Case report

Xanthogranulomatous pyelonephritis with associated renal cell carcinoma

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SUMMARY

Xanthogranulomatous pyelonephritis is associated with obstruction, stones and infection. CT is the mainstay of diagnosis, but appearances can mimic other conditions, including renal cell carcinoma. Nephrectomy is commonly recommended, but conservative treatment with antibiotics has been described after tissue diagnosis. We present a case of xanthogranulomatous pyelonephritis with concomitant renal cell carcinoma, which was an association that was suggested in 1988 and supported by subsequently reported cases. Conservative management of biopsy or cytology proven xanthogranulomatous pyelonephritis is unsafe, as an area of synchronous malignant tumour may be missed: we recommend it only in patients unfit for nephrectomy.

BACKGROUND

Xanthogranulomatous pyelonephritis (XGP) is a chronic granulomatous inflammation which destroys renal parenchyma, replacing it with a chronic inflammatory infiltrate, first described by Schlagenhauser in 1916.¹ Its propensity for mimicking other inflammatory and neoplastic renal pathologies such as renal cell carcinoma (RCC), leiomyosarcoma, malakoplakia and megalocytic interstitial nephritis is well established,² earning it a reputation as 'the great imitator'.³ The disease is more common in females.⁴ XGP is commonly treated by nephrectomy, but conservative management with antibiotics is described after tissue diagnosis with biopsy or cytology.⁵ We present a case of XGP associated with RCC in a patient operated on for suspected renal cancer, which led to diagnostic confusion when the patient suffered postoperative infection. Following review of the literature, we suggest an association between these conditions. We advise that needle biopsy or aspiration cytology proven XGP may not rule out the presence of RCC, and advise careful case selection for conservative treatment.

CASE PRESENTATION

A 72-year-old woman presented with a 2-month history of intermittent fever and rigours with urinary hesitancy, and two-stone weight loss. She was previously fit and well save for a history of duodenal ulcer and a previous transient ischaemic attack, had never smoked and had no history of chemical exposure nor any family history of urological malignancy.

INVESTIGATIONS

Ultrasound scan demonstrated an 8 cm heterogeneous vascularised mass in the mid to upper pole of the left kidney, suggestive of RCC, with dilatation of the collecting system. Staging CT confirmed this (figure 1), and suggested invasion of the renal sinus fat and renal vein. She was discharged on a course of ciprofloxacin for urinary tract infection and, following multidisciplinary team discussion, was listed for surgery for suspected T3aN0M0 RCC.

DIFFERENTIAL DIAGNOSIS

The working diagnosis at this stage was RCC, with the possibility of superadded infection, or of fever occurring as a paraneoplastic syndrome. Differential diagnoses of such a mass include other renal tumours such as renal adenoma, oncocytoma, angiomyolipoma, renal metastasis of a distant tumour, renal lymphoma, multilocular cystic nephroma and the rare solitary fibrous renal tumour. Renal pseudotumours include haemorrhagic or complex cysts, abscesses and inflammation including XGP, and renal infarct. An invasive adjacent tumour can also be mistaken for RCC, such as a transitional cell carcinoma, or renal tumour.

Figure 1 Staging CT showing mass in left upper pole.

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TREATMENT

Laparoscopic radical nephrectomy was performed with no immediate complications, but she did not recover as expected. On postoperative day 3, chest radiograph showed left lung base shadowing, so antibiotics were given for suspected pneumonia. Temperatures and leucocytosis settled, and she was discharged 7 days postoperatively. On postoperative day 10 she was readmitted with fever and rigours, and CT demonstrated a left retroperitoneal localised collection, with lung base atelectasis. CT-guided drain was placed, and she recovered slowly after a second drain and prolonged antibiotics. Blood cultures before initiation of antibiotics grew *Escherichia coli*, drain fluid during antibiotic therapy had no growth.

