Learning from errors

Xanthogranulomatous pyelonephritis in a young postpartal female

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Summary

Xanthogranulomatous pyelonephritis (XGP) is a rare chronic inflammatory disorder of the kidney characterised by an infectious phlegmon arising in the renal parenchyma. It is seen in patients who have urolithiasis, urinary tract infection and immunocompromised status. The clinical presentation is variable and renal neoplasm is considered as a differential due to its characteristic extrarenal visceral invasion. The treatment is almost universally extirpative and can pose a formidable challenge to the treating physician and surgeon. The authors report a rare case of XGP in a postpartal woman who presented with multiple visceral abscesses whose diagnosis was arrived on histopathological examination.

BACKGROUND

Xanthogranulomatous pyelonephritis (XGP) is a rare but life-threatening renal infection. Its presentation and management though is usually surgical; in our patient it presented with multiple intra-abdominal abscess, which misled us to a diagnosis of abdominal tuberculosis. In the Indian subcontinent, where such cases are rampant and treated as tuberculosis, diagnosing XGP is not common. We want to stress the need of keeping such differentials so that timely life-saving procedure can be undertaken successfully.

CASE PRESENTATION

A 22-year-old married woman presented to the emergency medical services of a tertiary hospital with progressive abdominal distention and continuous dull aching pain in the left hypochondrium for 2 months. The patient also had low grade fever, weight loss and loss of appetite since then. She had never had a history suggestive of tuberculosis or its contact, urinary tract infection or obstruction. There was a history of delivering a stillbirth baby vaginally 2 months ago in a rural hospital without instrumentation followed by hospital admission for 5 days. Obstetrics history was uneventful. There was not any history of past ailments or high risk behaviour.

On examination she was emaciated, afebrile, with a radial pulse of 90/min and blood pressure of 104/70 mm Hg. Pallor and grade I clubbing was noted. There was diffuse tenderness on deep palpation of the abdomen more marked in the left hypochondrium. A 4 cm soft and tender splenomegaly with 3 cm soft non-tender hepatomegaly was felt. Left kidney was ballotable and horseshoe-shaped dullness was present over the abdomen. Systemic and gynaecological examination was unremarkable.

INVESTIGATIONS

Investigations revealed a haemoglobin of 7.3 g% and leucocytosis of 32 400/mm³, which were predominantly polymorphonuclear. The erythrocyte sedimentation rate

was raised (–35 mm) at the end of the first hour. Liver and renal function tests were within normal limits. Urinalysis showed 2–3/hpf pus cells and red blood cells. No organisms or casts were seen. She was subjected to abdominal paracentesis and about 800 cc pale yellow hazy fluid was drained. Routine and microscopic examination showed 588 white blood cells/mm³, 70% lymphocytes, 30% polymorphs, proteins 4 g%, sugars 76 mg%, lactate dehydrogenase 6340 IU/l and serum albumin ascitic gradient 0.9 g/dl. On Gram staining, plenty of pus cells, Gram positive cocci and Gram negative bacilli were seen. No acid fast bacilli (AFB) were seen on Ziehl–Neelsen staining. HIV ELISA was negative and absolute CD4 count was 997 cells/μl. The tuberculin test read an induration of 15×20 mm. Serum protein electrophoresis, Brucella antigen and antinuclear antibody were negative. Contrast-enhanced CT (CECT) abdomen (figure 1) revealed the left kidney was grossly enlarged (16×8.5×5.4 cm) with pyonephrosis and perinephric collections. There was no excretion of contrast with a 1 cm terminal ureteral calculus. A 5.5×8×2.5 cm splenic infarct, moderate hepato-splenomegaly with gross ascites and necrotic retroperitoneal lymph nodes were seen.

On the basis of imaging lesions she underwent fluoroscopy guided percutaneous nephrostomy (PCN) insertion for left pyonephrosis. About 20 cc of yellow coloured, turbid fluid was drained, which on examination was full of pus cells without organisms but with raised adenosine deaminase levels (–744 IU/l).

