CASE OF THE MONTH

Cystic renal mass in a patient with previous Wilm's tumour

¹P DILKS, MRCP, FRCR, ¹E HELBREN, MRCS, FRCR and ²A SOHAIB, MRCP, FRCR

¹St Bartholomew's Hospital, West Smithfield, London, UK, and ²The Royal Marsden Hospital (London), London, UK

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Radiology

A 35-year-old female presented to her general practitioner with a 1-month history of moderate to severe continuous right flank pain associated with anorexia and weight loss. In the past, she had undergone partial right nephrectomy for a Wilm's tumour at the age of 8 months and was closely followed up for 16 years without recurrence.

On clinical examination there was a palpable mass in the right flank extending to the right iliac fossa. Initial work-up revealed a normochromic normocytic anaemia, normal white cell count, normal urea and creatinine, and mildly elevated C-reactive protein of 48. Glomerular filtration rate measured 92 ml min^{-1} , with the right kidney contributing 20% function on dimercaptosuccinic acid scintigraphy. A mid-stream specimen of urine had no white cells, although cultured *Escherichia coli*.

CT (Figure 1) and MRI (Figure 2) imaging was performed.

What was the diagnosis?

Figure 1. Single axial contrast-enhanced CT image at the level of the right renal hilum.

Address correspondence to: Dr Phillip Dilks, Speciality Registrar, Diagnostic Imaging, St Bartholomew's Hospital, West Smithfield, London EC1A 7BE, UK. E-mail: Philip.dilks@bartsandthelondon.nhs.uk

Figure 2. Axial MRI images from (a) T_2 weighted and (b) post-contrast T_1 weighted fat-suppressed sequences.

Imaging findings

Figure 1 shows a single axial contrast-enhanced CT image at the level of the right renal hilum, demonstrating diffuse reniform enlargement and parenchymal thinning of the right kidney. Peripherally, a mixture of cystic change and non-enhancing tissue is seen. No calcification is present. No extrarenal complications are demonstrated. The left kidney was normal. Figure 2 shows axial MRI images from T_2 weighted and post-contrast T_1 weighted fat-suppressed sequences. Multiple focal lesions within the right kidney are seen, including cysts and regions of non-enhancing tissue consistent with parenchymal destruction.

Diagnosis

The patient underwent a right radical nephrectomy. Histopathological analysis revealed a 12-cm mass comprising multiple cysts, some of which were filled with gelatinous material, and a 3-cm white/yellow solid component at one pole. There was a dense infiltrate of foamy histiocytes, lymphocytes and interweaving spindle cell proliferation. There was no evidence of necrotising granulomata, Wilm's tumour, nephroblastomatosis or renal cell carcinoma, and stains for acid fast bacilli and fungi were negative. The final diagnosis was xanthogranulomatous pyelonephritis (XGPN).

Discussion

In this case, making a diagnosis of XGPN based on imaging was challenging owing to the absence of visible obstruction or calcification, findings that are usually present in this disease [1–3]. There was no history of urinary tract infection or stones, and the coliform cultured from her urine had given no symptoms.

The differential diagnosis is wide and includes malignancy, such as renal cell carcinoma or lymphoma, and benign conditions, including bacterial pyelonephritis and necrotising granulomatous disorders such as tuberculosis. Given the history of a Wilm's tumour in childhood, nephroblastomatosis was considered. This

condition comprises multiple embryonic rests of tissue, often with concomitant cystic or dysplastic cortical malformations. It is a precursor condition for the development of Wilm's tumour [4].

Contrast-enhanced CT is the imaging modality of choice. The features of XGPN include diffuse reniform enlargement, parenchymal thinning, reduced perfusion and multifocal areas of fluid density, representing either dilated calyces or focal areas of parenchymal destruction containing pus or xanthogranulomata. There is usually evidence of urinary tract obstruction and nephrolithiasis [5].

In the multifocal form of nephroblastomatosis, areas of non-enhancing renal parenchyma are evident on contrast-enhanced CT, although renal enlargement is not usually a feature unless there is a concurrent Wilm's tumour [6]. Renal tuberculosis is usually unilateral and results in pericalyceal cyst formation, thickening and irregularity of the urothelium, debris within the collecting system and diffuse amorphous calcification. Ultimately it can progress to autonephrectomy. Bacterial pyelonephritis, on the other hand, is associated with wedge-shaped parenchymal defects or multiple masses and subsequent frank abscess formation [7]. Renal cell carcinoma most commonly manifests as an enhancing focal abnormality, often with areas of necrosis; however, the focal form of XGPN can often mimic malignancy. Renal lymphoma most commonly results in diffuse homogeneous reniform enlargement and does not typically result in focal areas of destruction.

In this case, the combination of diffuse reniform enlargement, cortical thinning and multifocal areas of parenchymal destruction despite the absence of calcification suggested XGPN as the most likely diagnosis.

Extrarenal involvement and the presence of complications are not uncommon and can be extensive. Imaging (particularly CT) can help to identify complications including haemorrhage, venous thrombosis, perirenal, psoas and hepatic abscess formation, and rarely nephrocutaneous and nephroenteric fistulation. Renal ultrasound may reveal renal enlargement, parenchymal and contour irregularity, obstruction and calcification. MRI can add further diagnostic value, including evaluation of the fat content and revealing the extent of involvement.

XGPN is an uncommon chronic destructive inflammatory disorder of the renal parenchyma characterised histologically by a chronic granulomatous infiltrate comprising xanthomatous histiocytes, lymphocytes, plasma cells and multinucleate giant cells [8, 9]. Presentation most commonly includes fever, abdominal pain, anorexia, weight loss, lower urinary tract symptoms and haematuria. Females are affected at least twice as commonly as males and presentation is often in the fifth or sixth decade. There is a usual association with long-standing obstruction, urolithiasis and bacterial (often coliform) infection. Unilateral involvement is the norm, although bilateral disease can occur and is often fatal. In addition, this entity is most commonly a diffuse process, although focal xanthogranulomatous involvement can occur [10] and management almost invariably involves nephrectomy.

In conclusion, XGPN can pose a diagnostic challenge but should be considered in the differential diagnosis in a patient with "cystic" renal tumour with a renal infection in the absence of nephrolithiasis.

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