OUTCOME AND FOLLOW-UP

Nephrectomy specimen was found to contain a clear cell RCC of overall Fuhrman grade 3, with no tumour necrosis or sarcomatoid areas. It extended into an endothelium-lined space within the renal sinus (pT3a), and surgical margins were clear. There were areas of xanthogranulomatous inflammation adjacent to the tumour (figure 2), extending into the renal sinus and calyces. Superimposed on this were areas of suppurative acute inflammatory reaction involving the peritumorous tissue, extending into the surrounding venous channels.

At the time of follow-up she had recovered well from her operation and the postoperative infections she experienced.

Figure 2 Pathological specimen and 5× objective magnification H&E stain showing renal cell carcinoma (RCC) on left and xanthogranulomatous pyelonephritis (XGP) on right, with dividing band of fibrotic tissue.

Learning points

- ▶ Xanthogranulomatous pyelonephritis is an uncommon disease which may be indistinguishable from renal cell carcinoma on ultrasound or CT imaging.
- ▶ Obstruction, most commonly by stones, is an aetiological factor in development of xanthogranulomatous pyelonephritis.
- ▶ Renal cell carcinoma and xanthogranulomatous pyelonephritis may coexist, possibly owing to obstruction of renal calyces by tumour.
- ▶ While conservative treatment of biopsy proven xanthogranulomatous pyelonephritis with antibiotics has been described, one should consider the possibility of dual pathology.
- ▶ In this patient with an initial presentation suggestive of infection, a higher index of suspicion for xanthogranulomatous pyelonephritis might have led us to extend antibiotics beyond surgical prophylaxis for a few days, and potentially averted the postoperative collections and infections she experienced.

DISCUSSION

The aetiology of XGP is unclear, but it is associated with chronic obstruction and infection. Urolithiasis and obstructive conditions such as ureteropelvic junction syndrome, duplex collecting system and bladder tumours may predispose, as well as chronic inflammation such as interstitial nephritis.² Misdiagnosis is common, as its clinical and radiographic features may mimic pyelonephritis, tuberculosis, perinephric abscess and RCC. Preoperative diagnostic rates of 22%–30% have been quoted in case series, nearly half of cases have a mass suggestive of RCC on imaging.² Adding to the diagnostic challenge, tissue specimens from XGP may resemble clear cell RCC, and it may be difficult to distinguish between them on frozen section.⁶

Elliott *et al* reported the first case of XGP with synchronous RCC in 1968,⁷ and 14 such cases are previously reported.^{3 8–17} The suggestion that XGP may be contributory to the development of tumours was made as early as 1988.¹⁸ While it is possible that in our patient's case the two pathologies evolved separately (a collision lesion), we suspect that the xanthogranulomatous inflammation was secondary to obstruction of the pelvicalyceal system by tumour. As can be seen from our specimen (figure 2), a biopsy or cytology needle could easily have sampled xanthogranulomatous inflammation without revealing the cancer: this is to our knowledge the first case to illustrate this possibility.

Despite advances in imaging and diagnostic tests, XGP is no less deceptive today than 30 years ago. Our case illustrates how preoperative imaging can distract attention from clinical signs of an inflammatory process, causing diagnostic confusion. It is also the first to demonstrate from pathology specimens that a biopsy reporting XGP might not rule out concomitant RCC. The association is now well documented, and we advise caution in suspected cases of either pathology: it may be one, the other, or both. Due to this possibility of synchronous malignancy we recommend against conservative management of XGP and nephrectomy should remain the principal treatment in 30 patients fit for surgery.

Contributors BFM performed the literature search and drafted the manuscript. He was involved with the patient care as an inpatient, and sought her consent for publication. LP reported the pathological specimens, provided photographs of these, advised on manuscript descriptions of the pathology found, proofread and

approved the manuscript. AC was the consultant responsible for the patient care as an inpatient, advised on and approved the manuscript and communicated with the patient to obtain a signed consent form. MK supervised the development of the manuscript, advised on literature search and approved the final manuscript.

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