flank pain, fever and an unusual diagnosis

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Summary

Xanthogranulomatous pyelonephritis (XGP) is a rare, serious, debilitating illness characterised by an infectious renal phlegmon. Most cases of XGP are unilateral and are often associated with urinary tract obstruction, infection, nephrolithiasis, diabetes, and/or immune compromise. This disease process ultimately results in focal or diffuse renal destruction and is characterised pathologically by lipid-laden foamy macrophages. XGP occurs in approximately 1% of all renal infections. The kidney is usually non-functional. XGP displays neoplasm like properties capable of local tissue invasion and destruction and has been referred to as a pseudotumour. Adjacent organs including the spleen, pancreas or duodenum may be involved. The gross appearance of XGP is a mass of yellow tissue with regional necrosis and haemorrhage, superficially resembling renal cell carcinoma. Renal cell carcinoma may be indistinguishable from XGP radiographically and clinically. The treatment of XGP is almost universally extirpative and can pose a formidable challenge to the surgeon.

BACKGROUND

Xanthogranulomatous pyelonephritis (XGP) is a rare condition which closely mimics a malignancy.

It displays neoplasm-like properties capable of local tissue invasion and destruction and has been referred to as a pseudotumour. Adjacent organs including the spleen, pancreas or duodenum may be involved.

The treatment of XGP is almost universally extirpative and can be extremely challenging to the surgeon. It is important to make a correct diagnosis preoperatively, as nephrectomy in this condition is very difficult, particularly by the laparoscopic approach.

We were able to diagnose the condition correctly by the clinical presentation and CT images. Given the propensity of this condition to invade surrounding structures, we electively opted for the open approach (despite our expertise in laparoscopic approach) and were proven right after we found the kidney to be densely adherent to surrounding structures like spleen and pancreas.

Figure 1 Plain skiagram kidney/ureter/bladder region showing a large obstructing stone at pelvi-ureteric junction with multiple secondary stones.

Figure 2 CT urogram showing functioning right kidney, multiple left renal stones with non-excretion of contrast and per cutaneous nephrostomy tube in left kidney.

Figure 3 CECT (transverse section) showing multiple left renal stones with extensive perinephric stranding. A, aorta; D, duodenum; I, inferior vena cava; L, liver; LK, left kidney; P, pancreas; RK, right kidney.

We are submitting the CT images in which classical features of XGP can be seen, these can be valuable teaching tools to correctly identify XGP preoperatively by CT images. Moreover, the cut open excised specimen also highlights the various macroscopic features of this rare disease.

CASE PRESENTATION

A 56-year-old hindu male presented with dull aching left flank pain and low-grade febrile episodes since 6 months. He had generalised malaise and weakness.

He was of average built and nutrition and belonged to average socio-economic status. There was no significant medical, surgical or family history.

INVESTIGATIONS

Routine blood investigations showed anaemia with leucocytosis and elevated erythrocytic sedimentation rate. Serum creatinine was marginally elevated (1.4 mg%) and urinalysis demonstrated pyuria and proteinuria.

A plain skiagram of kidney/ureter/bladder region revealed a large obstructing stone at pelvi-ureteric junction with multiple secondary stones in the left kidney (figure 1). An ultrasonogram (USG) revealed an enlarged left kidney harbouring multiple stones with multiple hypoechoic masses, irregular thinned parenchyma, and a dilated collecting system with internal echoes suggestive of pyonephrosis.

An USG-guided per cutaneous nephrostomy was done and some pus was aspirated.

One week after inserting nephrostomy tube, we subjected the patient to a contrast enhanced CT (CECT) scan of abdomen and pelvis with delayed images taken up to 24 h after administering contrast. It confirmed the findings

of plain skiagram and demonstrated a non-functioning left kidney (figure 2). The kidney showed perinephric stranding suggestive of extensive inflammation (figure 3). There were multiple pockets of pus collection and loss of fat planes between the kidney and surrounding structures like pancreas and spleen indicating adhesion to these structures (figures 4 and 5). There was also encasement of left renal artery by inflammatory reaction (figure 6).

DIFFERENTIAL DIAGNOSIS

Locally invasive renal malignancy.

TREATMENT

On observing the presence of internal echoes in left kidney on USG, we decided to perform a per cutaneous nephrostomy. An USG-guided per cutaneous nephrostomy was done under local anaesthesia.

On puncturing the kidney, frank pus was aspirated. We placed a 8 Fr. nephrostomy tube which initially drained some pus, but could not drain entire pus collection in the kidney as it was multiloculated. A culture of this pus from the kidney revealed infection by pseudomonas, appropriate antibiotics were started.

A CECT scan of abdomen and pelvis 1 week after inserting nephrostomy tube demonstrated a non-functioning left kidney and a decision was taken to remove the left kidney.

We decided to perform a nephrectomy by open method. A laparoscopic approach was ruled out considering the severe inflammation and adhesion to surrounding structures. The patient was placed in the flank position and a 12th rib cutting incision was made. The kidney was densely adherent to surrounding structures.

Figure 4 CECT (transverse section) showing multiple pockets of pus collection and loss of fat planes between the kidney and surrounding structures like pancreas and spleen indicating adhesion to these structures. A, aorta; C, coeliac trunk; L, liver; LK, left kidney; P, pancreas; RK, right kidney; S, spleen.

Figure 5 CECT (sagittal section) showing multiple pockets of pus collection and loss of fat planes between the kidney and surrounding structures like pancreas and spleen. Per cutaneous nephrostomy tube is also seen. LK, left kidney; P, pancreas; PCN, per cutaneous nephrostomy; S, spleen; St, stomach.

Figure 6 CECT (coronal section) showing left kidney adherent to spleen and encasement of left renal artery by dense inflammatory reaction.

