

Xanthogranulomatous pyelonephritis: the case of a perplexing kidney

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DESCRIPTION

Case report

A previously healthy 2-year-old girl presented with turbid urine and pallor. She was otherwise asymptomatic and afebrile. Clinical examination revealed a large ballotable right-sided abdominal mass.

Ultrasound revealed an enlarged right kidney with loss of the normal renal architecture, which was replaced with multiple hypoechoic nodules. A number of renal calculi were observed with typical acoustic shadowing (figure 1). A contrast-enhanced CT scan demonstrated the diagnostic 'bear's paw' sign, multiple calculi, retroperitoneal lymphadenopathy and a small right psoas abscess (figure 2). A dimercaptosuccinic acid (DMSA) scan demonstrated a non-functioning kidney (figure 3). Urine cultures tested positive for *Proteus mirabilis*.

The patient initially underwent a percutaneous nephrostomy and received antibiotics. Subsequently, a radical nephrectomy was performed. Extensive inflammatory adhesions to the inferior vena cava,

liver and surrounding tissues were found. She recovered fully postoperatively and remains well.

Discussion

Xanthogranulomatous pyelonephritis is a chronic process characterised by suppuration, renal parenchymal destruction and lipid-laden foamy macrophages. It is rare in children.¹ CT is the mainstay for diagnosis. Typically associated with a staghorn calculus, the kidney shows loss of the normal cortex with multiple low attenuation areas that represent areas of renal tissue replaced with lipid-laden macrophages.² Percutaneous drainage and adjunctive antibiotic therapy prior to nephrectomy are recommended to avoid complications.³ Nephrectomy is usually curative and confirms the diagnosis histologically.

The role of imaging is to confirm the presence of an enlarged, non-functioning kidney, which in the presence of renal calculi and the bear's paw sign, is diagnostic.

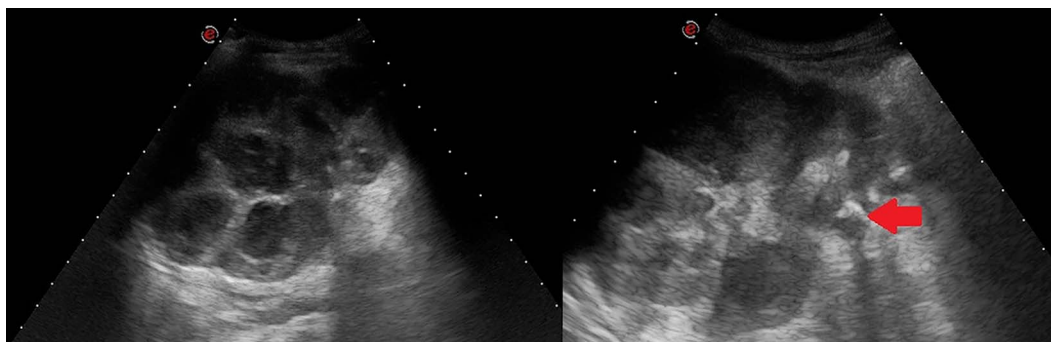


Figure 1 B-mode ultrasound images demonstrating an enlarged right kidney with multiple hypoechoic nodules and renal calculi demonstrating posterior acoustic shadowing (arrow).



Figure 2 Selected images of a contrast-enhanced CT of the abdomen. The typical 'bear's paw' sign is demonstrated (left arrow) together with the obstructing calculus (right arrow). The right psoas muscle is enlarged and enhances when compared with the left side in keeping with a psoas abscess (blue arrow).



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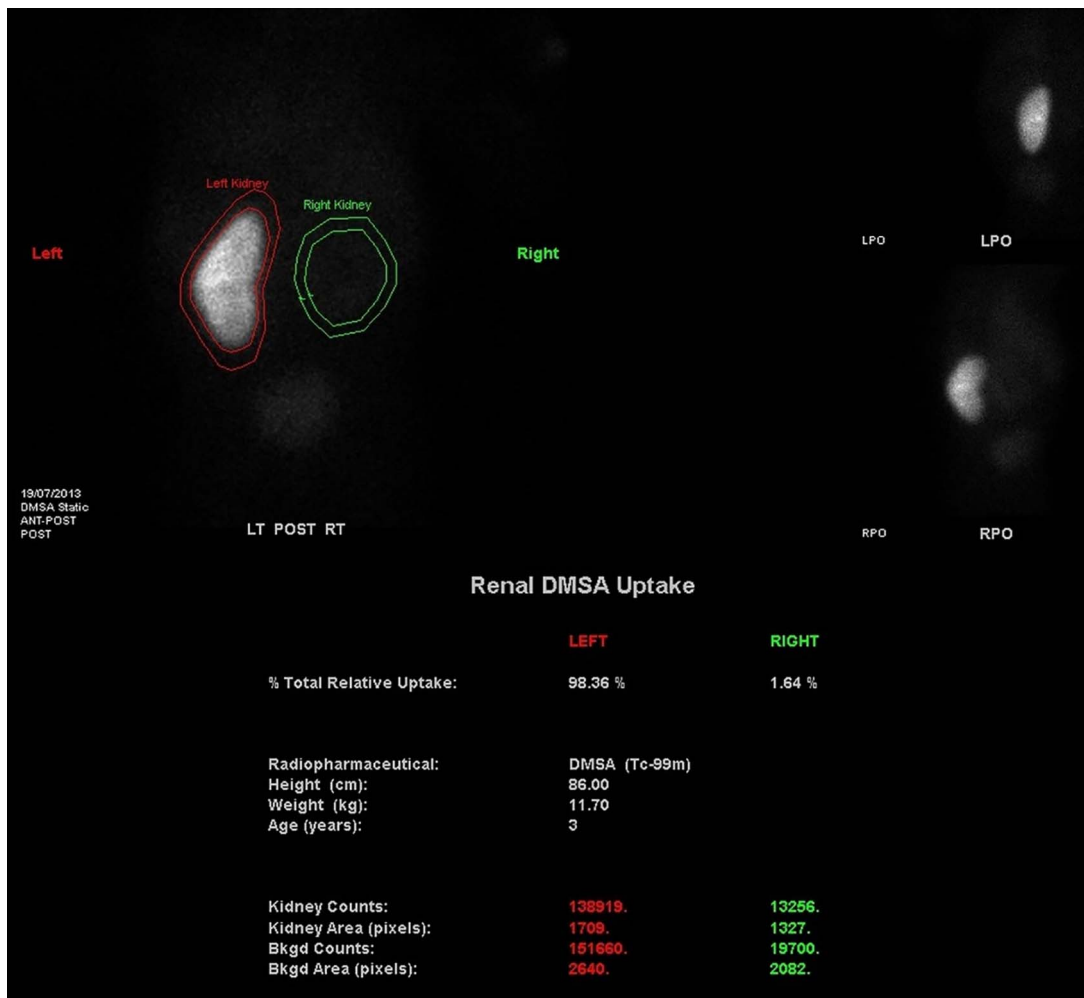


Figure 3 Dimercaptosuccinic acid scan demonstrating a non-functioning right kidney.

Learning points

- ▶ Imaging is key to the diagnosis.
- ▶ CT is the investigation of choice and demonstrates the typical bear's paw appearance.
- ▶ Nephrectomy is curative.

Contributors VSC and CA performed the literature review and drafted the write up of the article. CF and PC were involved in the clinical management of the patient.

Competing interests None.

Patient consent Obtained.

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