DIFFERENTIAL DIAGNOSIS

Tuberculous peritonitis with visceral abscess; secondary peritonitis with organisation to multiple visceral abscesses; nephrolithiasis, obstructive uropathy left pyonephrosis; and XGP.

TREATMENT

Suspecting abdominal tuberculosis, the patient was started on four-drug antituberculous treatment (isoniazid,

rifampicin, pyrazinamide, ethambutol) with intravenous amikacin and metronidazole. Her PCN drain was removed on day 4. With this treatment for 4 weeks her ascites, pain in abdomen and constitutional symptoms improved. But contrasting our diagnosis of abdominal tuberculosis her ascitic fluid, PCN drain fluid and urine culture were serially positive for *Escherichia coli* and negative for AFB; hence, an alternative diagnosis was sought.

OUTCOME AND FOLLOW-UP

After 6 weeks, her CECT of the abdomen was repeated (figure 2), which showed a non-functioning left kidney

with a rent in the cortex and a 5.5×2×3.9 cm hypodense lesion communicating with the splenic mass and the tail of the pancreas with bulky pancreas. Similar lesion extending up to the pouch of Douglas and left adnexa were noted. Coordinated evaluation with surgical and radiology services led us to consider the diagnosis of XGP. After confirming a non-functioning left kidney on a diethylene triamine pentaacetic acid (DTPA) scan she underwent elective laparotomy of the left nephrectomy with debridement of the phlegmon. The diagnosis was confirmed on histopathological examination (figure 3). Antitubercular drugs were discontinued and at 3-month follow-up the patient is

Figure 1 Axial post contrast-enhanced CT abdomen image prior to percutaneous nephrostom insertion. The big arrow shows enlarged left kidney, pyonephrosis with perinephric collections, without excretion of contrast and posterolateral splenic infarct. The small arrow shows necrotic retroperitoneal lymph nodes.

Figure 2 Gross section of the postnephrectomy specimen of left kidney showing complete loss of architecture replaced with a mass of yellow tissue with multiple necrotic areas, haemorrhages and focal abscesses.

Figure 3 Light microscopic view of a stained section showing the pathognomonic lipid-laden foamy macrophage—that is, 'xanthoma cells'. Also acute and chronic inflammatory cells with loss of nephrons are seen.

in complete health with repeat ultrasonography showing clearance of previous lesions.

DISCUSSION

XGP is a life-threatening chronic inflammatory disorder of the kidney characterised by destructive mass invading the renal parenchyma. It may extend to the adjacent viscera displaying neoplasm-like properties capable of local tissue invasion and destruction; hence, referred to as a 'pseudotumour'.¹ First described by Schlagenhauser in 1916, more than 400 cases have been reported in the literature.² It occurs in approximately 1% of all renal infections and is four times more common in women than men and usually noted in the fifth and sixth decades. Paediatric incidence is rare. Most cases are unilateral and right sided, although fatal bilateral disease has been reported.³ Involvement can be focal or diffuse accounting for 12.5% and 87.5% cases, respectively, with the latter form usually leaving the kidney non-functional.⁴ The exact aetiology is unknown, but long-term renal obstruction and added infection is accepted as a cause. Staghorn calculi may be seen in 75–86% of these patients, which was the case in our patient, but are not required to make the diagnosis.⁵ Congenital urinary obstruction in the paediatric age group makes it more frequent in boys. Association with *Proteus*, *E Coli* and, sometimes, *Pseudomonas* has been noted with positive urine cultures in 30–40% of cases.⁶ XGP is often seen in patients with diabetes, immunocompromised individuals and abnormal lipid metabolism.