Figure 7 Left renal fossa after nephrectomy, adrenal can be seen.

It was mobilised by meticulous dissection circumferentially. The renal vessels were ligated en-masse as it was not possible to dissect at the hilum due to inflammation (figure 6). The kidney was then removed leaving the adrenal behind (figure 7). On excising the kidney, it was found to be full of pus which was multiloculated and had multiple stones (figures 8–10). Since the pus collection was multiloculated, it could not be drained by inserting a nephrostomy.

A drain was placed and the wound was closed in layers.

OUTCOME AND FOLLOW-UP

The postoperative course was uneventful. Oral feeding was resumed the next day morning and the patient was

Figure 8 Left kidney specimen with pus exuding.

ambulated. The drain was removed on the second postoperative day and the patient was discharged the next day. Stitches were removed on the seventh postoperative day.

The histopathology confirmed the diagnosis of xanthogranulomatous pyelonephritis based on pathognomonic microscopic finding of lipid-laden foamy macrophage.

DISCUSSION

Xanthogranulomatous pyelonephritis (XGP) is a rare, serious, debilitating illness characterised by an infectious renal phlegmon.¹ It was first described by Schlagenhauser in 1916.

XGP is defined as a chronic inflammatory disorder of the kidney characterised by a destructive mass that invades the renal parenchyma. Most cases of XGP are unilateral and are

often associated with urinary tract obstruction, infection, nephrolithiasis, diabetes, and/or immune compromise. XGP occurs in approximately 1% of all renal infections. The condition is most commonly associated with *Proteus*

or *Escherichia coli* infection. *Pseudomonas* species have also been implicated.

Most cases of XGP involve a diffuse process; however, up to 20% are focal. The kidney is usually non-functional. The treatment of XGP is almost universally extirpative and can pose a formidable challenge to the surgeon.

XGP is four times more common in women than in men and is usually noted in the fifth and sixth decades of life. XGP affects both kidneys with equal frequency.

Although XGP is rare in the paediatric population, it is found in approximately 16% of paediatric nephrectomy specimens. In children, XGP is more common in boys and usually affects those younger than 8 years. Obstruction associated with XGP in the paediatric population is more due to congenital factors than obstructive calculi.

Patients with XGP often appear chronically ill. Symptoms include anorexia, fever, chills, weight loss and flank pain. The pain of XGP is not colicky in nature; it is usually dull and persistent. Urine typically contains both leucocytes and bacteria. The pH is often basic because *Proteus mirabilis* is a urease-producing organism. The erythrocyte sedimentation rate is frequently elevated.

XGP displays neoplasm like properties capable of local tissue invasion and destruction and has been referred to as a pseudotumour. Adjacent organs including the spleen, pancreas or duodenum may be involved. The gross appearance of XGP is a mass of yellow tissue with regional necrosis and haemorrhage, superficially resembling renal cell carcinoma.

XGP is notorious for fistulisation. Pyelocutaneous and ureterocutaneous fistulae have been well-described. Other organs are occasionally involved in this process, including surrounding viscera with resulting pyeloenteric fistulae.

Figure 9 Left kidney cut open along Brodel's line revealing multiple pus pockets and stones.

Figure 10 Cut open left kidney with multiple pus pockets and stones. Also note thickening of edges indicative of extensive inflammation.

Renal cell carcinoma may be indistinguishable from XGP radiographically and clinically.² A case of XGP involving thrombus of the renal vein has been reported.³ XGP and renal cell carcinoma have even been observed in the same specimen. Shah *et al* have reported a mistaken diagnosis of renal XGP that was subsequently proven to be renal tuberculosis.⁴ XGP must be diagnosed based on histology rather than based solely on radiographic imaging studies.

The pathognomonic microscopic feature is the lipid-laden foamy macrophage accompanied by both chronic and acute-phase inflammatory cells. Focal abscesses may be observed.

Illustrating the varied presentation of XGP, a recent case report by Hitti *et al* describes XGP located in a renal allograft.⁵ Another recent report describes a case of XGP associated with psoas abscess in a young pregnant woman in her third trimester.⁶

It is important to note that a CECT scan is not a reliable method to assess kidney function.

Generally, if the kidney does not show function on intravenous urogram or CECT and if hydronephrosis or pyonephrosis is present, then it should be decompressed by inserting a per cutaneous nephrostomy. After adequate decompression one should get a renal scan after 4–6 weeks and then only decide the future course of action.

This case, however, was different. USG showed presence of pyonephrosis along with thinned out renal parenchyma (both features suggestive of advanced renal damage). An attempt was made to drain the pus by inserting a nephrostomy tube. Thereafter the patient was subjected to a CECT scan with delayed images taken over 24 h. It showed multiple pus pockets and complete lack of function even after 24 h. Since the pus was multiloculated, it was not possible to drain it by inserting a nephrostomy tube. Moreover, a kidney with multiple pus pockets will invariably be non-functioning on a renal scan. This was the reason the renal scan was omitted.

Learning points

- ▶ If the kidney on imaging studies shows an obstructing stone at pelvi-ureteric junction along with multiple secondary stones with the possibility of multiloculated pyonephrosis and is non-excreting with a severe peri nephric inflammation resulting in adhesions to surrounding structures, then a provisional diagnosis of xanthogranulomatous pyelonephritis must be strongly entertained.
- ▶ In case of XGP, the treatment is almost universally extirpative.
- ▶ Nephrectomy in a case of XGP can be very difficult more so by the laparoscopic approach. In such cases, there should be no hesitation to adopt the open approach to remove the kidney.
- ▶ A per cutaneous nephrostomy in a case of XGP will not be able to drain all the pus because more often than not XGP has multi loculated pus collection.

Competing interests None.

Patient consent Obtained.

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