Patients appear chronically ill with anorexia, weight loss, fever, chills, dull persistent flank pain, dysuria and frequency. Due to its notoriety for fistulisation it can present as pyelocutaneous or pyeloenteric fistula. Some rare presentations as diverticulitis, psoas abscess and renal vein thrombosis too have been reported in the literature. CECT is a reliable

means to diagnose and establish the presence and extent of extrarenal involvement. The CT findings of XGP are calculi (75–86%), increased renal size (55–60%), pyelonephrosis (80%), hypodense areas with density representing focal areas of parenchymal destruction filled with pus and rim of enhancement, and extrarenal involvement in 15–20% cases making neoplasia as a close differential.⁵ Although ultrasonography, excretory urography, nuclear scintigraphy can be used, CECT abdomen is the investigation of choice with accuracy in the diagnosis of the entity. Still, XGP must be diagnosed on histology rather than solely on imaging studies as close differentials like renal tuberculosis and neoplasia demand different management.⁴ Grossly it appears as a mass of yellow tissue with regional necrosis, haemorrhages and focal abscesses resembling renal cell carcinoma superficially. The pathognomonic microscopic feature is the lipid-laden foamy macrophage accompanied by chronic and acute inflammatory cells.

Medical treatment alone is inadequate to treat XGP; antibiotics are a temporary measure for patients requiring medical investigation prior to nephrectomy. Total nephrectomy is the gold standard of treatment for XGP unless in bilateral cases where partial nephrectomy is performed with success.⁷ They can at times pose a formidable challenge to a surgeon particularly if there is local organ involvement where the aim is to remove all the involved granulomatous tissue. If unaccomplished, the remaining infected tissue may lead to progression and cutaneous fistulae.⁸ Our patient underwent left nephrectomy and debridement of the phlegmon under antibiotic cover, which resulted in complete recovery.

In conclusion, although rare as a diagnosis, XGP should be considered as a differential of renal masses especially if involving the other abdominal viscera. Prompt diagnosis and surgery can limit spread and progression of organ failure.

Learning points

- ▶ Xanthogranulomatous pyelonephritis is a life-threatening renal infection requiring timely diagnosis and surgical intervention.
- ▶ It can have varied presentation mimicking neoplasm.
- ▶ Rarely can it present as multiple intra-abdominal visceral abscesses, which need to be differentiated from a common diagnosis like abdominal tuberculosis or secondary peritonitis with visceral abscess.
- ▶ Timely surgical intervention is life saving and curative.

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REFERENCES

1. **Goldman SM**, Hartman DS, Fishman EK, *et al*. CT of xanthogranulomatous pyelonephritis: radiologic-pathologic correlation. *AJR Am J Roentgenol* 1984;**142**:963–9.
2. **Tolia BM**, Illoreta A, Freed SZ, *et al*. Xanthogranulomatous pyelonephritis: detailed analysis of 29 cases and a brief discussion of atypical presentations. *J Urol* 1981;**126**:437–42.
3. **Eastham J**, Ahlering T, Skinner E. Xanthogranulomatous pyelonephritis: clinical findings and surgical considerations. *Urology* 1994;**43**:295–9.
4. **Osca JM**, Peiro MJ, Rodrigo M, *et al*. Focal xanthogranulomatous pyelonephritis: partial nephrectomy as definitive treatment. *Eur Urol* 1997;**32**:375–9.
5. **Kim JC**. US and CT findings of xanthogranulomatous pyelonephritis. *Clin Imaging* 2001;**25**:118–21.
6. **MattaceRaso D**, Autorino R, Schiavo M, *et al*. Xanthogranulomatous pyelonephritis. Our experience and review of the literature. *Minerva Urol Nefrol* 2000;**52**:173–8.
7. **Peréz LM**, Thrasher JB, Anderson EE. Successful management of bilateral xanthogranulomatous pyelonephritis by bilateral partial nephrectomy. *J Urol* 1993;**149**:100–2.
8. **Chuang CK**, Lai MK, Chang PL, *et al*. Xanthogranulomatous pyelonephritis: experience in 36 cases. *J Urol* 1992;**147**:333–6.

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